1114

CALIFORNIA
TUMOR TISSUE REGISTRY

# STUDY CASES

# SEPTEMBER 1996

# "SOFT TISSUE TUMORS"

California Tumor Tissue Registry
c/o: Department of Pathology and Human Anatomy
Loma Linda University School of Medicine
11021 Campus Avenue, AH 335
Loma Linda, California 92350
(909) 824-4788
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**CONTRIBUTOR: Raymond Wuerker, M.D.**
**Long Beach, CA**

**CASE NO. 1 - SEPTEMBER 1996**

**TISSUE FROM: Posterior mediastinum**

**ACCESSION #26202**

**CLINICAL ABSTRACT:**

This 53-year-old Black male presented with a one year history of non-productive cough and mild shortness of breath and a three month history of mild malaise, chills and sweating. CT scan showed a 15.0 cm posterior mediastinal mass with central necrosis. The mass displaced the esophagus to the right and appeared to involve the aorta. The patient underwent thoracotomy.

**GROSS PATHOLOGY:**

The mass measured 14.0 x 11.0 x 7.0 cm, weighed 557 grams and had a 6 cm diameter necrotic central cavity.

SPECIAL STAINS:
S-100: positive Desmin: negative

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**CONTRIBUTOR: Loma Linda Pathology Group (drc)**
**Loma Linda, CA**

**CASE NO, 2 - SEPTEMBER 1996**

**TISSUE FROM: Left buttock**

**ACCESSION #27875**

**CLINICAL ABSTRACT:**

This 73-year-old Caucasian male patient presented with a lump on his left hip. The mass was resected.

**GROSS PATHOLOGY:**

The 245 gram, 12.0 x 9.5 x 4.5 cm specimen included superficial adipose tissue and deep skeletal muscle. Embedded within the resection specimen was a 5.0 x 3.5 x 4.0 cm white-tan well-circumscribed mobile mass.

**CONTRIBUTOR: Loma Linda Pathology Group (kt)** **CASE NO. 3 - SEPTEMBER 1996**
**Loma Linda, CA**

**TISSUE FROM: Right leg** **ACCESSION #27874**

**CLINICAL ABSTRACT:**

This 31-year-old Black male presented with a six month history of a painful mass in his right anterior thigh. This was resected but the mass recurred three months later. Physical examination showed a 25 cm long mass involving the right thigh to the patella. A right anterior compartment resection was performed.

**GROSS PATHOLOGY:**

The 2350 gram, 40.0 x 15.0 x 10 cm specimen included a 10.0 x 9.0 x 25.0 cm portion of irregular muscle with fascicles surrounded by dense fibrous tissue interlaced with fat. Within this were numerous vessels ranging from 0.2 to 0.3 cm in diameter.

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**CONTRIBUTOR: Loma Linda Pathology Group (drc)** **CASE NO. 4 - SEPTEMBER 1996**
**Loma Linda, CA**

**TISSUE FROM: Chest wall** **ACCESSION #27203**

**CLINICAL ABSTRACT:**

This 22-year-old Asian male presented with a 1 1/2 month history of a rapidly enlarging, tender mass in the right lateral chest wall. The mass was fixed to the 10th and 11th rib. A wide local excision was performed.

**GROSS PATHOLOGY:**

The 16 gram, 4.0 x 3.8 x 2.0 cm specimen included a well-demarcated 2.2 x 1.8 x 1.6 cm firm, bulging nodule surrounded by skeletal muscle. The mass had a glistening mucinous surface.

**CONTRIBUTOR: Solomon L. Riley, Jr., M.D.**
**Los Angeles, CA**

**CASE NO. 5 - SEPTEMBER 1996**

**TISSUE FROM: Thigh**

**ACCESSION #22007**

**CLINICAL ABSTRACT:**

This 15-year-old female presented with a one year history of vague lower thigh swelling. An excisional biopsy was performed.

**GROSS PATHOLOGY:**

The specimen was an 11.5 x 6.5 cm diameter mass.

SPECIAL STAINS
Keratin: ++ EMA: ++

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**CONTRIBUTOR: Usha Garg, M.D.**
**Oxnard, CA**

**CASE NO. 6 - SEPTEMBER 1996**

**TISSUE FROM: Chest wall**

**ACCESSION #23022**

**CLINICAL ABSTRACT:**

This 16-year-old male had a history of right chest discomfort, treated with antibiotics. Minor trauma produced a hemothorax and an infiltrative globular mass lesion was found in the right lung. It eventually progressed to minor discomfort, chest pain with shortness of breath, nausea and vomiting after minor chest trauma. The mass had increased in size on x-ray. A right thoracotomy found a 14 cm diameter tumor apparently arising from the 5$^{th}$ or 6$^{th}$ interspace and involving the lower lobe of the right lung. The tumor was resected.

**GROSS PATHOLOGY:**

The 360 gram specimen included a 13 cm diameter portion of lung and portions of two ribs firmly adherent to the visceral pleura. Within the lung parenchyma was a 6.2 cm diameter, well-encapsulated, hemorrhagic, focally necrotic mass.

