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*"Pediatric Pathology"*

**Case 1:**

An 8 m/o girl presented with a fibrous mass involving the posterior left upper arm. It was located in the subcutis, external to the triceps fascia.

**Dx:**

Fibrous Hamartoma, arm

Santa Barbara (Miramonte Laboratory) - Infiltrating lipoma  
Arkansas (The Laboratory of Pathology, P.A.) - Fibrous hamartoma of infancy  
Georgia, Atlanta - Fibrous hamartoma of infancy  
Kansas (Coffeyville Regional Medical Center) - Fibrous hamartoma of infancy  
Kansas (Peterson Laboratory Services) - Fibrous hamartoma of infancy (2)  
Michigan (Oakwood Hospital) - Fibrous hamartoma of infancy  
New Mexico, Albuquerque - Fibrous hamartoma of infancy  
Canada (Pasqua Hospital) - Infantile fibromatosis  
The Netherlands, Amstelveen - Fibrolipoma

**Refs:**

Cytologic diagnosis of fibrous hamartoma of infancy: a case report.  
Diagn Cytopathol 2003 May;28(5): p272-3  
Mohanty SK; Dey P

Fibrous hamartoma of infancy at the wrist and the use of MRI in preoperative planning.  
Pediatr Radiol 2001 Jun;31(6): p450-2  
Ashwood N; Witt JD; Hall-Craggs MA

Fibrous hamartoma of corpus cavernosum: a rare cause of congenital penile curvature associated with erectile dysfunction.  
J Urol 2004 Aug;172(2): p642-3  
Bertaccini A; Marchiori D, et al.

Fibrous hamartoma of infancy: an Italian multi-institutional experience.  
J Am Acad Dermatol 2006 May;54(5): p800-3  
Carretto E; Dall'Igna P, et al.

Fibrous hamartoma of infancy: a case report with associated cytogenetic findings.  
Arch Pathol Lab Med 2005 Apr;129(4): p520-2  
Lakshminarayanan R; Konia T; Welborn J

Cytology of fibrous hamartoma of infancy: a helpful approach for an uncommon soft tissue lesion.

Acta Cytol 2008 Mar-Apr;52(2): p123  
Sforza-Huffman C

Fibrous hamartoma of the scrotum in an infant.  
Minerva Pediatr 2005 Dec;57(6): p447-8  
Sakellaris G; Kafousi M; Charissis G

## **Case 2**

An 8 day old male infant had a 4 cm mass in the right cheek. The contributor called it a hemangiopericytoma.

### **Dx:**

Infantile myofibroma

Santa Barbara (Miramonte Laboratory) - Hemangiopericytoma  
Arkansas (The Laboratory of Pathology, P.A.) - Hemangioma  
Georgia, Atlanta - Angiomatoid fibrous histiocytoma  
Kansas (Coffeyville Regional Medical Center) - Solid variant of alveolar rhabdomyosarcoma  
Kansas (Peterson Laboratory Services) - Epithelioid hemangioendothelioma (1); Myofibroma (1)  
Michigan (Oakwood Hospital) - Myofibroma  
New Mexico, Albuquerque - Myofibroma  
Canada (Pasqua Hospital) - Infantile myofibromatosis  
The Netherlands, Amstelveen - Solitary fibrous tumor

### **Refs:**

Solitary intraosseous orbital myofibroma in four cases.  
Ophthal Plast Reconstr Surg 2006 Jul-Aug;22(4): p292-5  
Rodrigues EB; Shields CL, et al.

Myopericytoma: a unifying term for a spectrum of tumours that show overlapping features with myofibroma. A review of 14 cases.  
J Clin Pathol 2006 Jan;59(1): p67-73  
Dray MS; McCarthy SW, et al.

Intranasal myopericytoma. A tumour with perivascular myoid differentiation: the changing nomenclature for haemangiopericytoma.  
J Laryngol Otol 2007 Aug;121(8): p786-9  
Wilson T; Hellquist HB, et al.

Myopericytoma of skin and soft tissues: clinicopathologic and immunohistochemical study of 54 cases.  
Am J Surg Pathol 2006 Jan;30(1): p104-13  
Mentzel T; Dei Tos AP, et al.

## **Case 3**

A 4 m/o boy was born with a left kidney mass. Grossly it was 132 grams and 10.0 x 6.5 x 4.0. The cut surface bulged as a 6.0 x 5.0 x 2.5 cm yellow, trabeculated, rubbery tumor that obliterated

approximately half of the kidney parenchyma.

