AMR Seminar #53 – Short Summary of Cases:

Case 1: M.67 with a mass in the head of the pancreas.

Case 2: F.72 with diarrhea, weight loss and abdominal, retroperitoneal and mediastinal lymphadenopathy.

Case 3: F.41 with soft tissue mass in chest wall.

Case 4: M.80 with tumor in the parotid gland.

Case 5: F.43 with multinodular retroperitoneal masses, 4-5 cm. in diameter.

Case 6: F.68 with fever of unknown origin and mediastinal adenopathy; subsequently developed a liver mass.

Case 7: F.86 with rapidly growing skin lesion on her nose.

Case 8: M.60 with large tumor (15 cm) in the prostate.

Case 9: F.37 with 2 cm. palpable nodule in left breast.

Case 10: M.64 with large, thin-walled presacral cyst.

Case 11: M.21 with small bowel obstruction and multiple intraabdominal masses.

Case 12: M.74 admitted with renal failure that rapidly deteriorarated with fatal outcome. At autopsy a polyp was found at the gastroesophageal junction.

Case 13: F.52 with splenomegaly and anemia.

Case 14: M.45 with 2.1 cm. testicular mass.

Case 15: F.30 with tumor of right frontal lobe who developed 17 months later lumbosacral pain and bone lesions.

Case 16: M.79 with 9cm. hemorrhagic tumor of the kidney.

Case 17: M.4 month old with a mass in the neck at the bifurcation of carotid vessels.

Case 18: F.55 with 12 cm. soft tissue mass in her arm, unrelated to the humerus.

Case 19: F.38 with tumor in the right lung.

Case 20: M.94 with right lower lobe lung lesion.

Case 21: F.17 with polypoid mass in anterior surface of endocervix.

Case 22: M.26 with retroperitoneal lymph node dissection for a previous testicular germ cell tumor.

AMR SEMINAR #53
CASE 1

Contributed by: Philip Allen, M.D. (ID Number: FMC 07/S06345)

Case referred by Dr. Sabine Ernsting, Flinders Medical Centre, South Australia.

History: Male aged 67. Pancreatic mass found on CT scan. The reason for the CT scan was not stated. A Whipple's resection was performed. The specimen consisted of 70 mm of duodenum and small bowel with included enlarged head of pancreas measuring 100 x 65 x 60 mm. The ampulla of Vater was patent. On cutting, there was a well circumscribed, multiloculated cystic mass filled with yellowish granular material suggestive of keratin (see photograph). The lesion was completely encapsulated and did not infiltrate the adjacent residual pancreas, small bowel or pancreatic duct. The uninvolved pancreatic parenchyma was grossly normal.

Diagnosis: Benign lymphoepithelial cyst, head of pancreas.

Comments: This is the first pancreatic lymphoepithelial cyst that I have seen. It arrived at about the same time as Doctor Adsay joined the club, so it more or less selected itself for this seminar as a welcoming present. There are only about 50 published cases. The mean age in Dr. Adsay's series was 56 years, pain is the commonest presenting symptom although a few have been discovered incidentally. They are more common in men, can occur anywhere in the pancreas, are sharply delineated from the surrounding pancreatic tissue and are cured by conservative resection. The pathogenesis is unknown. Hypotheses include epithelial inclusions in intrapancreatic lymph nodes; cystic transformation of pancreatic ducts; limited differentiation in a teratoma; an ectopic branchial cleft cyst, and a process whereby lymphocyte related growth factors stimulate epithelial proliferation, as may occur in Warthin's tumor. Perhaps Juan will be able to update us further on that last hypothesis, which he proposed in 1995. Small foci of sebaceous differentiation were present in some of the slides from this case, a feature Dr Adsay found in one of his 12 cases.

Reference: Adsay NV, Hasteh F, Cheng JD et al. Lymphoepithelial cysts of the pancreas: a report of 12 cases and a review of the literature. Mod Pathol 2002;15(5):492-501.

AMR SEMINAR #53
CASE 2

Contributed by: Carlos E. Bacchi, MD; CB 196/08

Clinical History: This is a 72-year-old female who presented with episodes of diarrhea and weight loss. Her episodes of diarrhea last 4-5 days with about10 bowel movements a day with no mucus or blood. She has intervals with no diarrhea. Blood cell count showed mild anemia and elevation of platelets with no blasts in the peripheral blood. Bone marrow biopsy hasn't been performed yet. Image studies revealed abdominal, retroperitoneal and mediastinal lymphadenopathy. Several lymph nodes from the retroperitoneal region were removed. They all showed the same histological picture. One of those lymph nodes was sent to the AMR members.

Pathology Findings: The histological section of the lymph node shows distortion of the general architecture. There is delicate sclerosis, vessel proliferation, eosinophils and clusters of pale or clear cells with oval or reniform ("monocytoid") nuclei. These cells sometimes have bilobed nuclei with cytoplasm finely granular. There is a tendency of these cells to centered on arterioles or be located in perifollicular distribution often mixed with eosinophils. Immunohistochemistry studies revealed expression by these clear cells of CD45, CD117 (c-kit) and tryptase with negativity for S-100 protein, CD1a, CD3, CD20, CD30, CD10 and CD15.

Diagnosis: Mastocytosis.

