DIAGNOSTIC CASE PRESENTATIONS
CANP 54th annual meeting, Banff, Alberta
October 15th – 18th, 2014

Case C1.

P.W. Schutz[1], G.R.W. Moore[1], R. Akagami[2], I.R. Mackenzie[1]
[1]Department of Pathology [2]Division of Neurosurgery Vancouver General Hospital, University of British Columbia, Vancouver, BC, Canada

DIAGNOSIS :
Myxopapillary Ependymoma, multifocal presentation

REFERENCES:

Fassett DR, Pingree J, Kestle JR. The high incidence of tumor dissemination in myxopapillary ependymoma in pediatric patients. Report of five cases and review of the literature. *Journal of Neurosurgery* 2005;102(1 Suppl):59-64.

Higgins GS, Smith C, Summers DM, Statham PX, Erridge SC. Myxopapillary ependymoma with intracranial metastases. *Br J Neurosurg* 2005;19(4):356-358.

Shapey J, Barazi S, Bodi I, Thomas N. Myxopapillary ependymoma of the cerebellopontine angle: retrograde metastasis or primary tumour? *Br J Neurosurg* 2011;25(1):122-123.

Straus D, Tan LA, Takagi I, O'Toole JE. Disseminated spinal myxopapillary ependymoma in an adult at initial presentation: A case report and review of the literature. *Br J Neurosurg* 2014.

Case C2.

P Diamandis[1], MD Cusimano[2], K Kovacs[1], J Karamchandani[3]
[1]Department of Laboratory Medicine, Division of Pathology, St Michael's Hospital, University of Toronto, Toronto, ON, Canada. [2]Division of Neurosurgery, Department of Surgery, St. Michael's Hospital, University of Toronto, Toronto, ON, Canada.
[3]McGill University, Montreal Neurological Institute and Hospital, Montreal, Quebec, Canada.

DIAGNOSIS :
Phosphaturic mesenchymal tumor

REFERENCES:

Mathis DA, Stehel EJ Jr, Beshay JE, Mickey BE, Folpe AL, Raisanen J.
Intracranial phosphaturic mesenchymal tumors: report of 2 cases. *J Neurosurg*. 2013 Apr;118(4):903-7

Folpe AL, Fanburg-Smith JC, Billings SD, Bisceglia M, Bertoni F, Cho JY, *et al*.
Most osteomalacia-associated mesenchymal tumors are a single histopathologic entity: an analysis of 32 cases and a comprehensive review of the literature. *Am J Surg Pathol*. 2004 Jan;28(1):1-30.

David K, Revesz T, Kratimenos G, Krausz T, Crockard HA. Oncogenic osteomalacia
associated with a meningeal phosphaturic mesenchymal tumor. Case report.

J Neurosurg. 1996 Feb;84(2):288-92.

Weidner N, Santa Cruz D. Phosphaturic mesenchymal tumors. A polymorphous group causing osteomalacia or rickets. *Cancer*. 1987 Apr 15;59(8):1442-54.

Case C3.

Claire I. Coiré[1], And Warren P. Mason[2]
[1]Department of Pathology, Trillium Health Partners, Mississauga and [2]Department of Medical Oncology, Pencer Brain Tumor Centre, Princess Margaret Cancer Centre, University of Toronto.

DIAGNOSIS :
Secondary Gliosarcoma.with Epithelial and Pseudoepithelial Differentiation

REFERENCES:

Han SJ, Yang I, Otero JJ, Han BJ, Tihan T, McDermott MW, Berger MS, Chang SM, Parsa AT. Secondary Gliosarcoma After Diagnosis of Glioblastoma: Clinical Experience With 30 Consecutive Patients.
J Neurosurg 112:900-996, 2010.

Andaloussi-Saghir K, Oukabli M, El Marjany M, Sifat H, Hadadi K, Mansouri H. Secondary Gliosarcoma After the Treatment of Primary Glioblastoma Multiforme.
N Am J Med Sci 2011 November: 3(11): 527-530.

Rodigruez FJ, Scheithauer BW, Giannini C, Bryant SC, Jenkins RB.
Epithelial and Pseudoepithelial Differentiation in Glioblastoma and Gliosarcoma.
Cancer 2008;113:2779-89.

Pimentel J, Marques J, Pereira P, Roque L, Martins C, Campos A.
Sarcoma with True Epithelial Differentiation Secondary to Irradiated Glioblastoma.
Clinical Neuropathology, Vol 30 – N0 6/2011 (307-312).

Case C4.

J.P. Rossiter[1], A. Adeyinka[1] and J.G. Boyd[2]
Department of Pathology and Molecular Medicine[1] and Division of Neurology [2], Queen's University and Kingston General Hospital, Kingston, Ontario

DIAGNOSIS :
Extensive involvement of brain and spinal cord by primary diffuse large B-cell lymphoma ('Lymphomatosis cerebri et medullae spinalis').

REFERENCES:

Kitai R et al. (2013) Lymphomatosis cerebri: clinical characteristics, neuroimaging and pathological findings. *Brain Tumor Pathology* 29:47-53.

Courtois F et al. (2012) Lymphomatosis cerebri presenting as a recurrent leukoencephalopathy. *Case*

Reports in Neurology 4:181-186.

Rollins KE et al. (2005) Lymphomatosis cerebri as a cause of white matter dementia. Human Pathology 36:282-290.

