California Tumor Tissue Registry's
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"General Surgical Pathology"

Case 1

A 43 year old man with no significant past medical history was seen for right-sided chest pain and shortness of breath. A chest X-ray revealed a right pleural effusion and consolidation of the right lower lobe of lung. A CT scan showed an infiltrative mass extensively involving the diaphragmatic and costal margin of the pleura. A core biopsy showed a high-grade spindle cell neoplasm suspicious for a spindle cell sarcoma. An extrapleural pneumonectomy was performed. The specimen consisted of an extrapleural pneumonectomy with resection of the pericardium, right hemidiaphragm and right chest wall. The resection specimen showed a plaque-like thickening of the pleura that varied from 0.1 to 2.5 cm in thickness and involved the lower two-thirds of the anterior, posterior, medial, lateral and diaphragmatic pleura without involvement of the lung parenchyma. Positive for AE1/AE3 and focally for CK5/6, calretinin and SMA. Negative for CEA, MOC31, EMA, TTF1, CK7, CK20, S-100, desmin, H-caldesmon, HMB45, CD31, CD34, bcl-2, p63 and WT1. Mib-1 proliferation marker showed nuclear positivity in 50% of the tumor cells.

Dx: Spindle cell (sarcomatoid) malignant mesothelioma

(courtesy of Dr. Saul Suster, 123rd CTTR semi-annual seminar)

Suster S, Moran CA. Malignant mesothelioma: current status of histopathologic diagnosis and molecular profile. Exp Rev Mol Diagn 5:715-723, 2005.

Suster S, Moran CA. Applications and limitations of immunohistochemistry in the diagnosis of malignant mesothelioma. Adv Anat Pathol 13:316-329, 2006.

Alvarez-Fernandez E, Dieu-Nau MD. Malignant fibrosarcomatoid mesothelioma and benign pleural fibroma (localized fibrous mesothelioma) in tissue culture. Cancer 43:1658-1663.

Moran CA, Suster S, Koss MN. Smooth muscle tumors presenting as pleural neoplasms. Histopathology 27:227-234, 1995.

Nicholson AG, Goldstraw P, Fisher C et al. Synovial sarcoma of the pleura and its differentiation from other primary pleural tumors: a clinicopathological and immunohistochemical review of three cases. Histopathology 33:508-513, 1998.

Moran CA, Travis WD, de Christenseon M et al. Thymomas presenting as pleural tumors: report of 8 cases. Am J Surg Pathol 16:138-144, 1992.

Case 2

A 49 year old woman with no significant past medical history was seen for persistent cough. A chest X-ray revealed a spiculated right upper lobe mass. An endoscopic biopsy was carried out with a diagnosis of "undifferentiated carcinoma, favor small cell carcinoma". A right upper lobectomy was performed. Focal positivity for AE1/AE3 and MOC31, strong nuclear positivity for

p63. Negative for TTF1, chromogranin, synaptophysin, CEA, CK 5/6, monoclonal NSE, CD57, CK7 and CK20. MIP-1 showed 70% nuclear positivity.

Dx: Poorly differentiated squamous cell carcinoma, small cell variant
(courtesy of Dr. Saul Suster, 123rd CTTR semi-annual seminar)

Bernard WG. The nature of the "oat cell sarcoma" of the mediastinum. J Pathol Bacteriol 29:241-244, 1926.

Suster S, Moran CA. The Lung, in: Modern Surgical Pathology, 1st. Edition, Weidner N, Cote R, Suster S, Weiss LM (Eds), W.B.Saunders Co, Philadelphia, 2003, pp.350.

Carter D, Yesner R. Carcinomas of the lung with neuroendocrine differentiation. Semin Diagn Pathol 2:235-243, 1985.

Cerilli LA, Ritter JH, Mills SE et al. Neuroendocrine neoplasms of the lung. Am J Clin Pathol 116 (Suppl):S65-96, 2004.

Travis WD, Brambilla E, Muller-Hermelink HK, Harris C. Pathology & Genetics. Tumors of the Lung, Pleura, Thymus and Heart, WHO Classification of Tumors, IARC Press, Lyon, France, 2004, pp.31.