SPECIAL STAINS:

| | |
|---|---|
| Chromogranin/ desmin/ actin/ S-100: | negative |
| NSE/ 013 (Ewing's sarcoma)/ vimentin: | positive |

**CONTRIBUTOR: Lilibeth Guinto-Miranda, M.D.** **CASE NO. 7 - SEPTEMBER 1996**
**Bakersfield, CA**

**TISSUE FROM: Chest wall** **ACCESSION #25370**

**CLINICAL ABSTRACT:**

This 79-year-old male presented with an enlarging right upper lateral chest wall mass growing beneath the superficial muscles, between the ribs and possibly into the pleura. The mass was located several inches below the axilla, and over the area of a recent, 7th rib fracture. An excisional biopsy was performed.

**GROSS PATHOLOGY:**

The 14.5 x 12.0 x 9.5 cm tumor with four attached ribs weighed 837 grams. The main bulk of the tumor was homogenous pink-tan "fish flesh" in appearance.

SPECIAL STAIN:

| | |
|---|---|
| CEA/ myoglobin: | diffusely strong |
| PAS (for glycogen): | positive |
| Vimentin: | ++++ |
| Desmin/ actin/ keratin/ S-100: | negative |

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**CONTRIBUTOR: Drs. Douglas Kahn/Rita Hopper** **CASE NO. 8 - SEPTEMBER 1996**
**Sylmar, CA**

**TISSUE FROM: Abdominal mass** **ACCESSION #27971**

**CLINICAL ABSTRACT:**

This 28-year-old female presented with sharp, crampy right lower quadrant abdominal pain for four days. An abdominopelvic mass was palpable just above the umbilicus. Ultrasound revealed a 16.0 x 8.0 x 15.0 cm predominantly solid mass emanating from the pelvis. Clinical and radiographic impression was that of an ovarian germ cell tumor. At exploratory laparotomy, the mass was adherent to the surface of the ovary, possibly arising from the fallopian tube, cul de sac, or omentum. There was no apparent retroperitoneal involvement.

**GROSS PATHOLOGY:**

The 416 gram, 24.0 x 22.0 x 15 cm specimen consisted of yellow necrotic and red-brown, firm friable material. A more viable appearing part of the mass was yellow-tan and measured 9.0 x 7.5 x 5.0 cm.

**CONTRIBUTOR: Jozef Kollin, M.D.**
**Lakewood, CA**

**CASE NO. 9 - SEPTEMBER 1996**

**TISSUE FROM: Right pyriform sinus**

**ACCESSION #27908**

**CLINICAL ABSTRACT:**

This 32-year-old white male presented with respiratory stridor and was found to have a laryngeal mass almost completely obstructing the airway. An excision of the right pyriform sinus mass was performed. The patient had a past history of an industrial accident 10 years ago with resultant brain damage. A tracheostomy was performed during his acute care but was later removed.

**GROSS PATHOLOGY:**

The 14 gram specimen consisted of multiple slightly gritty gray-tan tissue fragments up to 4.5 cm in greatest diameter, some of which were covered by mucosa..

SPECIAL STAINS:

| | |
|---|---|
| Epithelial membrane antigen/keratin/ S-100/ desmin: | negative |
| Muscle specific actin/ smooth muscle actin/ Alpha-1 antichymotrypsin: | negative |
| Vimentin/ CD-34: | positive |

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**CONTRIBUTOR: Frank Hirose, M.D.**
**Torrance, CA**

**CASE NO. 10 - SEPTEMBER 1996**

**TISSUE FROM: Thigh**

**ACCESSION #25407**

**CLINICAL ABSTRACT:**

This 38 year old man had a 20 year history of a right posterior thigh mass which developed a constant sharp burning pain, with radiation down the back of the thigh and calf into the foot. The mass was found to be associated with the right sciatic nerve and was resected.

**GROSS PATHOLOGY:**

The 775 gram specimen consisted of a 29 x 8 cm fusiform tubular structure. The distal end branched into 3 smaller trunks, measuring 2.5, 3.0 and 4.0 cm in diameter. The tumor was grossly contained within a pink-tan, focally hemorrhagic capsule. Cross sections revealed a multiple rubbery, gelatinous, tubular structures, measuring up to 3.0 cm in diameter. Within the central fusiform swelling was a 5.0 cm diameter area of necrosis.

SPECIAL STAINS:

| | | | |
|---|---|---|---|
| S-100: | negative | NSE: | strongly positive |

CALIFORNIA
TUMOR TISSUE REGISTRY

# MINUTES

# FOR

# SEPTEMBER 1996

# "SOFT TISSUE TUMORS"

**SUGGESTED READING (General Topics from Recent Literature):**


Hubbard Ruth and Lewontin RC. (Harvard University, Cambridge, MA) Sounding Board. Pitfalls of Genetic Testing. *N Eng J of Med* 1996; 334(18):1192-1193.

Mettlin CJ, Murphy GP, Ho R, and Menck HR. The National Cancer Data Base Report on Longitudinal Observations on Prostate Cancer. *Cancer* 1996; 77(10):2162-2166.

Nash G, Hutter R, and Henson DE. Practice Protocol for Examination of Specimens from Patients with Lung Cancer. *Cancer* 1996; 77(10):2167-2174.

Grove Andy. Taking on Prostate Cancer. *Fortune* 1996; 133(9):55-72.

Rosai J, Akerman, M, Cin, DP, DeWever I, Fletcher CDM, et al. Combined Morphologic and Karyotypic Study of 59 Atypical Lipomatous Tumors . *Am J Surg Pathol* 1996; 20(10):1182-1189.