**Dx:**

Congenital mesoblastic nephroma, mixed pattern, predominantly classic type

Santa Barbara (Miramonte Laboratory) - Leiomyosarcoma

Arkansas (The Laboratory of Pathology, P.A.) - Mesoblastic nephroma

Georgia, Atlanta - Mesoblastic nephroma, classic type

Kansas (Coffeyville Regional Medical Center) - Congenital mesoblastic nephroma (leiomyomatous hamartoma)

Kansas (Peterson Laboratory Services) - Congenital mesoblastic nephroma (1); Mesoblastic nephroma (1)

Michigan (Oakwood Hospital) - Congenital mesoblastic nephroma

New Mexico, Albuquerque - Mesoblastic nephroma

Canada (Pasqua Hospital) - Congenital mesoblastic nephroma

The Netherlands, Amstelveen - Fibrosarcoma features

**Refs:**

Cellular mesoblastic nephroma in an infant: report of the cytologic diagnosis of a rare paediatric renal tumor.

Diagn Cytopathol 2009 May;37(5): p377-80

Gupta R; Mathur SR, et al.

Adult mesoblastic nephroma: a case with fatal recurrence.

Urol J 2008 Spring;5(2): p136-7

Moslemi MK

Mesoblastic nephroma--a report from the Gesellschaft fur Padiatrische Onkologie und Hamatologie (GPOH).

Cancer 2006 May 15;106(10): p2275-83

Furtwaengler R; Reinhard H, et al.

Prenatal diagnosis of congenital mesoblastic nephroma associated with renal hypertension in a premature child.

J Urol 2005 Mar;173(3): p983

Canning DA

[Congenital mesoblastic nephroma: a clinicopathologic study of five cases]

Med Wieku Rozwoj 2006 Jul-Sep;10(3 Pt 1): p677-86

Wozniak ZM; Sawicz-Birkowska K, et al.

**Case 4**

A 4 y/o boy presented with a left renal mass. The radical nephrectomy showed the kidney to contain a 10.0 x 8.0 x 8.0 cm tumor which was white-tan, but with intermixed firm/solid areas and areas of hemorrhage and necrosis.

**Dx:**

Wilms tumor, kidney

Santa Barbara (Miramonte Laboratory) - Wilms tumor

Arkansas (The Laboratory of Pathology, P.A.) - Wilms tumor

Georgia, Atlanta - Wilms tumor (nephroblastoma)

Kansas (Coffeyville Regional Medical Center) - Anaplastic nephroblastoma (Wilms tumor)  
Kansas (Peterson Laboratory Services) - Wilms tumor (1); Nephroblastoma (1)  
Michigan (Oakwood Hospital) - Wilms tumor  
New Mexico, Albuquerque - Nephroblastoma  
Canada (Pasqua Hospital) - Wilms tumor  
The Netherlands, Amstelveen - Wilms tumor

### **Refs:**

Amplification and expression of EGFR and ERBB2 in Wilms tumor.  
Cancer Genet Cytogenet 2009 Oct 15;194(2): p88-95  
Vasei M; Modjtahedi H, et al.

Secondary neoplasms after Wilms' tumor in Germany.  
Strahlenther Onkol 2009 Aug;185 Suppl 2:11-2  
Nourkami N; Furtwangler R; Alkassar M; Graf N

C-kit protein expression in Wilms' tumour: an immunohistochemical study.  
Eur J Surg Oncol 2009 Jun;35(6): p629-35  
Giordano G; Campanini N, et al.

Stroma-predominant Wilms tumor with teratoid features: report of a rare case and review of the literature.  
Pediatr Surg Int 2009 Mar;25(3): p293-5  
Gupta R; Sharma A; Arora R; Dinda AK

Cystic nephroma, cystic partially differentiated nephroblastoma and cystic Wilms' tumor in children: a spectrum with therapeutic dilemmas.  
Urol Int 2009;82(1): p65-70  
van den Hoek J; de Krijger R, et al.

### **Case 5**

An 8-year old female was found to have right abdominal mass. Imaging revealed a mass in the right kidney measuring 12 cm in greatest diameter. A right nephrectomy was performed. The nephrectomy specimen weighed 508 g and measured 12 cm in greatest diameter. It contained a 10 cm mass with a multinodular, variegated, yellow, soft, and focally hemorrhagic cut surface. The tumor bulged into the renal hilum and extended to within 0.1 cm of the inked soft tissue margin.