Brambilla E, Moro D, Velae D et al. Basal cell (basaloid) carcinoma of the lung. A new morphologic and phenotypic entity with separate prognostic significance. Hum Pathol 23:993-998, 1992.

Case 3

A 56 year old man was seen for shortness of breath and chest pain. A CT scan revealed a large anterior mediastinal mass. A video-assisted thoracoscopic biopsy with frozen section examination was read as a "small round blue cell neoplasm". Study of permanent sections showed scattered keratin-positive cells against a sea of lymphocytes favoring a diagnosis of thymoma. A total thymectomy was performed. The resected specimen consisted of a well-circumscribed, encapsulated tumor mass measuring 8.4 cm in greatest diameter. The cut section was gray-white, homogeneous and without areas of hemorrhage or necrosis. Positive for AE1/AE3, CK5/6 and CK19 with focal positivity for bcl-2 and p63. Negative for CD5, EMA, MOC31, SMA, S-100, vimentin, desmin and CD34.

Dx: Lymphocyte-rich spindle cell thymoma (WHO type AB)
(courtesy of Dr. Saul Suster, 123rd CTTR semi-annual seminar)

Bernatz PE, Harrison EG, Claggett OT. Thymoma: a clinicopathologic study. J Thorac Cardiovasc Surg 42:424-444, 1961.

Rosai J. Histological Typing of Tumors of the Thymus, (2nd. Ed), Berlin, Springer-Verlag, 1999, World Health Organization International Histological Classification of Tumors.

Travis WED, Brambilla E, Muller-Hermelink HK, Harris CC. Pathology and Genetics of Tumors of the Lung, Pleura, Mediastinum and Heart, World Health Organization Classification of Tumors, IARC Press, Lyon, 2004.

Suster S, Moran CA. Thymoma classification. Current status and future trends. Am J Clin Pathol 125:542-554, 2006.

Suster S, Moran CA. Problem areas and inconsistencies in the WHO classification of thymoma. Semin Diagn Pathol 22:188-197, 2005.

Chen G, Marx A, Wen-Hu C et al. New WHO histologic classification predicts prognosis of thymic epithelial tumors: a clinicopathologic study of 200 thymoma cases from China. Cancer 95:420-429, 2002.

Case 4

40 y/o man had a hx of high blood pressure and cramps in the legs. He also has had low serum potassium not responsive to K therapy. Electrolytes: Na 136, K 3.9, Cl 104, Co2 27. Ct scan revealed a small mass in the left adrenal gland, confirmed by adrenal venography. Aldosterone level in left renal vein was > 600 ng/dl, right renal vein was 24 ng/dl. A 2.2 cm encapsulated yellow mass was removed from the left adrenal.

Dx: Adrenal adenoma - Aldosterone producing

CT of primary hyperaldosteronism (Conn's syndrome): the value of measuring the adrenal gland. Lingam RK; Sohaib SA; Vlahos I; Rockall AG; Isidori AM; Monson JP; Grossman A; Reznek RH. AJR Am J Roentgenol 2003 Sep;181(3): p843-9.

Association between Crohn's disease and Conn's syndrome. A report of two cases. Astegiano M; Bresso F; Demarchi B; Sapone N; Novero D; Palestro G; Resegotti A; Pellicano R; Rizzetto M. Panminerva Med 2005 Mar;47(1): p61-4.

Primary hyperaldosteronism: effect of adrenal vein sampling on surgical outcome. Nwariaku FE; Miller BS; Auchus R; Holt S; Watumull L; Dolmatch B; Nesbitt S; Vongpatanasin W; Victor R; Wians F; Livingston E; Snyder WH. Arch Surg 2006 May;141(5): p497-502; discussion 502-3.