Turbett GR and Sellner LN. A Comprehensive Guide to Pathology Resources Available on the Internet. *Adv in Anat Pathol* 1996; 3(5):313-335.


NOTE: The new CTTR WEB Page will be operational shortly. It will include images from John Brooks Semi-Annual Seminar on Soft Tissue Tumors, the address is http://www.llu.edu/LLU/CTTR


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INLAND (RIVERSIDE/SAN BERNARDINO) - Schwannoma/neurilemoma (3)

BAKERSFIELD (Central Valley Study Group) - Sarcoma probably leiomyosarcoma

PLEASANTON/FREMONT - Cellular schwannoma (3); Neurofibroma (3)

LONG BEACH - Neurosarcoma (7)

SANTA BARBARA (St. Francis Med Ctr) - Schwannoma (3)

SANTA BARBARA (Cottage Hospital) - Cellular schwannoma with increased mitoses (vs MPNST)

SANTA ROSA - Schwannoma, r/o malignancy (2); Malignant peripheral nerve sheath tumor (1)

BAY AREA - Malignant schwannoma (3)

SACRAMENTO (UC Davis Medical Center) - Cellular schwannoma

ARIZONA (VA Medical Ctr) - Neurilemoma (schwannoma)

COLORADO (Konopka/Chu Study Group) - Epithelioid smooth muscle tumor

KANSAS (Univ of Kansas) - Schwannoma

NEBRASKA (Creighton University) - Neurilemoma (schwannoma)

MISSISSIPPI (Mississippi Baptist Hospital) - Cellular schwannoma (1)

FLORIDA (Tallahassee) - Schwannoma, malignant schwannoma

MARYLAND (Bethesda Naval Medical Center) - Cellular schwannoma (7); Malignant peripheral nerve sheath tumor (2)

MASSACHUSETTS (Berkshire Med Ctr) - Malignant peripheral nerve sheath tumor

NEW YORK (Impath Study Group) - Malignant peripheral nerve sheath tumor (4)

NEW JERSEY (Overlook Hospital Summit) - Malignant schwannoma, low grade (2); Schwannoma (3)

CONNECTICUT (Univ of Conn) - Benign peripheral nerve sheath tumor; Malignant peripheral nerve sheath tumor

AUSTRALIA (Sydney Group) - Cellular schwannoma (3); MPNST (4)

**DIAGNOSIS:**

**MALIGNANT SCHWANNOMA, POSTERIOR MEDIASTINUM**
**TY2340/M95603**

**CONSULTATION:**

AFIP (Departments of soft tissue pathology, and pulmonary and mediastinal pathology):
"Malignant schwannoma"

**FOLLOW-UP:**

Six years following surgery, he developed a liver metastasis and died soon after.

REFERENCES:

Moon WK, Im JG, Han MC. Malignant Schwannomas of the Thorax. CT Findings. *J Comput Assist Tomogr* 1993; 17(2):274-276.

Saenz NC, Schnitzer JJ, Eraklis AE, Hendren WH, Grier HE, et al. Posterior Mediastinal Masses. *J Pediatr Surg* 1993; 28(2):172-176.

D'Agostino AN, Soule EH, Miller RH. Primary Malignant Neoplasm of Nerves (Malignant Neurilemmomas) in Patients without Manifestations of Multiple Neurofibromatosis (von Reckinghausen's Disease). *Cancer* 1963; 16:1015-1027.

Akwarai OE, Payne WS, Onofrio BM et al. Dumbell Neurogenic Tumors of the Mediastinium. *Mayo Clin Proc* 1978; 53:353-358.

INLAND (RIVERSIDE/SAN BERNARDINO) - Cellular schwannoma (2); Malignant fibrous histiocytoma (1)

BAKERSFIELD (Central Valley Study Group) - Schwannoma

PLEASANTON/FREMONT - Fibrous histiocytoma (3); Ancient fasciitis (3)

LONG BEACH - Neurofibroma (7)

SANTA BARBARA (St. Francis Med Ctr) - Neurofibroma (3)

SANTA BARBARA (Cottage Hospital) - Myxoid lesion with recurring potential - ? giant cell fibroblastoma

SANTA ROSA - Myofibroblastic tumor (pseudotumor) (2); Myofibroblastic tumor (inflammatory pseudotumor)

BAY AREA - Malignant fibrous histiocytoma (myxoid type) (3)

SACRAMENTO (UC Davis Medical Center) - Benign fibrous tumor - Fibromatosis (extra-abdominal desmoid tumor)

ARIZONA (VA Medical Ctr) - Benign soft tissue lesion, probable neurofibroma

COLORADO (Konopka/Chu Study Group) - Sarcoma, NOS with HPC like areas

KANSAS (Univ of Kansas) - Nodular fasciitis

NEBRASKA (Creighton University) - Benign fibrous histiocytoma

MISSISSIPPI (Mississippi Baptist Hospital) - Solitary fibrous tumor (1)

FLORIDA (Tallahassee) - Schwannoma; Neurofibroma; Low grade neurosarcoma

MARYLAND (Bethesda Naval Medical Center) - Schwannoma (ancient) (9); Cat scratch disease (12)

MASSACHUSETTS (Berkshire Med Ctr) - Schwannoma

NEW YORK (Impath Study Group) - Benign nerve sheath tumor, ?neurofibroma (4)