Comments: The major differential diagnoses in this case include lymphoma (particularly marginal zone B-cell lymphoma), T-cell lymphoma, hairy cell leukemia and reactive histiocytic proliferations. The associated eosinophils, endothelial vessel proliferation and sclerosis provide a strong clue that the pale cell clusters are most likely mast cells rather than lymphoma/leukemia cells. These histological alterations in the lymph node are probably related to the effects of mediators released by mast cells. Besides these morphological findings, the diagnosis of mastocytosis can be confirmed with immunohistochemistry staining, which shows expression in this clear cell population of mast cell markers including trypsin and CD117; this result associated with absence of immunostaining for B and T cell markers also helps ruling out B and T-cell lymphomas. Langerhans cell histiocytosis may also enter into the differential diagnosis because of the monocytoid appearance of the mast cell nuclei and tissue eosinophilia. However, the mast cells are smaller, and the grooved nuclei and voluminous cytoplasm characteristic of Langerhans cells are lacking. This case probably represents systemic mastocytosis but

I have no information about infiltration in other organs like bone morrow. Clinically, this patient doesn't show any skin lesion, hepatosplenomegaly or peripheral lymphadenopathy. Interesting to mention is that gastrointestinal symptoms like presented by this patient are common seen in systemic mastocytosis, mainly abdominal pain, diarrhea, nausea, vomiting and peptic ulcer. As expected, many of these symptoms are attributable to histamine release by mast cells. At this moment, the work up in this lady hasn't been extensively performed, so she may have infiltration in other organs by mast cells as well. Currently, there is no cure for systemic mastocytosis and the prognosis is variable depending on the disease category into which the patient belongs. A major prognostic feature in systemic mastocytosis is the presence or absence of skin involvement. Patients with cutaneous lesions often experience an indolent course, whereas those with no detectable skin lesions usually have more aggressive and progressive disease.

References

Horny, H. P., E. Kaiserling, et al. (1992). Lymph node findings in generalized mastocytosis. Histopathology **21**(5): 439-46.

AMR SEMINAR #53
CASE 3

Contributed by: Ira Bleiweiss, M.D.

History: The patient is a 41-year-old Oriental woman with a 4 cm posterior chest wall mass of unknown duration. The lesion was completely surgically excised and consisted of a well circumscribed firm pink-white mass measuring 4x3x3 cm.

Diagnosis: Alveolar soft part sarcoma.

Comments: Not a diagnostic dilemma. No immuno, etc. needed or done. I just thought the members could use a nice example of this very rare lesion.

AMR SEMINAR #53
CASE 4

Contributed by: John Chan, M.D. (#07AH19745)

Clinical History: An 80-year-old man underwent excision for a 6 cm skin tumor in the temporal region in July 2007. Two months later, he developed parotid enlargement. The specimen is from the parotid gland.

Special Studies: The small cell component is positive for cytokeratin 20, chromogranin and synaptophysin. The spindle cell component is extensively positive for actin, desmin and myogenin, but negative for cytokeratin 20, chromogranin and synaptophysin.

Diagnosis: Parotid gland – Metastatic Merkel cell carcinoma, with rhabdomyosarcoma differentiation (metastatic "Merkel cell carcinosarcoma").

Point of interest: This patient did have a Merkel cell carcinoma of the temporal skin. The primary tumor showed typical morphology and immunophenotype. The point of interest is the appearance of the metastatic tumor in the parotid gland.

(1) There is a recognizable component of Merkel cell carcinoma. In areas, there are rosettes formed by fibrillary material. This component exhibits the classical immunophenotype of Merkel cell carcinoma.

(2) In addition, there is an intermingled malignant spindle cell neoplasm reminiscent of embryonal rhabdomyosarcoma – rhabdomyoblasts with cross striations are not difficult to find. In areas, the sarcomatous component occurs in the absence of the Merkel cell carcinoma component. The immunostains show that this is indeed a rhabdomyosarcoma.

Saul Suster has written a very nice and comprehensive review on the morphologic spectrum of Merkel cell carcinoma (Ann Diagn Pathol 2006;10:376-385). In that review, he does illustrate rosette formation that can be seen in this tumor type. He also emphasizes the "plasticity" of the tumor, in that aberrant or heterologous differentiation can occur, such as squamous cell carcinoma, melanocytic differentiation, eccrine differentiation, and sarcomatous (such as leiomyosarcoma and rhabdomyosarcoma) differentiation.

In the literature, there are a few reports of rhabdomyosarcomatous differentiation in Merkel cell carcinoma (such as Fernandez-Figueras MT, J Cutan Pathol 2002;29:619-622; Eusebi V, Am J Surg Pathol 2000;24:223-230), but the rhabdomyosarcomatous component in these cases takes the form of single or small bundles of rhabdomyoblasts among the Merkel cell carcinoma. The current case differs in that there is separable, embryonal rhabdomyosarcoma-like component.

Members of the club may have a deja-vu feeling, because there is a lot of histologic similarity to the case of pulmonary small carcinoma with skeletal muscle and myofibroblastic sarcoma component in case 13 of AMR Seminar #52. The only differences are: Merkel cell carcinoma instead of pulmonary small cell carcinoma, and pure rhabdomyosarcoma in the spindle cell component.

AMR SEMINAR #5
CASE 5

Contributed by: Kum Cooper, M.D.

Clinical History: A 43-year-old woman presented with multinodular retroperitoneal masses, measuring 4-5 cm in diameter. Her significant past history is that she underwent embolization of her multi fibroid uterus about three years ago.

Diagnosis: Metastatic low grade endometrial stromal sarcoma.

Discussion: The features on the submitted sections from the retroperitoneal tumor show typical low-grade endometrial stromal sarcoma characterized by small cells and numerous blood vessels/arterioles. The highlight of this case is the presence of syncytial meningothelial-like whorls as an integral part of the tumor. Other unusual features include the starburst-like hyalinization and sex-cord-like differentiation. The subsequent hysterectomy showed similar features.

The "kicker" in this case is that the original needle core biopsies of the retroperitoneal tumor were dominated by the whorled pattern and this entertained a broad differential diagnosis. To the best of our knowledge, this feature in endometrial stromal sarcomas has only been described twice before, both as letters to the editor.

References:

1. Mooney et al. Histopathol 2006; 49:312-314.
2. Zamecnik and Sultani. Pathol Int 2007; 57:632-633.

AMR SEMINAR #53
CASE 6

Contributed by: Ivan Damjanov, M.D.