Elevated blood pressure linked to primary hyperaldosteronism and impaired vasodilation in BK channel-deficient mice. Sausbier M; Arntz C; Bucurenciu I; Zhao H; Zhou XB; Sausbier U; Feil S; Kamm S; Essin K; Sailer CA; Abdullah U; Krippeit-Drews P; Feil R; Hofmann F; Knaus HG; Kenyon C; Shipston MJ; Storm JF; Neuhuber W; Korth M; Schubert R; Gollasch M; Ruth P. Circulation 2005 Jul 5;112(1): p60-8.

Primary aldosteronism - changing concepts in diagnosis and treatment. Young WF. Endocrinology 2003 Jun;144(6): p2208-13.

Case 5

19 y/o male with a rapidly growing 2 cm mass on the left temple.

Dx: Nodular fasciitis, face

Nodular fasciitis of the cheek. Kamiya H; Araki M; Kitajima Y. Eur J Dermatol 2003 Mar-Apr;13(2): p189-91.

Clonal rearrangement of 15p11.2, 16p11.2, and 16p13.3 in a case of nodular fasciitis: additional evidence favoring nodular fasciitis as a benign

neoplasm and not a reactive tumefaction. Donner LR; Silva T; Dobin SM. Cancer Genet Cytogenet 2002 Dec;139(2): p138-40.

Fine needle aspiration in nodular fasciitis of the face. Matusik J; Wiberg A; Sloboda J; Andersson O. Cytopathology 2002 Apr;13(2): p128-32.

Dermal nodular fasciitis: three case reports of the head and neck and literature review. Nishi SP; Brey NV; Sanchez RL. J Cutan Pathol 2006 May;33(5): p378-82.

Nodular fasciitis. Saqi A; Nassar A; Baloch Z. Diagn Cytopathol 2002 Jun;26(6): p407-8.

Differential expression of smooth muscle myosin, smooth muscle actin, h-caldesmon, and calponin in the diagnosis of myofibroblastic and smooth muscle lesions of skin and soft tissue. Perez-Montiel MD; Plaza JA; Dominguez-Malagon H; Suster S. Am J Dermatopathol 2006 Apr;28(2): p105-11.

Case 6

Elbow mass from a 13 y/o boy. Surgeon indicated that the tumor was adherent to muscle. 2.2 cm in greatest diameter.

Dx: Juvenile Hyaline fibromatosis

The gene for juvenile hyaline fibromatosis maps to chromosome 4q21. Rahman N; Dunstan M; Teare MD; Hanks S; Edkins SJ; Hughes J; Bignell GR; SO. Am J Hum Genet 2002 Oct;71(4): p975-80.

Juvenile hyaline fibromatosis: morphologic, immunohistochemical, and ultrastructural study of three siblings. Haleem A; Al-Hindi HN; Juboury MA; Husseini HA; Ajlan AA. Am J Dermatopathol 2002 Jun;24(3): p218-24.

Juvenile hyaline fibromatosis: report of a case and comparison with infantile systemic hyalinosis. Lim AA; Kozakewich HP; Feingold M; Padwa BL. J Oral Maxillofac Surg 2005 Feb;63(2): p271-4.

Juvenile hyaline fibromatosis and infantile systemic hyalinosis overlap associated with a novel mutation in capillary morphogenesis protein-2 gene. Antaya RJ; Cajaiba MM; Madri J; Lopez MA; Ramirez MC; Martignetti JA; Reyes-Mugica M. Am J Dermatopathol 2007 Feb;29(1): p99-103.

Juvenile hyaline fibromatosis: a report of two unrelated adult sibling cases and a literature review. Senzaki H; Kiyozuka Y; Uemura Y; Shikata N; Ueda S; Tsubura A. Pathol Int 1998 Mar;48(3): p230-6.

Juvenile hyaline fibromatosis: ultrastructural study. Winik BC; Boente MC; Asial R. Am J Dermatopathol 1998 Aug;20(4): p373-8.

Case 7

39 y/o woman with a 6 month hx of a mass on her back. Past hx positive for breast lumpectomy, removal of an ovarian cyst and right foot surgery.