NEW JERSEY (Overlook Hospital Summit) - Neurofibroma (3); Low grade fibromyxoid sarcoma (1)

CONNECTICUT (Univ of Conn) - Nodular fasciitis; Fibrous histiocytoma

AUSTRALIA (Sydney Group) - Neurofibroma, hyalinized form

DIAGNOSIS:

**SPINDLED NEOPLASM MOST CONSISTENT WITH HEMANGIOPERICYTOMA, LEFT BUTTOCK**

Note: Tumor was S-100 protein negative. CD-34 was not performed

**TY1600/M91501**

REFERENCES:

Nappi O, Ritter JH, Pettinato G, and Wick MR. Hemangiopericytoma. Histopathological Pattern or Clinicopathologic Entity? *Sem in Diag Pathol* 1995; 12(3):221-232.

Enzinger FM, Smith BH. Hemangiopericytoma. A Review of 106 Cases. *Hum Pathol* 1976; 7:61-82.

Yu CC-W, Hall PA, Fletcher CDM, et al. Hemangiopericytomas. The Prognostic Value of Immunohistochemical Staining with a Monoclonal Antibody to Proliferating Cell Nuclear Antigen (PCNA). *Histopathology* 1991; 19:29-33.

McMaster M, Soule E, Ivins J. Hemangiopericytoma. A Clinicopathologic Study of Long-Term Follow-Up of 60 Patients. *Cancer* 1975; 36:2232.

Nielson GP, Dickersin GR, Provenzal JM, Rosenberg AE. Lipomatous Hemangiopericytoma. A Histologic, Ultrastructural and Immunohistochemical Study of a Unique Variant of Hemangiopericytoma. *Am J Surg Pathol* 1995; 19(7):748-756.

Al-Khalifa S, Paulose KO, Shenoy P, Sharma RK. Hemangiopericytoma of the Nasal Septum. *J Laryngol Otol* 1988; 102(12):1161-1163.

**CASE NO. 3, ACCESSION NO. 27874** **SEPTEMBER 1996**

INLAND (RIVERSIDE/SAN BERNARDINO) - Arteriovenous hemangioma (2); Infiltrating angiolipoma of muscle (1)

BAKERSFIELD (Central Valley Study Group) - Hemangioma, intramuscular

PLEASANTON/FREMONT - Intramuscular angiolipoma (5); Intramuscular cavernous hemangioma with fatty infiltration (1)

LONG BEACH - Intramuscular angiolipomatous malformation (7)

SANTA BARBARA (St. Francis Med Ctr) - Intramuscular hemangioma (3)

SANTA BARBARA (Cottage Hospital) - Angiomatosis of soft tissue

SANTA ROSA - Hemangioma (2); Intramuscular hemangioma, large vessel type (1)

BAY AREA - Intramuscular angiolipoma (3)

SACRAMENTO (UC Davis Medical Center) - Angiomatosis vs cavernous intramuscular hemangioma

ARIZONA (VA Medical Ctr) - Intramuscular hemangioma

COLORADO (Konopka/Chu Study Group) - AV hemangioma

KANSAS (Univ of Kansas) - Intramuscular cavernous hemangioma

NEBRASKA (Creighton University) - Fibromatosis (extra-abdominal desmoid)

MISSISSIPPI (Mississippi Baptist Hospital - Angiomatosis (1)

FLORIDA (Tallahassee) - Angiomatosis; AVM

MARYLAND (Bethesda Naval Medical Center) - Intramuscular hemangioma (5); Intramuscular angiomatosis (4)

MASSACHUSETTS (Berkshire Med Ctr) - Intramuscular angiolipoma (2); vs well-differentiated liposarcoma (4)

NEW YORK (Impath Study Group) - Intramuscular angiomatosis (4)

NEW JERSEY (Overlook Hospital Summit) - Intramuscular hemangioma (angiomatosis) (4)

CONNECTICUT (Univ of Conn) - Lymphangiectasis; A-V malformation

AUSTRALIA (Sydney Group) - Benign infiltrating angiolipoma of muscle

**DIAGNOSIS:**

**INTRAMUSCULAR ANGIOMATOSIS ("DIFFUSE HEMANGIOMA"), RIGHT LEG**
**TY9410/M91200**

REFERENCES:

Allenby PA, et al. Diffuse Angiomatosis of the Extremities Presenting as a Sarcoma. *Arch Pathol Lab Med* 1990; 114:987-990.

Vos LD, Bom EP, Vroegindeweij D, Tielbeek AV. Congenital Pelvic Arteriovenous Malformation. A Rare Cause of Sciatica. *Clin Neurol Neurosurg* 1995; 97(3):229-232.