Clinical History: This 68-year-old woman presented with fever of unknown etiology in January, which receded on antibiotic treatment. The fever recurred 4 months later and a more detailed work-up disclosed mild one sided mediastinal lymphadenopathy. An extensive immunologic work-up disclosed no abnormalities. A biopsy was performed and the diagnosis of Hodgkin's lymphoma was made. She received the standard treatment, but three month after the diagnosis she developed jaundice and a rapidly growing hepatic mass was identified. The liver biopsy confirmed that the mass was Hodgkin's lymphoma. 11 months after the onset of first symptoms (fever) she died, presumably of liver failure. The material submitted for the AMR seminar is a mediastinal lymph node from the site where the primary diagnosis was made.

Diagnosis: Hodgkin's lymphoma.

Comment: Although we are not a major hematopathology center we see every year more than a few lymph nodes with Hodgkin's lymphoma. Nevertheless, I do not remember seeing Hodgkin's lymphoma with such a rapid downhill course. Obviously, the present patient was over 60 years of age, and the elderly with Hodgkin's lymphoma have a less favorable prognosis. The immunohistochemistry did not help us to answer this question. RS cells stained with antibodies to CD15 and CD30. In situ hybridization disclosed extensive EB virus reactivity. Do you see any histologic hallmarks in this slide that could have predicted such a poor prognosis?

AMR SEMINAR #53
CASE 7

Contributed by: Otto Dietze, M.D.

Clinical History: Rapidly growing skin lesion from the nose of an 86-year-old woman, suspicious of a keratoacanthoma.

Pathologic Findings: The lesion is completely excised and 3 transverse sections on the slide show a well-circumscribed centrally exulcerated tumor with a peripheral epidermal collarette. The tumor cells are arranged in broad fascicles with scanty stroma and show cytoplasma-rich predominately clear cells with several giant cells and mitotic activity including some atypical mitoses.
Immunostaining is positive with vimentin , CD68, CD99 and negative with different keratins, CD34, CD117, S-100, HMB-45 and F XIIIa.

Diagnosis: Clear cell atypical fibroxanthoma.

Comment: I did not see this type of AFX before and there are some cases in the literature e.g. a case report including EM and with a review of 7 other cases by Murali R and Palfreeman S (J Cutan Pathol 2006: 33: 343-348). AFX is considered by several authors as the superficial form of MFH, however, B.Zelger told me that to his experience AFX is a pleomorphic-anaplastic reaction, in many cases corresponding to a dedifferentiated squamous-cell carcinoma with loss of keratin expression.

AMR SEMINAR #53
CASE 8

Contributed by: Hugo Dominguez-Malagon, M.D.

History: A 60-year-old male with obstructive urinary symptoms, A large prostatic tumor measuring 15 cm was excised.

Histologically the tumor is composed of spindle to plump cells arranged in a patternless pattern, the nuclei show variation in shape and size, fine chromatin and up to 10 mitoses per 10 HPFs are seen in the more cellular areas, there are dense bundles of collagen. Tumor necrosis was seen in some areas (not shown in the slide).

Immunohistochemical stains were positive for CD34, Bcl-2, calponin (weak) and Ki67 (10% of the cells). Negative for S100.

Diagnosis: Solitary fibrous tumor of the prostate.

Discussion: Solitary fibrous tumor of the prostate is a rare neoplasm with nearly 20 cases published to date, mostly as case reports and a larger series or 12 cases. By immunohistochemistry: SFT in the prostate are positive for CD34 (>90%), Bcl-2 (88 to 100%), and CD99 (70 to 100%). However, CD34 is also expressed in a variety of spindle cell tumors involving the prostate including tumors of specialized stroma, GIST and peripheral nerve tumors. For that reason is important to use a complete panel in order to rule out other entities.

Criteria for malignancy: High cellularity with nuclear crowding and overlapping, increased mitotic rate (>4 per 10 HPFs), atypical mitoses, necrosis and hemorrhage, pleomorphism, fibrosarcoma-like growth pattern. Ki67 is not very useful to distinguish benign from malignant SFT of the prostate, expression of >20% can be seen in benign as well as malignant tumors.

However, these histological features do not necessarily correlate with malignant behavior, in pleura 50% of malignant SFTs are cured by simple excision. In 16 cases of SFT of the prostate reported by Herawi and Epstein, follow-up of 6/9 with borderline to malignant tumors, none recurred after follow-up from 1 to 10 years. The most difficult differential diagnosis of SFT is with specialized stromal tumors of the prostate. Stromal tumors of uncertain malignant potential (STUMP) are characterized by 1 of 4 patterns: (1) Scattered cytologically atypical cells associated with benign glands, (2) resembling glandular-stromal hyperplasia with hypercellular stroma, (3) extensive myxoid stroma, and (4) phyllodes pattern. In addition STUMP lack the dense ropy (wire-like) collagen and hemangiopericytoma-like pattern of SFT.

References.

1. Herawi M, Epstein J. Solitary fibrous tumor on needle biopsy and transurethral resection of the prostate. A clinicopathological study of 13 cases. Amk J Surg Pathol 2007;31:870-76.

2. Hansel DE, Herawi M, Montgomery E, Epstein J. Spindle cell tumors of the adult prostate. Mod Pathol 2007;20:148-158.

3. Herawi M, Epstein J. Specialized stromal tumors of the prostate: A clinipathologic study of 50 cases. Am J Surg Pathol 2006;30:694-704.

AMR SEMINAR #53
CASE 9

Contributed by: Vincenzo Eusebi, M.D.

Clinical History: A 37-year-old woman underwent fine needle aspiration cytology (FNAC) for a 2 cm palpable nodule located in the upper inner quadrant of the left breast. The diagnosis was poorly differentiated breast carcinoma. Frozen section was performed during definitive surgery and the intraoperative diagnosis of poorly differentiated malignant tumor was confirmed. A mastectomy was performed with axillary dissection. Residual tumour was fount around the FS biopsy, but the rest of the breast was non neoplastic. Axillary lymph nodes (12) were reactive.