### **Dx:**

Wilms tumor with diffuse anaplasia

Santa Barbara (Miramonte Laboratory) - Neuroblastoma  
Arkansas (The Laboratory of Pathology, P.A.) - Neuroblastoma  
Georgia, Atlanta - PNET  
Kansas (Coffeyville Regional Medical Center) - PNET (primitive neuroectodermal tumor)  
Kansas (Peterson Laboratory Services) - Peripheral neuroectodermal tumor (1); Clear cell sarcoma (1)  
Michigan (Oakwood Hospital) - PNET  
New Mexico, Albuquerque - Neuroblastoma  
Canada (Pasqua Hospital) - Wilms vs. clear cell sarcoma  
The Netherlands, Amstelveen - Malignant tumor small round blue tumor

**Refs:**

Loss of 11q and 16q in Wilms tumors is associated with anaplasia, tumor recurrence, and poor prognosis.

Genes Chromosomes Cancer 2007 Feb;46(2): p163-70

Wittmann S; Zirn B, et al.

Bilateral Wilms' tumor with anaplasia: lessons from the National Wilms' Tumor Study.

J Pediatr Surg 2006 Oct;41(10): p1641-4

Hamilton TE; Green DM; Perlman EJ; Argani P, et al.

Molecular cytogenetic anomalies and phenotype alterations in a newly established cell line from Wilms tumor with diffuse anaplasia.

Cancer Genet Cytogenet 2008 Jul;184(1): p22-30

Faussillon M; Murakami I, et al.

Treatment of anaplastic histology Wilms' tumor: results from the fifth National Wilms' Tumor Study.

J Clin Oncol 2006 May 20;24(15): p2352-8

Dome JS; Cotton CA; Perlman EJ; Breslow NE; Kalapurakal JA; Ritchey ML; Grundy PE; Malogolowkin M; Beckwith JB, et al.

Anaplastic nephroblastomas express transketolase-like enzyme 1.

J Clin Pathol 2009 May;62(5): p460-3

Wu HT; Allie N; Myer L; Govender D

**Case 6**

A radical nephrectomy was performed on this 4 y/o child who had hematuria and a right renal mass. The kidney tumor was 12.0 x 8.0 with numerous tan nodules bulging from a white, fibrous stroma. The nodules ranged from 0.7 to 4.0 cm.

**Dx:**

Clear Cell Sarcoma of Kidney

Santa Barbara (Miramonte Laboratory) - Malignant rhabdoid tumor

Arkansas (The Laboratory of Pathology, P.A.) - Clear cell sarcoma of kidney

Georgia, Atlanta - Clear cell sarcoma of kidney

Kansas (Coffeyville Regional Medical Center) -Metanephric stromal tumor

Kansas (Peterson Laboratory Services) - Clear cell sarcoma (1); Sarcoma with rhabdoid features (1)

Michigan (Oakwood Hospital) - Clear cell sarcoma of kidney

New Mexico, Albuquerque - Clear cell sarcoma

Canada (Pasqua Hospital) - Clear cell sarcoma

The Netherlands, Amstelveen - Clear cell sarcoma

**Refs:**

Clear cell sarcoma of the kidney--a study of seven cases over a period of three years.

Indian J Pathol Microbiol 2007 Apr;50(2): p270-3

Viswanathan S; Dave BK; Desai SB

Clear cell sarcoma: a dilemma on pathological staging and clinical management.

Pediatr Hematol Oncol 2005 Apr-May;22(3): p257-61

Ng A; Jenkinson H; Morland B; Grundy R

Fine-needle aspiration cytology of clear-cell sarcoma of the kidney: study of eight cases.

Diagn Cytopathol 2005 Aug;33(2): p83-9

Iyer VK; Agarwala S; Verma K

Recurring translocation (10; 17) and deletion (14q) in clear cell sarcoma of the kidney.

Arch Pathol Lab Med 2007 Mar;131(3): p446-51

Brownlee NA; Perkins LA, et al.

Clonal divergence and genetic heterogeneity in clear cell renal cell carcinomas with sarcomatoid transformation.

Cancer 2005 Sep 15;104(6): p1195-203

Jones TD; Eble JN, et al.

### **Case 7**

A 4 y/o boy presented with a tumor on the floor of the mouth. It was curretted, resulting in an 8 gram, 6 x 2 x 1.6 cm aggregate of multiple white-tan soft tissue fragments admixed with fragments of teeth. Negative for: desmin, MSA, SMA, S100, EMA, Cam5.2.

Elevated Ki-67 - 10% nuclei positive.