**CASE NO. 4, ACCESSION NO. 27203** **SEPTEMBER 1996**

INLAND (RIVERSIDE/SAN BERNARDINO) - Nodular fasciitis, intramuscular type (1)

BAKERSFIELD (Central Valley Study Group) - Nodular fasciitis

PLEASANTON/FREMONT - Nodular fasciitis (4)

LONG BEACH - Nodular fasciitis (7)

SANTA BARBARA (St. Francis Med Ctr) - Nodular fasciitis (3)

SANTA BARBARA (Cottage Hospital) - Nodular fasciitis

SANTA ROSA - Nodular fasciitis (3)

BAY AREA - Malignant fibrous histiocytoma, low grade (1); Atypical fibrous histiocytoma - ? fasciitis (1); ? myositis (1)

SACRAMENTO (UC Davis Medical Center) - Nodular fasciitis

ARIZONA (VA Medical Ctr) - Kaposi's sarcoma

COLORADO (Konopka/Chu Study Group) - Nodular fasciitis

KANSAS (Univ of Kansas) - Benign fibroblastic tumor (neurofibroma)

NEBRASKA (Creighton University) - Nodular fasciitis

MISSISSIPPI (Mississippi Baptist Hospital) - Fasciitis (1)

FLORIDA (Tallahassee) - Nodular fasciitis

MARYLAND (Bethesda Naval Medical Center) - Nodular fasciitis (9)

MASSACHUSETTS (Berkshire Med Ctr) - Synovial sarcoma vs chondrosarcoma vs osteosarcoma

NEW YORK (Impath Study Group) - Nodular fasciitis (4)

NEW JERSEY (Overlook Hospital Summit) - Nodular fasciitis (4)

CONNECTICUT (Univ of Conn) - Nodular fasciitis

AUSTRALIA (Sydney Group) - Nodular fasciitis

**DIAGNOSIS:**

**NODULAR FASCIITIS, CHEST WALL**
**TY2150/M76130**

REFERENCES:

Bernstein K, Lattes R. Nodular (Pseudosarcomatous) Fasciitis, a Non-Recurrent Lesion: Clinicopathologic Study of 134 Cases. *Cancer* 1982; 49:1668-1678.

Faber LP, Somers J, Templeton AC. Chest Wall Tumors. *Current Prob in Surg* 1995; 32(8):661-756.

King RM, Pairolero PC, Trastek VF, Piehler JM, Payne WS, Bernatz PE. Primary Chest Wall Tumors. Factors Affecting Survival. *Ann Thorac Surg* (United States), 1986; 41(6):597-601.

**CASE NO. 5, ACCESSION NO. 22007** **SEPTEMBER 1996**

INLAND (RIVERSIDE/SAN BERNARDINO) - Synovial sarcoma (2); Clear cell sarcoma (1)
BAKERSFIELD (Central Valley Study Group) - Fibrosarcoma
PLEASANTON/FREMONT - Monophasic synovial sarcoma (6)
LONG BEACH - Synovial sarcoma (7)
SANTA BARBARA (St. Francis Med Ctr) - Monophasic synovial sarcoma (3)
SANTA BARBARA (Cottage Hospital) - Synovial sarcoma, monophasic
SANTA ROSA - Rhabdomyosarcoma (1); Synovial sarcoma (2)
BAY AREA - Synovial sarcoma (2); Epithelioid sarcoma (1).
SACRAMENTO (UC Davis Medical Center) - Monophasic synovial sarcoma
ARIZONA (VA Med Ctr) - Synovial sarcoma
COLORADO (Konopka/Chu Study Group) - Synovial sarcoma
KANSAS (Univ of Kansas) - Synovial sarcoma
NEBRASKA (Creighton University) - Monophasic synovial sarcoma
MISSISSIPPI (Miss Baptist Hospital) - Synovial sarcoma (1)
FLORIDA (Tallahassee) - Synovial sarcoma
MARYLAND (Bethesda Naval Medical Center) - Synovial sarcoma (9)
MASSACHUSETTS (Berkshire Medical Ctr) - Synovial sarcoma
NEW YORK (Impath Study Group) - Synovial sarcoma (4)
NEW JERSEY (Overlook hospital Summit) - Monophasic synovial sarcoma (4)
CONNECTICUT (Univ of Conn) - Synovial sarcoma, monophasic fibrous type; Rhabdomyosarcoma, embryonal; Sarcoma, NOS
AUSTRALIA (Sydney Group) - Synovial sarcoma, predominantly monophasic

**DIAGNOSIS:**

**SYNOVIAL SARCOMA, THIGH**
**TY9100/M90403**

REFERENCES:

Clark J, Rocques PJ, Crew AJ, et al. Identification of Novel Genes, SYT and SSX, Involved in the t (x;18) (p11.2q-112.2) Translocation Found in Human Synovial Sarcoma. *Nat Genet* 1994; 7:502-508.
Cagle LA, Mirra JM, et al. Histologic Features Relating to Prognosis in Synovial Sarcoma. *Cancer* 1987; 59:1810.

**CASE NO. 6, ACCESSION NO. 23022** **SEPTEMBER 1996**

INLAND (RIVERSIDE/SAN BERNARDINO) - Ewing's sarcoma (3)

BAKERSFIELD (Central Valley Study Group) - Ewing, extra skeletal

PLEASANTON/FREMONT - PNET (6)

LONG BEACH - Ewing's (7)

SANTA BARBARA (St. Francis Med Ctr) - Ewing's sarcoma (3)

SANTA BARBARA (Cottage Hospital) - PNET/Ewing's sarcoma (Askin tumor)

SANTA ROSA - Perifesal neuroectodermal tumor (1); Primitive neuroectodermal tumor (2)

BAY AREA - Extraskeletal Ewing's sarcoma (3)

SACRAMENTO (UC Davis Medical Center) - Ewing's sarcoma/peripheral primitive neuroectodermal - AKA Askin tumor.

