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*"CNS & Neuroendocrine"*

**Case 1:**

A 49 y/o woman presented with this 20 cm mass in the buttock. There was no history of previous tumors.

Santa Barbara (Miramonte Laboratory) - Neurofibroma  
Santa Rosa (Santa Rosa Memorial Hospital) - Neurofibroma, Diffuse  
Woodland Hills (Kaiser Permanente) - Neurofibroma  
Arkansas (The Laboratory of Pathology, P.A.) - Neurofibroma  
Georgia, Atlanta - Neurofibroma  
Illinois, Oak Brook - Neurofibroma  
Kansas (Peterson Laboratory Services) - Neurofibroma  
Louisiana, Shreveport (LSU Shreveport) - Neurofibroma  
Maryland, Baltimore - Neurofibroma  
New Mexico, Albuquerque - Soft tissue neurofibroma  
Ohio, Columbus - Soft tissue, buttock, neurofibroma  
Texas, Victoria - NF, buttock  
Hong Kong (Hong Kong Baptist Hospital) - Neurofibroma  
India, Rochi - Neurofibroma of buttock  
The Netherlands, Amstelveen - Neurofibroma

**Dx:**

Neurofibroma, buttock

**Refs:**

Cytogenetics of neurofibromas: two case reports and literature review.

Cancer Genet Cytogenet 2010 Jan 1;196(1): p93-5

Liu K; De Angelo P, et al.

Intravascular embolization and surgical resection of a giant neurofibroma with intratumoral hemorrhage.

Br J Radiol 2010 Nov;83(995): pe225-9

Jones RG; Kiatisevi P, et al.

Mast cells and the neurofibroma microenvironment.

Blood 2010 Jul 15;116(2): p157-64

Staser K; Yang FC; Clapp DW

Dendritic cell neurofibroma with pseudorosettes: a report of 18 cases of a distinct and hitherto unrecognized neurofibroma variant.

Am J Surg Pathol 2001 May;25(5): p587-94

Michal M; Fanburg-Smith JC, et al.

**Case 2:**

A 36 y/o man presented with a 3 cm mass involving the right knee. The tumor was strongly positive for S100 protein, NSE and vimentin. Note focal plexiform pattern.

Santa Barbara (Miramonte Laboratory) - Neurofibroma  
Santa Rosa (Santa Rosa Memorial Hospital) - Neurofibroma, totally plexiform (2) /  
Neurofibroma, NOS  
Woodland Hills (Kaiser Permanente) - Neurofibroma  
Arkansas (The Laboratory of Pathology, P.A.) - Neurofibroma  
Georgia, Atlanta - Plexiform neurofibroma  
Illinois, Oak Brook - Neurofibroma  
Kansas (Peterson Laboratory Services) - Neurofibroma  
Louisiana, Shreveport (LSU Shreveport) - Neurofibroma  
Maryland, Baltimore - Neurofibroma, focally myxoid  
New Mexico, Albuquerque - Intraneural neurofibroma  
Ohio, Columbus - Soft tissue, right knee, neurofibroma  
Texas, Victoria - NF, plexiform, intraneural  
Hong Kong (Hong Kong Baptist Hospital) - Plexiform neurofibroma  
India, Roohi - Myxoid neurofibroma of knee  
The Netherlands, Amstelveen - Myxoid neurofibroma

**Dx:**

Neurofibroma with myxoid change (myxoid neurofibroma), knee

**Refs:**

Periungual myxoid neurofibroma.

Cutis 2002 Jan;69(1): p54-6

Gmyrek RF; Beer R, et al.

Subungual myxoid neurofibroma on the thumb.

Acta Derm Venereol 2001 Jun-Jul;81(3): p210-1

Baran R; Haneke E

Myxoid neurofibromas of the breast: mammographical, sonographical  
and MRI appearances.

Br J Radiol 2007 Oct;80(958): p234-7

Gokalp G; Hakyemez B, et al.

Solitary myxoid neurofibroma of the soft palate.