Dx: Atypical lipoma/well differentiated lipoma-like liposarcoma

Atypical lipoma as a potential pitfall in the cytodiagnosis of subcutaneous tumors. A report of two cases. Woyke S; Kapila K; Goswami KC. Acta Cytol 1997 May-Jun;41(3): p897-902.

Atypical subcutaneous fatty tumors. Challis D. Adv Anat Pathol 2000 Mar;7(2): p94-9.

Atypical subcutaneous fatty tumors: a review of 37 referred cases. Allen PW; Strungs I; MacCormac LB. Pathology 1998 May;30(2): p123-35.

Nonrandom pattern of telomeric associations in atypical lipomatous tumors with ring and giant marker chromosomes. Mandahl N; Mertens F; Willen H; Rydholm A; Kreicbergs A; Mitelman F. Cancer Genet Cytogenet 1998 May;103(1): p25-34.

Atypical lipomatous tumors with smooth muscle differentiation: report of two cases. Zamecnik M; Michal M; Sulc M. Pathology 1999 Nov;31(4): p425-7.

Case 8

A 28 year-old woman presented with a rapidly growing nodule on the right thumb. Radiographs taken prior to surgery demonstrated an ill-defined soft tissue mass without calcification or periosteal reaction. *(Note: this is the CTTR's Case of the Month for 12/08 at www.cttr.org).*

Dx: Fibroosseous pseudotumor of digit

Fibro-osseous pseudotumour of the digit--amputation for a benign but aggressive lesion. Coleman RA. Hand Surg [Br 2005 Oct;30(5): p504-6.

Chan KW, Khoo US, Ho CM. Fibro-osseous tumor of the digits: report of a case with immunohistochemical and ultrastructural studies. *Pathology* 25(2):193-196, 1993.

deSilva MV, Reid R. Myositis ossificans and fibroosseous pseudotumor of digits: a clinicopathological review of 64 cases with emphasis on diagnostic pitfalls. *Int J Surg Pathol* 11(3):187-195, 2003.

Dupree WB, Enzinger FM. Fibro-osseous tumor of the digits. *Cancer* 58: 2103-2109, 1986.

Fletcher CDM, Unni KK, Mertens F. World Health Organization Classification of Tumors. Pathology and Genetics of Tumours of Soft Tissue and Bone. IARC Press: Lyon 2002.

Nishio J, Iwasaki H, Soejima O, Naito M, Kikuchi, M. Rapidly growing fibro-osseous pseudotumor of the digits mimicking extraskeletal osteosarcoma. *J Orthop Sci* 7(3):410-413, 2002.

Sleater J, Mullins D, Chun K, Hendricks J. Fibro-osseous pseudotumor of the digit: a comparison to myositis ossificans by light microscopy and immunohistochemical methods. *J Cutan Pathol* 23(4): 373-374, 1996.

Case 9

Two cm mass in deep tissues of distal medial thigh in an 18 y/o male. Cytokeratin negative.

Dx: Localized deep granuloma annulare

Pseudorheumatoid nodules in adults: a juxta-articular form of nodular granuloma annulare. Barzilai A; Huszar M; Shpiro D; Nass D; Trau H. Am J Dermatopathol 2005 Feb;27(1): p1-5.

Localized granuloma annulare and autoimmune thyroiditis in adult women: a case-control study. Vazquez-Lopez F; Pereiro M; Manjon Haces JA; Gonzalez Lopez MA; Soler Sanchez T; Fernandez Coto T; Perez Oliva N; Toribio J. J Am Acad Dermatol 2003 Apr;48(4): p517-20.

Subcutaneous granuloma annulare in childhood: clinicopathologic features in 34 cases. Grogg KL; Nascimento AG. Pediatrics 2001 Mar;107(3):pE42.

Deep granuloma annulare (pseudorheumatoid nodule) in children: clinicopathologic study of 35 cases. McDermott MB; Lind AC; Marley EF; Dehner LP. Pediatr Dev Pathol 1998 Jul-Aug;1(4): p300-8.

HIV-associated granuloma annulare (HAGA): a report of six cases. O'Moore EJ; Nandawni R; Uthayakumar S; Nayagam AT; Darley CR. Br J Dermatol 2000 May;142(5): p1054-6.