ARIZONA (VA Med Ctr) - Extraskeletal Ewing's sarcoma

COLORADO (Konopka/Chu Study Group) - Malignant small cell tumor of the thoracopulmonary region in childhood (Askin tumor)

KANSAS (Univ of Kansas) - "Small blue cell tumor" - favor Ewing

NEBRASKA (Creighton Univ) - Ewing's sarcoma, extraskeletal

MISSISSIPPI (Mississippi Baptist Hosp) - Ewing's sarcoma (1)

FLORIDA (Tallahassee) - Ewing's sarcoma

MARYLAND (Bethesda Naval Medical Center) - Primitive neuroectodermal tumor (PNET/Askin's tumor) (5); Extraskeletal Ewing's sarcoma (4)

MASSACHUSETTS (Berkshire Med Ctr) - Ewing's sarcoma

NEW YORK (Impath Study Group) - Extraskeletal Ewing's sarcoma (4)

NEW JERSEY (Overlook Hospital Summit) - Ewing's sarcoma (1); PNET (1); Malignant small cell tumor of thoracopulmonary origin

CONNECTICUT (Univ of Conn) - Primitive neuroectodermal tumor

AUSTRALIA (Sydney Group) - Askin Tumour (PNET)

**DIAGNOSIS:**

**PRIMITIVE NEUROECTODERMAL TUMOR / EXTRASKELETAL EWING'S SARCOMA, CHEST WALL**
**TY2150/M92603**

**CONSULTATION:**

AFIP (Sharon W. Weiss, M.D.) - "Sarcoma, not further classified, chest wall."

REFERENCES:


May WA, Gishizlary ML, Lessnich SL, et al. Ewing Sarcoma 11; 22 Translocation Produces a Chimeric Transscription Factor that requires the DNA-Binding Domain Encoded by FLI1 for Transformation. *Proc Natl Acad Sci* USA 1993; 90:5752-5756.

Zucman J, Melot T Demaze C, et al. Combinational Generation of Variable Fusion Proteins in the Ewing Family of Tumors. *EMBOJ* 1993; 12:4481-4487.

Ladanyi M. The Emerging Molecular Genetics of Sarcoma Translocations. *Diag Molecular Pathol* 1995; 4(3):162-173.

Schmidt D, Herrman C, Jurgens H and Harms D. Malignant Peripheral Neuroectodermal Tumor and its Necessary Distinction from Ewing's Sarcoma. A Report from the Kid Pediatric Tumor Registry. *Cancer* 1991; 68:2251-2259.

INLAND (RIVERSIDE/SAN BERNARDINO) - Poorly differentiated rhabdomyosarcoma (2); Alveolar soft part sarcoma (1)

BAKERSFIELD (Central Valley Study Group) - Rhabdomyosarcoma

PLEASANTON/FREMONT - Rhabdomyosarcoma (6)

LONG BEACH - Rhabdomyosarcoma (7)

SANTA BARBARA (St. Francis Med Ctr) - Rhabdomyosarcoma (3)

SANTA BARBARA (Cottage Hospital) - Pleomorphic rhabdomyosarcoma

SANTA ROSA - Rhabdomyosarcoma (2); Rhabdomyosarcoma, pleomorphic (1)

BAY AREA - Malignant rhabdoid tumor (2); Rhabdomyosarcoma (1)

SACRAMENTO (UC Davis Medical Center) - Pleomorphic rhabdomyosarcoma

ARIZONA (VA Med Ctr) - Rhabdomyosarcoma

COLORADO (Konopka/Chu Study Group) - Malignant (rhabdomyosarcoma vs carcinoma)

KANSAS (Univ of Kansas) - Rhabdomyosarcoma

NEBRASKA (Creighton University) - Rhabdomyosarcoma

MISSISSIPPI (Mississippi Baptist Hospital) - Pleomorphic rhabdomyosarcoma

FLORIDA (Tallahassee) - Rhabdomyosarcoma

MARYLAND (Bethesda Naval Medical Center) - Extrarenal malignant rhabdoid tumor (7); Malignant granular cell tumor (2)

MASSACHUSETTS (Berkshire Med Ctr) - Rhabdomyosarcoma (4); vs Mets renal cell carcinoma (2)

NEW YORK (Impath Study Group) - Extraskeletal malignant rhabdoid tumor (4)

NEW JERSEY (Overlook Hospital Summit) - Malignant rhabdoid tumor (1); Rhabdomyosarcoma (3)

CONNECTICUT (Univ of Conn) - Myosarcoma ; Malignant granular cell tumor ; Embryonal rhabdomyosarcoma

AUSTRALIA (Sydney Group) - Epithelioid mesothelioma; Inflammatory MFH

**DIAGNOSIS:**

**MALIGNANT NEOPLASM WITH RHADBOID PHENOTYPE, CHEST WALL**
Note: further study produced tissues included as "Case 11" of October 1996 study set.
**TY2150/M80003**

**FOLLOW-UP:**

Twelve hours following the resection of the tumor the patient went into sudden uncontrolled bleeding episodes at the thoracotomy site. The patient expired and no autopsy was performed.

REFERENCES:

Chase DR. Rhabdoid vs Epithelioid Sarcoma (Letter to Editor). *Am J Surg Pathol* 1990; 14:792.