Comment: I received the tumor in consultation and to make a long story short , immuno was negative for all type of keratins, EMA, actin, P63, ER, PR, GCDFP-15, HER2, CD 31, CD 34, HMB45, MART 1. And CD 68. Neoplastic cells were consistently positive for Vimentin, S-100 Protein and CD 21 (beautiful).

An extensive search did not reveal any tumour in other sites of the body. The case is recent and no FU is available. Probably chemotherapy is going to be administered.

In view of H&E findings and of the immuno results, the present tumour was diagnosed as follicular dendritic cell tumor primary in breast of which at least two cases have been published (3; 5). This specific tumour has to be distinguished from interdigitating dendritic cell sarcoma (one case only described (7)) as this latter is negative for CD 21 . Lymphoepithelial- like carcinoma(LEC) (2) can be identical to FDC, but fortunately LEC is positive for keratins and negative for CD 21. Extranodal FDC outside breast are not very common(6), often seen in association with Castleman disease(1; 4), vascular type. FDC behave aggressively being the recurrence free survival 27.4%(6) . Treatment so far is not standardized.

References:

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3. Fisher C, Magnusson B, Hardarson S, Smith MEF. Myxoid variant of follicular dendritic cell sarcoma arising in the breast. Ann Diagn Pathol 1999;3:92-98.

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5. Pruneri G, Masullo M, Renne G, Taccagni G, Manzotti M, Luini A, Viale G. Follicular dendritic cell sarcoma of the breast. Virchows Arch 2002;441:202-204.

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AMR SEMINAR #53
CASE 10

Contributed by: Cyril Fisher, Royal Marsden Hospital, London UK.

Clinical History: A 64-year-old male presented with long standing urinary symptoms. Imaging showed a large unilocular, thin walled cyst within the left mesorectum and extending to the left pelvic side wall. An additional finding was congenital absence of left kidney and suprarenal gland. The cyst contained numerous floating multifaceted spherical densities up to 1.5 cm in diameter. Section is from the cyst wall.

Pathology Findings: Macroscopically, the cyst was 14 x 11 x 10 cm and contained multiple yellow spheroids formed of compacted keratin laminae. (Fig 1). Histologically, the lesion is a dermoid cyst lined by keratinizing squamous epithelium with adnexa. There is extensive pagetoid infiltration by adenocarcinoma cells, in places with glandular lumen formation, and focally extending into intradermal eccrine ducts. The cells are immunoreactive for AE1/AE3 and, in contrast to the surrounding squamous epithelium, for CK7 (Fig 2), EMA, CEA, and GCDFP (Fig 2, inset). They are non-immunoreactive for CK5/6, CK14, CK20, CDX2, PSA, TTF-1 and S100 protein.

Diagnosis: Presacral (retrorectal) dermoid cyst with Paget's disease in a patient with possible Currarino syndrome.

Comment: Submitted as a change from soft tissue tumors, this case presents no diagnostic difficulty but is of interest because of the rare combination of findings. The retrorectal (presacral) space can be the site of dermoid cysts, which can be interpreted as mature teratomas. In this site they are most frequently diagnosed in infancy, and are rare with an incidence of between 1 per 30,000 to 43,000 live births.[1] In the adult, they occur mostly in middle-aged females and are usually asymptomatic; malignant change in such lesions is rare but invasive adenocarcinoma[2]and neuroendocrine tumor[3] have been described.

In this case, the immunophenotype of the carcinomatous infiltrate is consistent with Paget's disease of extramammary type. The clinical and radiological findings did not reveal an associated internal malignancy, and this was interpreted as an in-situ version of an apocrine carcinoma rather than pagetoid spread from another local or distant neoplasm. Three cases of extramammary Paget's disease have been reported within mature cystic (monodermal) teratomas of the ovary[4-6] and one case has been described in a retroperitoneal teratoma.[7] However, eextramammary Paget's disease arising in a presacral dermoid cyst appears to be novel and the report of this case will appear soon in AJSP.

Currarino syndrome is an inherited (autosomal dominant) genetic disorder comprising a triad of presacral mass, partial sacral agenesis and anorectal defects.[8, 9] The presacral mass may be a teratoma, dermoid cyst, hamartoma, neuroenteric cyst, anterior meningomyelocele or a combination of these. Clinical manifestations of the syndrome are variable. Not all components of the triad need be present and the spectrum of additional manifestations includes renal, urinary tract and gynecologic anomalies.[3] The syndrome is often subclinical, with up to 33% of patients being asymptomatic, and it can appear in adult life.[10, 11] This patient had congenital absence of the left kidney and adrenal gland. One case of single pelvic kidney in association with the Currarino anomaly has been reported in a term neonate with teratoma arising from the conus medullaris.[10] However, in the current case there was no sacral abnormality on imaging, and no other stigmata of the complex were found. No family history is known. Conceivably, however, it might represent an incompletely expressed variant of the Currarino syndrome.

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AMR SEMINAR #53
CASE 11

Contributed by: Christopher Fletcher, M.D. (Case No. 56080)

Clinical History: A 21-year-old male presented with large bowel obstruction and was found to have multiple intra abdominal masses, one of which (attached to the serosa of the colon) was excised.

Diagnosis: Deciduoid malignant mesothelioma.

Comment: This case is such a nice example of this rare mesothelioma variant that I thought that members of our group would enjoy seeing it. Since first being described mainly in the peritoneal cavity of young women, this subset of mesotheliomas has also been described in older patients and in the pleura, although overall the peritoneum still seems to be more common (at least in my experience). When I first saw this lesion, I initially wondered about the possibility of PEComa (given the nesting and granular/clear cytoplasm); however, immunostains showed strong positivity for keratin, EMA, calretinin and WT-1, leaving no doubt as to the diagnosis. As one might imagine, these lesions are generally associated with a poor outcome.