J Craniofac Surg 2011 Sep;22(5): p1901-3

Choi JS; Jang TJ

Myxoid neurofibroma in a child. An asymptomatic nodule on the  
finger.

Eur J Dermatol 2011 Mar-Apr;21(2): p267-8

Patrizi A; Medri M, et al.

**Case 3:**

A 47 y/o woman had this 2 cm mass in the right knee region.

Santa Barbara (Miramonte Laboratory) - Myxoid schwannoma  
Santa Rosa (Santa Rosa Memorial Hospital) - Neurofibroma, plexiform (3)  
Woodland Hills (Kaiser Permanente) - Dermal nerve sheath myxoma  
Arkansas (The Laboratory of Pathology, P.A.) - Myxoid neurofibroma  
Georgia, Atlanta - Nerve sheath myxoma  
Illinois, Oak Brook - Nerve sheath myxoma

Kansas (Peterson Laboratory Services) - Neurothekeoma  
Louisiana, Shreveport (LSU Shreveport) - Myxoid neurofibroma vs neurothekeoma  
Maryland, Baltimore - Myxoid plexiform schwannoma vs. neurothekeoma  
New Mexico, Albuquerque - Plexiform neurofibroma  
Ohio, Columbus - Soft tissue, right knee, nerve sheath myxoma  
Texas, Victoria - Nerve sheath myxoma  
Hong Kong (Hong Kong Baptist Hospital) - Neurothekeoma  
India, Roohi - Neurothekeoma of knee  
The Netherlands, Amstelveen - Neurothekeoma

**Dx:**

Neurothekeoma (nerve sheath myxoma), knee

**Refs:**

Neurothekeoma and plexiform fibrohistiocytic tumor: mere histologic resemblance or histogenetic relationship?

Am J Surg Pathol 2009 Jun;33(6): p905-13

Jaffer S; Ambrosini-Spaltro A; Mancini AM; Eusebi V; Rosai J

Is plexiform fibro-histiocytic tumor a deep form of cellular neurothekeoma?

J Cutan Pathol 2009 Oct;36(10): p1123-5

Leclerc-Mercier S; Brousse N; Fraitag S

Cellular neurothekeoma with fascicular growth features mimicking cellular dermatofibroma.

Am J Dermatopathol 2011 May;33(3): p281-4

Thakral B; Gleason BC; Thomas AB; Billings SD; Victor TA; Cibull TL

Neurothekeoma: an analysis of 178 tumors with detailed immunohistochemical data and long-term patient follow-up information.

Am J Surg Pathol 2007 Jul;31(7): p1103-14

Fetsch JF; Laskin WB; Hallman JR; Lupton GP; Miettinen M

Cellular neurothekeoma: detailed characterization in a series of 133 cases.

Am J Surg Pathol 2007 Mar;31(3): p329-40

Hornick JL; Fletcher CD

An update on plexiform fibrohistiocytic tumor and addition of 66 new cases from the Armed Forces Institute of Pathology, in honor of Franz M. Enzinger, MD.

Ann Diagn Pathol 2007 Oct;11(5): p313-9

Moosavi C; Jha P; Fanburg-Smith JC

**Case 4:**

This 1.5 cm nodule is from the palmar aspect of the left middle finger of a 27 y/o female. Clinically it was felt to be a cyst. Case seen by Dr. Chris Fletcher and contributed by Dr. Mark Carter.

Santa Barbara (Miramonte Laboratory) - Neurofibroma

Santa Rosa (Santa Rosa Memorial Hospital) - Neurothekeoma (myxoid nerve sheath tumor) (3)

Woodland Hills (Kaiser Permanente) - Cellular neurothekeoma  
Arkansas (The Laboratory of Pathology, P.A.) - Perineurioma  
Georgia, Atlanta - Neurothekeoma  
Illinois, Oak Brook - Nerve sheath myxoma  
Kansas (Peterson Laboratory Services) - Myxoid neurofibroma  
Louisiana, Shreveport (LSU Shreveport) - Perineurioma  
Maryland, Baltimore - Perineurioma  
New Mexico, Albuquerque - Nerve sheath myxoma  
Ohio, Columbus - Soft tissue, left middle finger, benign myxoid tumor  
Texas, Victoria - Schwannoma, finger  
Hong Kong (Hong Kong Baptist Hospital) - Nerve sheath myxoma  
India, Rochi - Perineurinoma of finger  
The Netherlands, Amstelveen - Perineurioma?