### **Dx:**

Myofibroblastic Sarcoma of Mouth

Case seen by Dr. Cheryl Coffin: "Myofibroblastic Sarcoma"

Santa Barbara (Miramonte Laboratory) - Poorly differentiated malignant neoplasm

Arkansas (The Laboratory of Pathology, P.A.) - Nodular fasciitis

Georgia, Atlanta - Nodular fasciitis

Kansas (Coffeyville Regional Medical Center) - Myxoid low grade sarcoma

Kansas (Peterson Laboratory Services) - Nodular fasciitis with epulis (1); Nodular fasciitis (1)

Michigan (Oakwood Hospital) - Inflammatory myofibroblastic tumor

New Mexico, Albuquerque - Infantile fibrosarcoma

Canada (Pasqua Hospital) - Nodular fasciitis

The Netherlands, Amstelveen - Fibromatosis

### **Refs:**

Myofibroblastic sarcoma vs nodular fasciitis: a comparative study of chromosomal imbalances.

Am J Clin Pathol 2009 May;131(5): p701-9

Meng GZ; Zhang HY, et al.

Low-grade myofibroblastic sarcoma of the parapharyngeal space.

Int J Oral Maxillofac Surg 2006 Oct;35(10): p965-8

Takahama A; Nascimento AG, et al.

Inflammatory myofibroblastic tumor and low-grade myofibroblastic sarcoma: a comparative study of clinicopathologic features and further observations on the immunohistochemical profile of myofibroblasts.

Hum Pathol 2008 Jun;39(6): p846-56

Qiu X; Montgomery E; Sun B

Low-grade abdominopelvic sarcoma with myofibroblastic features (low-grade myofibroblastic sarcoma): clinicopathological, immunohistochemical, molecular genetic and ultrastructural study of two cases with literature review.

J Clin Pathol 2008 Mar;61(3): p301-6

Agaimy A; Wunsch PH, et al.

Epithelioid sarcoma.

Arch Pathol Lab Med 2009 May;133(5): p814-9

AU - Armah HB; Parwani AV

### **Case 8**

A 5-year old female was found to have a left retroperitoneal mass. The mass measured 14 cm in greatest diameter. The resected mass weighed 554 g and measured 14 cm in greatest diameter. The dark red mass had a vaguely nodular tan and red cut surface.

### **Dx:**

Neuroblastoma, differentiating subtype with low mitotic-karyorrhectic index

Case seen by Dr. Cheryl Coffin: "Neuroblastoma, differentiating subtype with low mitotic-karyorrhectic index"

Santa Barbara (Miramonte Laboratory) - Rhabdomyosarcoma

Arkansas (The Laboratory of Pathology, P.A.) - PNET

Georgia, Atlanta - Ganglioneuroblastoma

Kansas (Coffeyville Regional Medical Center) - Nephroblastoma (Wilms tumor)

Kansas (Peterson Laboratory Services) - Neuroblastoma (1); Ganglioneuroblastoma (1)

Michigan (Oakwood Hospital) - Neuroblastoma, undifferentiated

New Mexico, Albuquerque - Yolk sac tumor

Canada (Pasqua Hospital) - Rhabdomyosarcoma

The Netherlands, Amstelveen - Neuroblastoma

### **Refs:**

High Myc pathway activity and low stage of neuronal differentiation associate with poor outcome in neuroblastoma.

Proc Natl Acad Sci U S A 2008 Sep 16;105(37): p14094-9

Fredlund E; Ringner M, et al.

The calcium-sensing receptor and parathyroid hormone-related protein are expressed in differentiated, favorable neuroblastic tumors.

Cancer 2009 Jun 15;115(12): p2792-803

de Torres C; Beleta H, et al.

Neuroblastoma is a prototype of those malignancies which can progress very rapidly but can also be differentiated into a benign variant. In this issue. Pathol Oncol Res 2007;13(4): p267  
Kopper

Computer-assisted grading of neuroblastic differentiation. Arch Pathol Lab Med 2008 Jun;132(6): p903-4; author reply 904  
Kong J; Sertel O, et al.

PPARbeta agonists trigger neuronal differentiation in the human neuroblastoma cell line SH-SY5Y. J Cell Physiol 2007 Jun;211(3): p837-47  
Di Loreto S; D'Angelo B, et al.

SIRT1 regulates tyrosine hydroxylase expression and differentiation of neuroblastoma cells via FOXO3a. FEBS Lett 2009 Apr 2;583(7): p1183-8  
Kim MJ; Ahn K, et al.

### **Case 9**

A 13-year old boy with neurofibromatosis type 1 developed pain in the temporomandibular joint. A tumor mass was identified occupying the pterygomaxillary fossa. The mass grew progressively and was biopsied.