AMR SEMINAR #53
CASE 12

Contributed by: Andrew Folpe, M.D.

Clinical History: A 74-year-old male presented to the Emergency Department with a recent onset of vomiting and diarrhea. He was found to be in acute renal failure, which rapidly progressed to multiorgan failure and death. An autopsy was performed. The submitted slide is from a 2.5 cm mass present at the gastroesophageal junction.

Pathological Findings: The submitted section from the GE junction mass shows a submucosal tumor, composed of bland spindled to stellate cells, embedded in a myxohyaline matrix. A well-formed capillary-sized vasculature is present, as is a mixed inflammatory cell infiltrate, including eosinophils. Careful inspection of larger blood vessels in the submucosa of the stomach and within the submucosal mass reveals numerous highly atypical lymphoid cells, with enlarged, irregular, hyperchromatic nuclei and occasional prominent nucleoli. By immunohistochemistry, these cells were uniformly positive for CD20.

Diagnosis: Intravascular large B-cell lymphoma involving an inflammatory fibroid polyp.

Comment: As I am sure you have all surmised, the intravascular B-cell lymphoma (rather than the inflammatory fibroid polyp) was the cause of this patient's multiorgan failure and subsequent demise. The lymphoma was not suspected antemortem (validating once again the utility of the autopsy, even in sophisticated tertiary care centers!). Although neither inflammatory fibroid polyps nor intravascular lymphomas are terribly uncommon, I haven't ever seen this particular combination together on one slide, and I hope you will all appreciate having this curiosity in your slide collections.

AMR SEMINAR #53
CASE 13

Contributed by: Jeronimo Forteza Vila, M.D.

Clinical Findings: A 52-year-old woman presented with splenomegaly. Her laboratory examinations showed anaemia as the only significant finding. A splenectomy was performed. Grossly, the spleen showed a nodular pattern. Three years later, the patient presented with cervical lymphadenopathy. A cervical lymph node and a bone marrow biopsy were performed.

Diagnosis: Splenic marginal zone lymphoma with progression to a diffuse B cell lymphoma with overexpression of MYC.

Comments: Histologically, the spleen (slide A) showed a nodular infiltration of white and red pulp with tumor cells located at pre-existent germinal centers. There are two populations of neoplastic cells, one corresponding to small lymphocytes and the other to large cells with irregular nuclei and pale cytoplasm located towards the periphery of the nodules. Immunohistochemical study showed expression for CD20, bcl-2 and IgD in the tumor cells but not for cyclin D1. The differential diagnosis of this entity includes mantle cell lymphoma, follicle center lymphoma and hairy cell leukemia. Three years later, a lymph node (slide B) and a bone marrow biopsy were performed. Microscopically, both cases showed a neoplastic infiltration characterized by large cells, with prominent nucleoli. Immunohistochemical, expression for CD20, bcl.-2 and a high proliferation index. The translocation of MYC was detected by FISH. The differential diagnosis of this entity is with "Burkitt-like" lymphoma.

References:

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AMR SEMINAR #53
CASE 14

Contributed by: Allen Gown, M.D.

History: The patient is a 45-year-old male who presented with a 2.1 cm testicular mass. An orchiectomy was performed, and the enclosed slide is a representative H&E stained section of the tumor.

Diagnosis: Sertoliform adenoma of the rete testis.

Discussion: The histology of this tumor raises an interesting differential diagnosis, which includes sex cord stromal tumor, including Sertoli cell tumor. However, the immunophenotype of this tumor is not typical of either of these diagnoses:

Antibodies Specific To:	**Result:**
Cytokeratins [OSCAR, AE1./AE3])	Positive
Estrogen receptor	Positive
CD10	Positive
PAX2	Positive
S100 protein	Positive
WT-1	Positive
MART1	Positive
Calretinin	Negative
Inhibin alpha	Negative
OCT3/4	Negative
CD30	Negative
Prostatic specific antigen	Negative
Ki67 antigen	<1%

I was so very puzzled by the immunophenotype of this tumor, and I shared this with case with Dr. Mahul Amin, Cedars Sinai Medical Center, in Los Angeles, and it is he who suggested the diagnosis of sertoliform adenoma of the rete testis, and thought this tumor was quite similar to one he had seen. This is a rare tumor which manifests "bidirectional differentiation", both towards sex cord stromal (e.g., S100, MART1) as well as mesonephric (e.g., PAX-2, CD10) differentiation.

Only a handful of these tumors of the rete testis have been described, and referred to as adenomas, cystadenomas, or adenofibromas, in patients ranging from 12 to 51 years old; the lesions can be cystic, solid, or both. All were characterized by sertoliform tubules. (Altaffer LF et al., J Urol 127:332-, 1982; Jones MA and Young RH., J Urol Pathol 7:47-53, 1997; Murad T and Tanahashi T. Acta Pathol Jap 38:105-12, 1988). All previously reported cases had a benign outcome.

I wonder if members of this slide club agree with this diagnosis, or might suggest an alternative.

AMR SEMINAR #53
CASE 15

Contributed by: Janez Lamovec, M.D. (Case #433-03 (15a) and #3598-05 (15b))

History: A 30-year-old woman was admitted to the hospital because of a sudden epileptic attack of grand mal type. MRI of the brain revealed a tumor in the left frontal lobe. She has been operated upon, and the tumor was reportedly excised in its entirety. It was diagnosed as anaplastic oligoastrocytoma. She received no additional treatment and was in good health for eleven months. After that period she experienced attacks of sudden headache; a recurrent tumor was found on MRI; she was reoperated, and foci of residual tumor were confirmed histologically. Subsequently, she was irradiated and also received adjuvant chemotherapy. Seventeen months after second surgery she was admitted again because of severe lumbosciatic pain; two radiolucent lesions in the sacrum and diaphysis of the left femur were found. The drill biopsy of the latter lesion was performed. The specimen of bone biopsy was submitted to our laboratory; paraffin block of the recurrent brain tumor was referred to us from another institution.