Case 10

A 38 year old woman with no significant past medical history was seen for a slow-growing soft tissue mass in the subcutaneous tissue of her right hip. A wide excision was performed for the suspicion of malignancy. Positive for vimentin, CD34 and bcl-2. Negative for AE1/AE3, SMA, h-caldesmon, calponin, S-100, EMA, CD10 and CD117. MIB-1 stain showed nuclear positivity in 5% of tumor cells.

Dx: Solitary Fibrous Tumor of Soft Tissues, Right Hip

(courtesy of Dr. Saul Suster, 123rd CTTR semi-annual seminar)

Fletcher CDM. Hemangiopericytoma – a dying breed. Reappraisal of an "entity" and its variants: a hypothesis. Curr Diagn Pathol 1:19-23, 1994.

Gengler C. Gillou L. Solitary fibrous tumor and hemangiopericytoma: evolution of a concept. Histopathology 48:63-74, 2006.

Nappi O, Ritter JH, Pettinato G, Wick MR. Hemangiopericytoma: Histopathological pattern or clinicopathologic entity? Semin Diagn Pathol 12:221-232, 1995.

Fletcher CDM. The evolving classification of soft tissue tumors: an update based on the new WHO classification. Histopathology 18:3-12, 2006.

Guillou L, Fletcher CDM, Mandahl N. Extrapleural solitary fibrous tumor and hemangiopericytoma, In: World Health Organization Classification of Tumors, Pathology and Genetics of Tumors of Bone and Soft Tissue; Eds: Fletcher CDM, Unni KR, Mertens F; IARC Press, Lyon, 2002; pp.86-90.

Klemperer B, Rabin CB. Primary neoplasms of the pleura: a report of five cases. Arch Pathol 11:385, 1931-1936.

Moran CA, Suster S, Koss MN. The spectrum of histologic growth patterns in benign and malignant fibrous tumors of the pleura. Semin Diagn Pathol 9:169-180, 1992.

Suster S, Nascimento AG, Miettinen M, et al. Solitary fibrous tumor of soft tissue. A clinicopathologic study of 12 cases. Am J Surg Pathol 19:1257-1266, 1995.

Bonus Case 11 - courtesy of Dr. Resa Chase, Loma Linda VA Medical Center
(no answer required … look in appendiceal wall to see abnormality)

Abdominal surgery for a leiomyomatous uterus in a 35 y/o woman. This is from an incidental appendectomy. No history of her travels.

Dx: Schistosoma eggs in appendix (see schistosoma involving *gallbladder* in Sub C, vol1, case1)

Acute abdomen associated with schistosomiasis of the appendix. Gabbi C; Bertolotti M; Iori R; Rivasi F; Stanzani C; Maurantonio M; Carulli N. Dig Dis Sci 2006 Jan;51(1): p215-7.

Diagnosis of genital cervical schistosomiasis: comparison of cytological, histopathological and parasitological examination. Poggensee G; Sahebali S; Van Marck E; Swai B; Krantz I; Feldmeier H. Am J Trop Med Hyg 2001 Sep;65(3): p233-6.

The epidemiology of schistosomiasis in Egypt: summary findings in nine governorates. El-Khoby T; Galal N; Fenwick A; Barakat R; El-Hawey A; Nooman Z; Habib M; Abdel-Wahab F; Gabr NS; Hammam HM; Hussein MH; Mikhail NN; Cline BL; Strickland GT. Am J Trop Med Hyg 2000 Feb;62(2 Suppl): p88-99.

Schistosomiasis. Halkic N; Gintzburger D. Comment On: RefSource:N Engl J Med. 2002 Apr 18; 346(16):1212-20.

Appendicitis and infections of the appendix. Lamps LW. Semin Diagn Pathol 2004 May;21(2): p86-97

Human schistosomiasis. Gryseels B; Polman K; Clerinx J; Kestens L. Lancet 2006 Sep 23;368(9541): p1106-18.