**Dx:**

Perineurioma, finger

**Refs:**

Perineurioma: a distinctive and underrecognized peripheral nerve sheath neoplasm.

Arch Pathol Lab Med 2007 Apr;131(4): p625-36

Macarenco RS; Ellinger F; Oliveira AM

Hybrid schwannoma/perineurioma: clinicopathologic analysis of 42 distinctive benign nerve sheath tumors.

Am J Surg Pathol 2009 Oct;33(10): p1554-61

Hornick JL; Bundock EA; Fletcher CD

Longitudinal study of intraneural perineurioma--a benign, focal hypertrophic neuropathy of youth.

Mauermann ML; Amrami KK; Kuntz NL; Spinner RJ; Dyck PJ; Bosch EP;

Engelstad J; Felmlee JP; Dyck PJ

Intraneural perineurioma: a systematic review with illustrative cases.

Arch Pathol Lab Med 2007 Sep;131(9): p1382-92

Boyanton BL; Jones JK; Shenaq SM; Hicks MJ; Bhattacharjee MB

Extra-acral cutaneous/soft tissue sclerosing perineurioma: an under-recognized entity in the differential of CD34-positive cutaneous neoplasms.

J Cutan Pathol 2010 Oct;37(10): p1053-6

Fox MD; Gleason BC; Thomas AB; Victor TA; Cibull TL

**Case 5:**

A 30 y/o woman was found to have this 6 cm mass in the axilla. Two weeks earlier she had been stung by a bee on the ipsilateral hand. Seen by A. Folpe, Mayo Clinic.

Santa Barbara (Miramonte Laboratory) - Low-grade leiomyosarcoma

Santa Rosa (Santa Rosa Memorial Hospital) - Malignant peripheral nerve sheath tumor (3)

Woodland Hills (Kaiser Permanente) - Malignant peripheral nerve sheath tumor

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

Georgia, Atlanta - Perineurioma

Illinois, Oak Brook - Neurofibroma cellular  
 Kansas (Peterson Laboratory Services) - Malignant peripheral nerve sheath tumor  
 Louisiana, Shreveport (LSU Shreveport) - Perineurioma  
 Maryland, Baltimore - Malignant, favor MPNST  
 New Mexico, Albuquerque - Malignant tumor of peripheral nerve  
 Ohio, Columbus - Soft tissue, axilla, cellular spindle cell neoplasm  
 Texas, Victoria - MPNST, axilla  
 Hong Kong (Hong Kong Baptist Hospital) - Histiocytic proliferation ? infection  
 India, Roohi - Malignant perineurinoma of axilla  
 The Netherlands, Amstelveen - Mesenchymal proliferation?

**Dx:**

Malignant perineurioma, axilla

**Refs:**

Low-grade malignant soft-tissue perineurioma: interphase fluorescence in situ hybridization.

Pathol Int 2008 Nov;58(11): p718-22

Sato K; Ueda Y; Miwa S; Yokogawa A; Ozaki M; Katsuda S

Atypical soft tissue perineurioma in the tongue of a young girl.

Pathol Int 2010 Dec;60(12): p787-91

Adachi S; Doi R, et al.

Malignant peripheral nerve sheath tumor with perineurial differentiation: "malignant perineurioma"

J Cutan Pathol 2002 Jul;29(6): p362-7

Rosenberg AS; Langee CL; Stevens GL; Morgan MB

Low-grade malignant perineurioma (perineurial sarcoma) of soft tissue: a potential diagnostic pitfall on fine needle aspiration.