Pathologic Findings: The major part of the bone specimen is represented by compact bone tissue and subperiosteal fibrous and osseous tissue; on the inner aspect of the specimen there is a dense population of oval to spindle cells, with moderately pleomorphic hyperchromatic nuclei and relatively abundant eosinophilic cytoplasm. Some mitoses may be seen. Immunohistochemically, tumor cells were negative for pankeratin, SMA and S-100 protein and positive for GFAP (see attached micro photo).

The recurrent brain tumor (we were not able to obtain the primary) is morphologically glioblastoma showing both astrocytic and oligodendrocytic, predominantly anaplastic components. Astrocytic component was GFAP positive, oligodendrocytic negative.

Diagnosis: Metastatic glioblastoma to bone.

Follow up: The patient's bone lesions were irradiated; 26 months after the latter treatment and 58 months after first craniotomy she shows no evidence of disease.

Comment: The metastases of malignant brain tumors to extraneural sites are very rare; among metastasizing tumor glioblastoma is relatively more frequently observed while metastases of oligodendroglioma are exceptional. The extraneural spread is supposed to be the consequence of craniotomy, tumor resection, radiotherapy and/or shunt operations, all of them enabling tumor cells to cross blood-brain barrier. However, spontaneous extraneural metastases without craniotomy are also possible. This possibility should be kept in mind, since young adults succumbing to malignant brain tumors are potential organ donors and cases of transmission of either glioblastoma or medulloblastoma with the transplanted organ are on record.
In our case, the metastasizing component of the original oligoastrocytoma was exclusively astrocytic, GFAP positive; the bioptic specimen was small and one can only speculate whether any remnants of oligodendrocytic component were also present in the metastasis.

I am submitting this case because of the extreme rarity of the event and to demonstrate how difficult it would be to render any specific diagnosis in the situation (quite possible) if no history of previous brain surgery were available. Unfortunately, in some slides there is very little tumor tissue left.

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4. Detry O, et al. Organ donors with primary nervous system tumors.Transplantation 70; 244-248. 2000

AMR SEMINAR #53
CASE 16

Contributed by: Michal Michal, M.D., (M6514/03)

History: 78-year-old male. Hemorrhagic tumor of the kidney, diam. 9 cm.

Immunohistochemical Examination: racemase (AMACR) +, AE1-AE3 +, EMA +, CD 10+, vimentin +.

Ultrastructure: cytoplasm of the tumorous cells was packed by numerous mitochondria.

FISH: Fluorescent in situ hybridization analysis on formalin fixed-paraffin embedded tissue showed three or more signals for chromosome 7 and 17, signals of chromosome Y was absent.

Diagnosis: Oncocytic papillary renal cell carcinoma (1).

Comment: This neoplasm is one of the eosinophilic tumors of the kidney, which is usually difficult to be distinguished from renal oncocytoma and chromophobe cell carcinoma. The typical features are: 1) it is composed of cells with morphological, immunohistochemical and ultrastructural features of oncocytes; 2) shows frequently nuclear grade 3 as type 2 papillary RCC; 3) can rarely develop metastases as type 1 papillary RCC; 4) shows trisomies of chromosomes 7 and 17 and loss of chromosome Y as the other types of papillary RCC; 5) expresses racemase as both type 1 and type 2 papillary RCC; 6) is more frequent in males rather than females as both type 1 and type 2 papillary RCC; 7) in difficult cases FISH on formalin fixed-paraffin embedded material could be a useful diagnostic tool.

Even if one of the papers describing identical tumors could not find the trisomies of chromosomes 7 and 17 typical of ordinary papillary carcinoma (2), another paper published last month found them including loss of chromosome Y (2/4 cases) (3) and confirmed our results. Interestingly, our paper was submitted for publication to Am J Surg Pathol in the first half of 2005, held by the reviewer for over half year a and then rejected. Even if published in 2006, it was written and submitted for publication prior to the other paper (2).

References:

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2. Lefevre M et al. Adult papillary renal tumor with oncocytic cells: clinicopathologic, immunohistochemical, and cytogenetic features of 10 cases. Am J Surg Pathol 2005:29:1576-81
3. Kunju P et al. Papillary renal cell carcinoma with oncocytic cells and nonoverlapping low grade nuclei. Hum Pathol 2008:39:96-101

AMR SEMINAR #53
CASE 17

Contributed by: Michal Michal, M.D. (#66989/04)

Clinical data: The patient was a 4-month-old boy patient, who presented with a mass in the neck. The tumor proved to be located in the left bifurcation of carotid vessels, as established by clinical investigation and surgery. The tumor was 5x3x3 cm in size, elastic in consistency, it was of white color and endowed with a thin capsule. The tumor currently recurred in the same place. I have not seen slides from the recurrence.

The tumor was composed of several cell types. The predominant cell type was small round to fusiform dark blue cells with a small amount of inconspicuous cytoplasm, which were arranged in poorly formed alveoli. Focally, these dark blue cells acquired a greater amount of pale oxyphilic cytoplasm so that the alveoli formed rare rosettes. In other places, the dark blue cells formed distinct epithelial cords with gland-like formations. Some of these gland-like spaces contained small amount mucus, which stained PAS- and mucicarmine-positive. Another distinctive component of the tumors was a mesenchymal one. The mesenchymal areas appeared very benign and could be likened to a fibroma having a densely collagenous stroma, or they had spindle cells set in the myxoid background, rendering a myxoma-like appearance. Focally there were apparent transitions between the myxoid spindled areas and the dark blue cells or the borders of the two components were sharply demarcated. Relatively large areas were composed of mats of eosinophilic epithelioid cells, probably representing intermediate differentiation between the dark blue cells and the myxoid spindle-cell-shaped areas. All of the above cell types formed peculiar, variously concentric structures.