Kodet R, Newton WA, et al. Rhabdoid Tumors of Soft Tissues. A Clinicopathological Study of 26 Cases Enrolled in the Intergroup Rhabdomyosarcoma Study. *Hum Pathol* 1991; 22:674.

Tsuneyoshi M, et al. The Existance of Rhabdoid Cells in Specified Soft Tissue Sarcomas. Histopathological, Ultrastructural, and Immunohistochemical Evidence. *Virchows Arch (Pathol Anat)* 1987; 411:509.

INLAND (RIVERSIDE/SAN BERNARDINO) - Poorly differentiated liposarcoma (3)

BAKERSFIELD (Central Valley Study Group) - Liposarcoma

PLEASANTON/FREMONT - Pleomorphic liposarcoma (6)

LONG BEACH - Liposarcoma, dedifferentiated (high-grade) (7)

SANTA BARBARA (St. Francis Med Ctr) - Myxoid liposarcoma (3)

SANTA BARBARA (Cottage Hospital) - Liposarcoma, round cell type

SANTA ROSA - Myxoid liposarcoma (2); Myxoid liposarcoma with round cell area (1)

BAY AREA - Myxoid liposarcoma (3)

SACRAMENTO (UC Davis Medical Center) - Liposarcoma, round cell type

ARIZONA (VA Med Ctr) - Myxoid liposarcoma

COLORADO (Konopka/Chu Study Group) - Myxoid liposarcoma

KANSAS (Univ of Kansas) - Liposarcoma vs Yolk sac carcinoma

NEBRASKA (Creighton University) - Liposarcoma, myxoid and round cell

MISSISSIPPI (Mississippi Baptist Hospital) - Pleomorphic liposarcoma (1)

FLORIDA (Tallahassee) - Liposarcoma, Grade II/III, myxoid and round cell type

MARYLAND (Bethesda Naval Medical Center) - Pleomorphic liposarcoma (9)

MASSACHUSETTS (Berkshire Med Ctr) - Myxoid liposarcoma and pleomorphic liposarcoma

NEW YORK (Impath Study Group) - Myxoid liposarcoma with round cell (pleomorphic) dedifferentiation. (4)

NEW JERSEY (Overlook Hospital Summit) - Liposarcoma (1); Round cell liposarcoma (1); Dedifferentiated liposarcoma (2)

CONNECTICUT (Univ of Conn) - Liposarcoma

AUSTRALIA (Sydney Group) - Round cell liposarcoma and myxoid liposarcoma

**DIAGNOSIS:**

**MYXOID AND ROUND CELL LIPOSARCOMA ("HIGH GRADE MYXOID LIPOSARCOMA"), LEFT ADNEXAL**
**T01300/M88523**

**FOLLOW-UP:**

Post-chemotherapy CT scan of the abdomen showed a 3.0 cm recurrent tumor in the region of the left adnexa.

REFERENCES:

Scott E, Kilpatrick, doyon J, Peter FM, et al. The Clinicopathologic Spectrum of Myxoid and Round Cell Liposarcoma. A Study of 55 Cases. *Cancer* 1996; 77:1450-1458.

Rabbitts TH, Forsler A, Larson R, Nathan P. Fusion of the Dominant Negative Transcription Regulator CHOP with a Novel Gene FUS by Translocation t (12; 16) in Malignant Liposarcoma. *Nat Genet* 1993; 4:175-180.

Allen PW. Myxoid Tumors of Soft Tissues. *Pathol Ann* 1980; 15(pt1):156-160.

Frable W. Pathologic Classification of Soft Tissue Sarcomas. *Sem Surg Oncol* 1994; 10(5):332-339.

McCormick D, Mentzel T, Beham A and Fletcher C. Dedifferentiated Liposarcoma. Clinicopathologic Analysis of 32 Cases Suggesting a Better Prognostic Subgroup Among Pleomorphic Sarcomas. *Am J Surg Pathol* 1994; 18(12):1213-1223.

**CASE NO. 9, ACCESSION NO. 27908** **SEPTEMBER 1996**

INLAND (RIVERSIDE/SAN BERNARDINO) - Hemangiopericytoma (3)
BAKERSFIELD (Central Valley Study Group) - Hemangiopericytoma
PLEASANTON/FREMONT - Hemangiopericytoma (6)
LONG BEACH - Solitary fibrous tumor (7)
SANTA BARBARA (St. Francis Med Ctr) - Angiofibroma (3)
SANTA BARBARA (Cottage Hospital) - Hemangiopericytoma
SANTA ROSA - Hemangiopericytoma (3)
BAY AREA - Angiofibroma (3)
SACRAMENTO (UC Davis Medical Center) - Spindle cell hemangioendothelioma
ARIZONA (VA Med Ctr) - Hemangiopericytoma
COLORADO (Konopka/Chu Study Group) - Angiofibroma
KANSAS (Univ of Kansas) - Spindle cell hemangioendothelioma (benign fibrous histiocytoma)
NEBRASKA (Creighton University) - Hemangiopericytoma
MISSISSIPPI (Mississippi Baptist Hospital) - Hemangioperictyoma (1)
FLORIDA (Tallahassee) - Angioleiomyoma
MARYLAND (Bethesda Naval Medical Center) - Hemangiopericytoma (9)
MASSACHUSETTS (Berkshire Med Ctr) - Solitary fibrous tumor
NEW YORK (Impath Study Group) - Solitary fibrous tumor of nasal cavity (4)
NEW JERSEY (Overlook Hospital Summit) - Hemangiopericytoma (4)
CONNECTICUT (Univ of Conn) - Hemangioperictyoma, benign
AUSTRALIA (Sydney Group) - Hemangiopericytoma

**DIAGNOSIS:**

**SOLITARY FIBROUS TUMOR, RIGHT PYRIFORM SINUS**
**T60320/M88100**

**FOLLOW-UP:**

The patient was seen three times since the surgery and shows no evidence of recurrent tumor.