Ann Diagn Pathol 2005 Aug;9(4): p197-201

Suster D; Plaza JA; Shen R

Unusual malignant perineurioma of soft tissue.

Virchows Arch 2001 Aug;439(2): p212-4

Fukunaga M

**Case 6:**

A 33-year-old woman recently noticed a bump on her posterior scalp. Examination revealed a 3 x 2.5 cm, soft, mobile mass in the midline posterior scalp clinically thought to be an epidermoid cyst.

EMA	Positive
AE1/AE3	Focal positive
Vimentin	Positive
CD99	Positive
Progesterone Receptor	Positive
Estrogen Receptor	Negative
GCDFP	Negative
S100	Negative
Ber-EP4	Negative

Mammoglobin

Negative

Santa Barbara (Miramonte Laboratory) - Glomus tumor  
Santa Rosa (Santa Rosa Memorial Hospital) - Extracranial meningioma (3)  
Woodland Hills (Kaiser Permanente) - Meningioma  
Arkansas (The Laboratory of Pathology, P.A.) - Meningioma  
Georgia, Atlanta - Atypical meningioma  
Illinois, Oak Brook - Ectopic meningioma  
Kansas (Peterson Laboratory Services) - Meningioma  
Louisiana, Shreveport (LSU Shreveport) - Meningioma  
Maryland, Baltimore - Extracranial meningioma (cutaneous meningeal hamartoma)  
New Mexico, Albuquerque - Ectopic meningioma  
Ohio, Columbus - Soft tissue, scalp, meningioma  
Texas, Victoria - Lipomatous perineuroma  
Hong Kong (Hong Kong Baptist Hospital) - Cutaneous meningothelial tumor  
India, Roohi - Extracranial meningioma  
The Netherlands, Amstelveen - Meningioma?

**Dx:**

Extracranial meningioma, posterior scalp

**Refs:**

Ectopic extra-cranial meningioma presenting as an aural polyp.  
Eur Arch Otorhinolaryngol 2003 Jul;260(6): p322-4  
Uppal HS; Kabbani M; Reddy V; Kaur S

A case of neck ectopic meningioma.  
Neuropathology 2008 Apr;28(2): p157-9  
Sanei MH; Berjis N; Mahzouni P; Naimi A

Pathology and genetics of meningiomas.  
Semin Diagn Pathol 2011 Nov;28(4): p314-24  
Alahmadi H; Croul SE

A meningioma and its consequences for American history and the rise  
of neurosurgery.  
J Neurosurg 2011 Dec;115(6): p1067-71  
Ansari SF; Gianaris NG; Cohen-Gadol AA

**Case 7:**

A 57 y/o woman had a lower cervical-spinal cord tumor.

Santa Barbara (Miramonte Laboratory) - Chordoma  
Santa Rosa (Santa Rosa Memorial Hospital) - Hemangioblastoma (3)  
Woodland Hills (Kaiser Permanente) - Schwannoma  
Arkansas (The Laboratory of Pathology, P.A.) - Hemangioblastoma  
Georgia, Atlanta - Hemangioblastoma  
Illinois, Oak Brook - Hemangioblastoma  
Kansas (Peterson Laboratory Services) - Hemangioblastoma  
Louisiana, Shreveport (LSU Shreveport) - Hemangioblastoma  
Maryland, Baltimore - Hemangioblastoma  
New Mexico, Albuquerque - Ependymoma  
Ohio, Columbus - Spinal cord, cervical region, hemangioblastoma

Texas, Victoria - Paraganglioma  
Hong Kong (Hong Kong Baptist Hospital) - Hemangioblastoma  
India, Roohi - hemangioblastoma of spinal cord  
The Netherlands, Amstelveen - Hemangioblastoma

**Dx:**

Hemangioblastoma, spinal cord

**Refs:**

Spinal hemangioblastoma.