Another distinctive feature was ganglion cell differentiation, which were usually rare, but seen in several slides given to the AMR seminar. The ganglion cells were in different stages of development; they grouped together or appeared singly. Brisk mitotic figures, including atypical forms, were found only in the small cell component, whereas the mesenchyme components were devoid of any necroses and mitotic figures. All above types of neoplastic cells were negative for smooth muscle actin, calponin, desmin, HMB45, neurofilament protein, CD99/MIC2, Melan A, tyrosinase, serotonin, CD56, Melan A, GFAP and S-100 protein. Cytokeratin, synaptophysin, FLI1 protein and chromogranin antibodies reacted only in the primitive small round cells, while all the other components were cytokeratin negative. In contrast, vimentin stained all cell types but the small round cell component. EMA stained a small proportion of the small round cells. MIB1 reacted in 15 and 20% of the small round cell component, while it was positive in less than 1% of the spindle cell mesenchymal component. Ultrastructurally, the cells endowed with well-formed intercellular desmosomes and numerous cells with cytoplasm containing a varying amount of membrane-bound secretory granules were found. Some cells had cytoplasm packed with mitochondria. Ganglion cells were not sampled.

Using probe EWSR1 in both cases, yellow fusion signals were observed in more than 90 % of the nuclei, indicating tumors without the EWSR1 gene translocation. Analysis of chromosome 12p gain was negative in Case 1, too. Ratio 12p:CEP12 were 1.0.

Diagnosis: Primitive small cell tumor with epithelial, gangliocytic, neuroendocrine and mesenchymal differentiation.
Two cases of this unusual tumor were published recently (*International Journal of Surgical Pathology 2007:15:429-436*).

Comment: The small round cell component in our cases seems to represent an authentic epithelial differentiation, a conclusion supported by the presence of well-formed desmosomes, cytokeratin positivity and vimentin negativity and formation of epithelial cords with gland-like spaces containing PAS and focal mucicarmine positive mucus. Based on the presence of atypical mitoses, the considerable MIB1 proliferation rate and presence of necroses, it seems that the epithelial small round cell component is the one which proliferates, while the spindle cell mesenchymal and gangliocytic components seem to be senescent ones. Neuroendocrine and gangliocytic differentiation of the tumor and the young age of the patient suggests that we might deal with a sort of paraganglioblastoma. Of note, in this respect is the publication of "adrenocortical blastoma" of the adrenals by Molberg et al **(***Molberg K, Vuitch F, Stewart D, Albores-Saavedra J. Adrenocortical blastoma. Hum Pathol 1992;23:1187-90***)**. The adrenal medulla is very similar in histogenesis and function to paraganliocytic extra-adrenal tissues. Morphologically the tumor of Molberg et al has some similarities to our cases, namely the round cell epithelial-like component and spindle cell stroma, but immunohistochemically it differed in being cytokeratin and EMA negative and vimentin positive (including the epithelial-like component).

AMR SEMINAR #53
CASE 18

Contributed by: Markku Miettinen, M.D.

History: A 55-year-old-woman with a large, 12 x 7 cm tumor in the arm separate from the humerus. Radiologic studies show moderate calcification, especially in the central part of the tumor.

Diagnosis: Extraskeletal osteosarcoma.

Comment: This tumor shows a combination of unusual trabecular clusters, fibromatosis-like areas, as well as foci of osteocartilaginous-osteosarcomatous differentiation. S100 positivity is limited to cartilaginous foci. I have considered this an osteosarcoma variant, but note that cases like this have sometimes been considered (potentially) related to ossifying fibromyxoid tumor. Cannot be sure that this is the case, especially in the absence of areas of typical OFT. I am afraid this tumor is closer to high than low grade. Large tumor size and proximity to a neurovascular bundle (see slide) made it impossible to proceed in a limb salvage mode resulting in arm amputation. Would be interested to hear other opinions.

AMR SEMINAR #53
CASE 19

Contributed by: Cesar Moran, M.D.

Clinical History: 38-year-old woman presented with shortness of breath and thoracic pain. A radiographic examination disclosed the presence of a "tumor" in the right lung. Resection of the lesion was performed.

Diagnosis: Endometriosis of the lung.

AMR SEMINAR #53
CASE 20

Contributed by: James Strauchen, M.D.

History: The patient is a 94-year-old man presenting with a right lower lobe lung lesion. Past medical history was remarkable for a Clark level III malignant melanoma (1.3 mm) of the left arm resected five years previously. Sentinel lymph node biopsy had shown a single cluster of S-100 positive, Melan-A positive cells. This was interpreted as a focus of malignant melanoma by us, but as benign nevus cells at Memorial Sloan-Kettering, and no further therapy was recommended. Three years later enlarged left axillary lymph nodes developed. Left axillary lymph node dissection was performed, revealing a malignant spindle cell neoplasm, consistent with metastatic malignant melanoma, involving 2 of 12 lymph nodes. Immunohistochemical stains were not performed. Wedge resection of the right lower lobe lesion was performed.

Pathology: Sections show a lung lesion composed of plexiform branching vascular-like structures in an inflamed collagenous stroma. Immunohistochemical stains revealed these cells were positive for S-100 with negative staining for Melan-A and HMB-45. Scattered entrapped endothelial cells were positive for CD34 and FVIIIR.

Diagnosis: Angiotropic metastasis of malignant melanoma.

Comment: Angiotropic malignant melanoma refers to a pattern of infiltration of melanoma cells in and along the walls of blood vessels resulting in a phenomenon referred to as "extravascular migratory metastasis." It differs from vascular invasion in that the melanoma cells remain outside the endothelium and result in formation of an "angio-tumoral complex." The phenomenon of angiotropic malignant melanoma is considered to be under appreciated (certainly by me!) and is said to be not uncommon if one looks for it. In this case, the resemblance of the lesion in the lung to an epithelioid vascular tumor was great; however, the morphology and immunophenotype were identical to the previous metastatic malignant melanoma in the axillary lymph nodes. The patient subsequently developed a metastatic skin nodule with an identical histology.