### **Dx:**

Embryonal Rhabdomyosarcoma

Case seen by Dr. Cheryl Coffin: "Embryonal Rhabdomyosarcoma"

Santa Barbara (Miramonte Laboratory) - Rhabdomyosarcoma  
Arkansas (The Laboratory of Pathology, P.A.) - Malignant peripheral nerve sheath tumor  
Georgia, Atlanta - Triton tumor  
Kansas (Coffeyville Regional Medical Center) - MPNST (malignant peripheral nerve sheath tumor)  
Kansas (Peterson Laboratory Services) - Malignant peripheral nerve sheath tumor (1); Synovial sarcoma (1)  
Michigan (Oakwood Hospital) - Malignant peripheral nerve sheath tumor  
New Mexico, Albuquerque - Embryonal rhabdomyosarcoma  
Canada (Pasqua Hospital) - Malignant peripheral nerve sheath tumor  
The Netherlands, Amstelveen - Cystic lesion surrounded by mesenchymal cells

### **Refs:**

Trisomy 8 as a sole aberration in embryonal rhabdomyosarcoma (sarcoma botryoides) of the vagina. Cancer Genet Cytogenet 2009 Dec;195(2): p172-4  
Manor E; Bodner L, et al.

Intranuclear cytoplasmic inclusions--an extremely unusual finding in embryonal rhabdomyosarcoma: Report of a case diagnosed by fine-needle aspiration biopsy. Diagn Cytopathol 2009 Oct;37(10): p740-3  
Kumar S; Siddaraju N, et al.



Translocation (2; 8)(q35; q13): a recurrent abnormality in congenital embryonal rhabdomyosarcoma.

Cancer Genet Cytogenet 2009 May;191(1): p43-5

Meloni-Ehrig A; Smith B, et al.

Embryonal rhabdomyosarcoma with a novel t(2; 6)(p23; p21.1).

Cancer Genet Cytogenet 2008 Nov;187(1): p39-42

Pressey JG; Mroczek-Musulman E, et al.

Rhabdomyosarcoma subtyping by immunohistochemical assessment of myogenin: tissue array study and review of the literature.

Pathol Oncol Res 2008 Sep;14(3): p233-8

Morgenstern DA; Rees H, et al.

### **Case 10**

A 9-year old female had a soft tissue mass in the left neck. Multiple samples were obtained from the neck mass and consisted of soft red and focally hemorrhagic and granular tissue up to 6.5 cm in greatest diameter.

### **Dx:**

Malignant rhabdoid tumor, extra-renal

Case seen by Dr. Cheryl Coffin: "Malignant rhabdoid tumor, extra-renal"

Santa Barbara (Miramonte Laboratory) - Melanoma

Arkansas (The Laboratory of Pathology, P.A.) - Hemangioendothelioma

Georgia, Atlanta - Rhabdomyoblastoma

Kansas (Coffeyville Regional Medical Center) - Rhabdomyosarcoma

Kansas (Peterson Laboratory Services) - Rhabdomyosarcoma (2)

Michigan (Oakwood Hospital) - Embryonal rhabdomyosarcoma

New Mexico, Albuquerque - Squamous cell carcinoma

Canada (Pasqua Hospital) - Rhabdomyosarcoma

The Netherlands, Amstelveen - Malignant lymphoma

### **Refs:**

Congenital malignant rhabdoid tumor.

Diagn Cytopathol 2005 Feb;32(2): p135-6

Dehner LP

Extrarenal malignant rhabdoid tumour of the heel--a case report.

Anticancer Res 2005 Nov-Dec;25(6C): p4573-6

Mazzocchi M; Chiummariello S, et al.

Malignant mixed epithelial and stromal tumor of the kidney with rhabdoid features: report of a case including immunohistochemical, molecular genetic studies and comparison to morphologically similar renal tumors.

Hum Pathol 2007 Sep;38(9): p1432-7

Sukov WR; Cheville JC, et al.

Rhabdoid tumour: a malignancy of early childhood with variable primary site, histology and clinical behaviour.  
Pathology 2008 Dec;40(7): p664-70  
Wu X; Dagar V; Algar E, et al.

Proximal-type epithelioid sarcoma vs. malignant rhabdoid tumor of the vulva: a case report, review of the literature, and an argument for consolidation.  
Gynecol Oncol 2007 Oct;107(1): p130-5  
Argenta PA; Thomas S; Chura JC

Malignant teratoid/rhabdoid tumour: long-term survival.  
Can J Neurol Sci 2007 May;34(2): p245-7  
Fisher BJ; Dennis KE; Ang LC