REFERENCES:


Wilkin CB, Rosai J. Solitary Fibrous Tumors of the Upper Respiratory Tract. A Report of Six Cases. *Am J Surg Pathol* 1991; 15:842-848.

Zukerberg LR, Rosenberg AE, Randolf G, et al. Solitary Fibrous Tumor of the Nasal Cavity and Paranasal Sinuses. *Am J Surg Pathol* 1991; 15:126-130.

Flint A, Weiss SW, CD-34 and Keratin Expression Distinguishes solitary Fibrous Tumor (Fibrous Mesothelioma) of Pleura From Desmoplastic Mesothelioma. *Hum Pathol* 1995; 26:428-431.

Briselli M, Mark EJ, Dickersin GR. Solitary Fibrous Tumors of the Pleura. Eight New Cases and Review of 360 Cases in the Literature. *Cancer* 1981; 47:2678-2689.

INLAND (RIVERSIDE/SAN BERNARDINO) - Malignant peripheral nerve sheath tumor (3)

BAKERSFIELD (Central Valley Study Group) - Nerve sheath tumor

PLEASANTON/FREMONT - Malignant peripheral nerve sheath tumor (MPNST) with epithelioid cell differentiation (6)

LONG BEACH - Malignant peripheral neuroectodermal tumor (arising in patients with neurofibromatosis) (7)

SANTA BARBARA (St. Francis Med Ctr) - Ependymoma (3)

SANTA BARBARA (Cottage Hospital) - Peripheral malignant nerve sheath tumor, high grade, probable Triton tumor

SANTA ROSA - Sarcoma, probably neurofibrosarcoma (1); Sarcoma, probably epithelioid (1); Epithelioid hemangioendothelioma vs epithelioid sarcoma (1)

BAY AREA - Malignant peripheral nerve sheath tumor (3)

SACRAMENTO (UC Davis Medical Center) - Malignant peripheral nerve sheath tumor arising in a plexiform neurofibroma or ganglioneuroma.

ARIZONA (VA Med Ctr) - Malignant peripheral nerve sheath tumor

COLORADO (Konopka/Chu Study Group) - Malignant schwannoma

KANSAS (Univ of Kansas) - Malignant peripheral nerve sheath tumor

NEBRASKA (Creighton University) - Malignant peripheral nerve sheath tumor

MISSISSIPPI (Mississippi Baptist Hospital) - Epithelioid MPNST (1)

FLORIDA (Tallahassee) - Malignant peripheral nerve sheath tumor.

MARYLAND (Bethesda Naval Medical Center) - Epithelioid malignant peripheral nerve sheath tumor (9)

MASSACHUSETTS (Berkshire Med Ctr) - Malignant peripheral nerve sheath tumor

NEW YORK (Impath Study Group) - Primitive neuroectodermal tumor associated with neurofibromatosis adjacent nerves (4)

NEW JERSEY (Overlook Hospital Summit) - Malignant mesenchymoma (nerve sheath origin) (1); Sarcoma (NOS) (2); Malignant epithelioid peripheral nerve sheath tumor (1)

CONNECTICUT (Univ of Conn) - Peripheral nerve sheath tumor, malignant; Sarcoma, NOS

AUSTRALIA (Sydney Group) - Malignant epithelioid schwannoma

**DIAGNOSIS:**

**MALIGNANT PERIPHERAL NERVE SHEATH TUMOR ("MALIGNANT SCHWANNOMA"), SCIATIC NERVE**

*Note: Rare slides showed the presence of rhabdomyoblasts ("Malignant Triton Tumor").

**TX9440/M95603**

REFERENCES:

Hruban RH, Shiu MH, Senie RT, et al. Malignant Peripheral Nerve Sheath Tumors of the Buttock and Lower Extremity. *Cancer* 1990; 66:1253.

Matsunou H, Shimoda T, Kakimoto S, et al. Histopathologic and Immunohistochemical Study of Malignant Tumors of Peripheral Nerve Sheath (Malignant Schwannoma). *Cancer* 1985; 56:2269.

Menon AG, Anderson KM, Riccardi VM, et al. Chromosome 17p Deletions and p53 Gene Mutations Associated with the Formation of Malignant Neurofibrosarcomas in von Recklinghausen's Neurofibromatosis. *Proc Natl Acad Sci* USA 1990; 87:5435.

Payne RA. Metaplasia in a Nerve Sheath Sarcoma in von Recklinghausen's Disease. *Br J Surg* 1960; 47:688.

Coindre JM, Terrier P, Bui B, Bonichon F, et al. Prognostic Factors in Adult Patients with Locally Controlled Soft Tissue Sarcoma. A Study of 546 Patients from the French Federation of Cancer Centers - Sarcoma Group. *J Clin Oncol* 1996; 14:869-877.