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AMR SEMINAR #53
CASE 21

Contributed by: Paul Wakely, Jr., M.D.

History: A 17-year-old G0 woman presented with dysfunctional uterine bleeding for 8 months that was severe enough to require a transfusion. Conservative management with birth control pills decreased her bleeding. A polypoid mass arising from the anterior endocervix was eventually biopsied.

Pathology: The specimen was labeled "aborting cervical mass" and consisted of multiple tissue fragments & blot clot - 7 x 6 x 2 cm. in aggregate. Variable amounts of stromal proliferation, decidual metaplasia and endocervical gland dilatation are present. Well-developed muscular arteries are easily found at the base. Small foci of malignant small round cells are scattered within the endocervical stroma and in a subepithelial "cambium" layer just beneath endocervical glandular epithelium. I have placed at least 1 green dot on all slides to highlight these foci since they are easily overlooked (and to save you time). Some of these cells also exist in edematous stroma. Immunohistology showed these to be myogenin, myoglobin positive.

Diagnosis: Embryonal rhabdomyosarcoma [RMS] of the uterine cervix.

Comment: This case was originally diagnosed by one of our pathologists as an "inflamed decidual cast with Aria-Stella reaction." Another 6 months went by before a subsequent biopsy revealed the correct diagnosis. She was treated with chemotherapy and 11 months after this initial biopsy, a hysterectomy was performed. That specimen showed a single nodule of metaplastic hyaline cartilage, but no evidence of residual RMS.

Cervical RMS is rare. Any discussion of pure RMS in the gynecologic tract typically revolves around *vaginal* RMS which is almost exclusive to infants and children <5 years (x age = 1.8 yrs.). A recent study from Memorial Sloan-Kettering uncovered only 15 adults (≥ 16 years) with pure gynecologic RMS in a 40-year review. *Ferguson et al.* RMS in the cervix arises in older teenagers and young adults. Vaginal RMS is almost unheard of in this age group. Mean age for 13 patients with cervical RMS was 18 yrs., *Daya & Scully* for 21 patients was 17 yrs., *Brand et al.* 14.5 yrs., *Hays et al* and for 8 patients 42 yrs. of age. *Ferguson et al.* RMS seems to ascend in the gynecologic tract with each major transition period in female reproduction: vaginal RMS → infants/children, cervical RMS → adolescents/ pre-menopausal women, uterine RMS → postmenopausal period. With regard to the cervix, several references list leiomyosarcoma as the most common primary sarcoma with RMS next in line. When it does occur in adults, the cervix appears to be the most common site of RMS. *Ferguson, Daya, Zeisler* Typical clinical presentation is DUB, vaginal discharge, or "something" protruding from the introitus. Less frequent is the passing of tissue. Four cases of cervical RMS have been reported in association with ovarian Sertoli-Leydig tumor - significance unknown. Cartilaginous metaplasia has been reported in several cases – we saw it only in the post-chemoRx treated specimen of this case.

This case was adulterated by the prior administration of birth control pills for 8 months, and is more typical of a cervical polyp with thick walled vessels at the base, a rich vascular and fibrous stroma, decidualized LUS stromal cells, and focally dilated endocervical glands. Presumably because the foci of RMS were small and widely scattered, the diagnosis was overlooked, and it was not until 6 mos. later when another biopsy unveiled the correct diagnosis. If any club members have encountered an example(s) of cervical RMS, please comment on your case(s) particularly with regard to whether your example was straightforward RMS, or had a variety or other features making the diagnosis more challenging.

References:

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AMR SEMINAR #53
CASE 22

Contributed by: Lawrence Weiss, M.D., City of Hope, Duarte, CA

Short History: 26-year-old man with a retroperitoneal lymph node dissection for a germ cell tumor.

Long History: This 26-year old man presented one year prior with a 12 cm. right testicular mass. Radical orchiectomy was performed, revealing a malignant mixed germ cell tumor, with 70% yolk sac tumor, 25% immature and mature teratoma, and 5% embryonal carcinoma. The teratoma included a mesenchymal component consisting of fibrous tissue and lobules of mature-appearing cartilage. Although there was no evidence of angiolymphatic space invasion, the spermatic cord foci of mature teratoma. Staging showed extensive nodal disease as high as the right supraclavicular lymph nodes. He received 4 cycles of chemotherapy with standard PEB. Post-treatment, he again developed extensive nodal metastases as far up as the upper mediastinum. He underwent an eventful retroperitoneal lymph node dissection, which required vena caval resection, aortic resection, and over 80 units blood transfusion.

Gross: Pathology received multiple lymph node specimens, most of which showed mature cystic teratoma. However, the largest mass measured 17 cm. and had a 6.5 cm. fleshy area within what otherwise appeared to be a mature cystic teratoma.

Special Stains:	Keratin	negative
	S-100	negative
	CD99	weak cytoplasmic positive
	Desmin	focal positive
	Myogenin	focal positive
	GFAP	negative
	OCT-4	negative

Comment: There is a small round blue cell tumor adjacent to areas of mature cystic teratoma. This is extensive necrosis. Focal areas show clear-cut rhabdomyoblastic differentiation.

Diagnosis: Mature cystic teratoma with sarcomatous component (embryonal rhabdomyosarcoma)

Discussion: I received this case on the exact day I opened up my September issue of Am J Surg Pathol, and perused the outstanding paper on this topic by Drs. Malagon, Valdez, Moran, and Suster (Am J Surg Pathol 2007;31:1356-1362). I present this case not so much as a diagnostic dilemma, but more as a nice illustration to the recent paper. The patient is 4 months S/P his surgery, and has not yet been able to recover from the surgery enough to receive his planned chemotherapy for the embryonal rhabdomyosarcoma.