Am J Med Sci 2011 Sep;342(3): p240

Pruksakorn P; Siwanuwatn R; Snabboon T

Intraspinal hemangioblastoma: analysis of 22 cases.

J Clin Neurosci 2008 Dec;15(12): p1366-9

Dwarakanath S; Sharma BS; Mahapatra AK

Surgical treatment of intramedullary spinal cord tumors: prognosis and complications.

Spinal Cord 2008 Apr;46(4): p282-6

Nakamura M; Ishii K, et al.

Metastatic renal cell carcinoma to hemangioblastoma in von Hippel-Lindau disease.

Arch Pathol Lab Med 2007 Apr;131(4): p641-5

Polydorides AD; Rosenblum MK; Edgar MA

Spinal cord hemangioblastoma with extensive syringomyelia.

J Chin Med Assoc (China 2005 Jan;68(1): p40-4

Wu TC; Guo WY, et al.

The natural history of hemangioblastomas of the central nervous system in patients with von Hippel-Lindau disease.

J Neurosurg 2003 Jan;98(1): p82-94

Wanebo JE; Lonser RR; Glenn GM; Oldfield EH

**Case 8:**

A 22 y/o woman presented with seizures and headache. A CT showed a 3 cm left parietal tumor which was biopsied and resected. The tumor was GFAP positive and strongly p53 immunoreactive. Immunostains for EMA and synaptophysin were negative. Seen by neuropathologist Ravi Raghavan, MD, Loma Linda, California.

Santa Barbara (Miramonte Laboratory) - Astrocytoma

Santa Rosa (Santa Rosa Memorial Hospital) - Anaplastic ependymoma, WHO grade III (2) /

Anaplastic ependymoma

Woodland Hills (Kaiser Permanente) - Anaplastic astrocytoma

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

Georgia, Atlanta - Astroblastoma

Illinois, Oak Brook - Ependymoma

Kansas (Peterson Laboratory Services) - Anaplastic ependymoma

Louisiana, Shreveport (LSU Shreveport) - Astroblastoma

Maryland, Baltimore - Astroblastoma

New Mexico, Albuquerque - Papillary meningioma

Ohio, Columbus - Brain, left parietal, gemistocytic astrocytoma  
Texas, Victoria - Astrocytoma, pilocytic  
Hong Kong (Hong Kong Baptist Hospital) - Ependymoma  
India, Rochi - Astroblastoma  
The Netherlands, Amstelveen - Astrocytoma?

**Dx:**

Astroblastoma, left parietal lobe

**Refs:**

Two cases of pediatric high-grade astroblastoma with different clinical behavior.  
Tumori 2010 Jan-Feb;96(1): p160-3  
Mastrangelo S; Lauriola L; Coccia P; Puma N; Massimi L; Riccardi R

Malignant astroblastoma with rhabdoid morphology.  
J Neurooncol 2007 Jul;83(3): p277-8  
Bannykh SI; Fan X; Black KL

Astroblastoma in childhood: pathological and clinical analysis.  
Childs Nerv Syst 2005 Mar;21(3): p211-20  
Navarro R; Reitman AJ, et al.

Astroblastoma with unusual signet-ring-like cell components: a case report and literature review.  
Neuropathology 2002 Sep;22(3): p200-5  
Sugita Y; Terasaki M, et al.

**Case 9:**

A 15 y/o boy suffered from headaches, nausea and vomiting. A CT & MRI showed a 4.0 cm partially enhancing tumor. The resected tumor was tan to brown, weighed 7 grams, and formed a 3.2 x 2.5 x 2.4 cm aggregate.

Santa Barbara (Miramonte Laboratory) - Metastasis  
Santa Rosa (Santa Rosa Memorial Hospital) - Pineal parenchymal tumor of indeterminate differentials (3)  
Woodland Hills (Kaiser Permanente) - Anaplastic oligodendroglioma  
Arkansas (The Laboratory of Pathology, P.A.) - Anaplastic ependymoma  
Georgia, Atlanta - Germinoma w/o metastasis  
Illinois, Oak Brook - Oligodendroglioma  
Kansas (Peterson Laboratory Services) - Oligodendroglioma, anaplastic  
Louisiana, Shreveport (LSU Shreveport) - Ependymoma  
Maryland, Baltimore - Ependymoma, favor anaplastic  
New Mexico, Albuquerque - Pituitary adenoma with necrosis  
Ohio, Columbus - Brain, malignant neoplasm  
Texas, Victoria - Medulloblastoma  
Hong Kong (Hong Kong Baptist Hospital) - ? Germinoma  
India, Rochi - Anaplastic ependymoma of brain  
The Netherlands, Amstelveen - Medulloblastoma?

**Dx:**

Anaplastic ependymoma, brain

**Refs:**

Parenchymal anaplastic ependymoma with intratumoral hemorrhage: a case report.

Brain Tumor Pathol 2003;20(2): p85-8

Kojima A; Yamaguchi N, et al.

The histologic grade is a main prognostic factor for patients with intracranial ependymomas treated in the microneurosurgical era: an analysis of 258 patients.

Cancer 2004 Mar 15;100(6): p1230-7

Korshunov A; Golanov A; Sycheva R; Timirgaz V

Myxopapillary ependymoma with anaplastic features.

Pathol Int 2003 Oct;53(10): p700-3

Awaya H; Kaneko M, et al.

Highly anaplastic extraventricular ependymoma arising in an adult, mimicking metastatic adenocarcinoma with heavy stromal inflammation and emperipoiesis.

Pathol Int 2003 Aug;53(8): p539-46

Moritani S; Kushima R, et al.

Anaplastic ependymoma with cartilaginous and osseous metaplasia: report of a rare case and review of literature.

J Neurooncol 2007 Mar;82(1): p75-80

Mridha AR; Sharma MC, et al.

**Case 10:**

A 22 y/o man had a three day history of progressive confusion, generalized weakness, nausea and vomiting. A CT scan revealed a 4.0 x 3.5 cm mass in the right cerebellum. The tumor was positive for synaptophysin, S100, GFAP and Vimentin. It was negative for AE1/AE3/CAM5.2/CK8/EMA and SMA.

Santa Barbara (Miramonte Laboratory) - Medulloblastoma

Santa Rosa (Santa Rosa Memorial Hospital) - Medulloblastoma (3)

Woodland Hills (Kaiser Permanente) - Medulloblastoma

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

Georgia, Atlanta - Medulloblastoma

Illinois, Oak Brook - Medulloblastoma

Kansas (Peterson Laboratory Services) - Medulloblastoma

Louisiana, Shreveport (LSU Shreveport) - Medulloblastoma

Maryland, Baltimore - Medulloblastoma

New Mexico, Albuquerque - Medulloblastoma

Ohio, Columbus - Cerebellum, right, small cell malignant neoplasm consistent with medulloblastoma

Texas, Victoria - Medulloblastoma

Hong Kong (Hong Kong Baptist Hospital) - Medulloblastoma

India, Roohi - Medulloblastoma

The Netherlands, Amstelveen - Medulloblastoma

**Dx:**

Medulloblastoma, brain, posterior fossa

**Refs:**

Medulloblastoma.

J Child Neurol 2009 Nov;24(11): p1418-30

Dhall G

Medulloblastomas: update on a heterogeneous disease.

Curr Opin Oncol 2011 Nov;23(6): p630-7

Bourdeaut F; Miquel C; Alapetite C; Roujeau T; Doz F

Histology predicts a favorable outcome in young children with desmoplastic medulloblastoma: a report from the children's oncology group.

Cancer 2011 Jul 15;117(14): p3262-7

Leary SE; Zhou T; Holmes E; Geyer JR; Miller DC

Definition of disease-risk stratification groups in childhood medulloblastoma using combined clinical, pathologic, and molecular variables.

J Clin Oncol 2011 Apr 10;29(11): p1400-7

Ellison DW; Kocak M, et al.