Sato H et al. (2013) Lymphomatosis cerebri with intramedullary spinal cord involvement. Internal Medicine 52:2561-2565.

Case C5.

M. Alturkustani[1,2], F. AlSufiani[1], L-C. Ang[1]
[1] Division of Neuropathology, London Health Sciences Centre; University of Western, London, Canada;
[2] Department of Pathology, King Abdulaziz University and Hospital, Jeddah, Saudi Arabia

DIAGNOSIS :
Myopathy with Hexagonally Cross-Linked Crystalloid Inclusions

REFERENCES:

B. Lach *et al*. Sarcoplasmic hexagonally cross-linked tubular arrays immunostain for caveolin-3: an excess caveolinopathy? *Acta Neuropathol* (2009) 117:339–341

K .G. Claeys *et al*. Myopathy with hexagonally cross-linked crystalloid inclusions: Delineation of a clinico-pathological entity. *Neuromuscular Disorders* 20 (2010) 701–708

Case C6.

N. Sinha[1], R. Fraser[2], CJ. Fallet-Bianco[3] & A. Oviedo[2]
[1] Division of Anatomical Pathology, Queen Elizabeth II Health Sciences Centre and Dalhousie University, Halifax, Nova Scotia, Canada; [2] Department of Pathology & Laboratory Medicine, IWK Health Centre, Halifax, Canada [3] Department of Pathology and Neuropathology, Hôpital Sainte-Justine, Montreal, Canada

DIAGNOSIS :
Bilateral brain stem tegmental calcification (brainstem watershed infarcts)

REFERENCES:

Sarnat HB. Watershed infarcts in the fetal and neonatal brainstem. An aetiology of central hypoventilation, dysphagia, Möbius syndrome and micrognathia. *Eur J Paed Neurol* (2004) 8, 71–87.

Bavinck JN, Weaver DD. Subclavian artery supply disruption sequence: hypothesis of a vascular etiology for Poland, Klippel-Feil and Moebius anomaly. *Am J Med Genet* (1986) 23:903–918.

Matsunaga Y, Amamoto N, Kondoh T et al. A severe case of Moebius syndrome with calcification on the fourth ventricular floor. *Hum Genet* (1998) 43:62–64.

Case C7.

Ana Nikolic[1], Kevin Fonseca[2], Alberto Severini[3], Konstantin Koro[4], Sandra Lee[4] and Jeffrey T. Joseph[1]
[1] Department of Pathology and Laboratory Medicine, Division of Neuropathology [4] Department of Pathology and Laboratory Medicine, Division of Anatomic Pathology, University of Calgary, Calgary, Alberta [2] Provincial Laboratory of Public Health, University of Calgary, Alberta [3] National Microbiology Laboratory, Winnipeg, Manitoba

DIAGNOSIS :
Subacute sclerosing panencephalitis

Note: This autopsy is interesting because the patient had many SSPE viral inclusions remaining in her brain at the time of death. These are very obvious on H&E. It is also interesting because her SSPE seems to have been induced by her pregnancy. In our discussion, we will comment on the strain of measles identified and the risk of transmission to the infant.

REFERENCES:

Ogbuanu IU, Zeko S, Chu SY, Muroua C, Gerber S, De Wee R, Kretsinger K, Wannemuehler K, Gerndt K, Allies M, Sandhu HS, Goodson JL. Maternal, fetal, and neonatal outcomes associated with measles during pregnancy: Namibia, 2009-2010. *Clin Infect Dis*. 2014 Apr; 58(8):1086-92

Prashanth LK, Taly AB, Ravi V, Sinha S, Arunodaya GR. Adult onset subacute sclerosing panencephalitis: clinical profile of 39 patients from a tertiary care centre. *J Neurol Neurosurg Psychiatry*. 2006 May;77(5):630-3

Wirguin I, Steiner I, Kidron D, Brenner T, Udem S, Rager B, Abramsky O. Fulminant subacute sclerosing panencephalitis in association with pregnancy. *Arch Neurol*. 1988 Dec;45(12):1324-5

Case C8.

J. Ng[1], P. Alcaide-Leon[1], D. Ghazarian[2], DG Munoz[1]
[1] Department of Laboratory Medicine, Division of Pathology, St Michael's Hospital, University of Toronto, Toronto, ON, Canada.

DIAGNOSIS :
Psammomatous melanotic schwannoma

REFERENCES:

Carney JA. Psammomatous melanotic schwannoma. A distinctive, heritable tumor with special associations, including cardiac myxoma and the Cushing syndrome. *Am J Surg Pathol 1990*;14:206-22.

Shields LB, Glassman SD, Raque GH, Shields CB. Malignant psammomatous melanotic schwannoma of the spine: A component of Carney complex. *Surg Neurol Int*. 2011;2:136.

Marton E, Feletti A, Orvieto E, Longatti P. Dumbbell-shaped C-2 psammomatous melanotic malignant schwannoma. Case report and review of the literature. *J Neurosurg Spine*. 2007;6(6):591-9.

Wilkes D, McDermott DA, Basson CT. Clinical phenotypes and molecular genetic mechanisms of

Carney complex. *Lancet Oncol*. 2005 Jul;6(7):501-8

Horvath A, Bossis I, Giatzakis C, Levine E, Weinberg F, Meoli E, *et al*. Large deletions of the PRKAR1A gene in Carney complex. *Clin Cancer Res*. 2008 Jan 15;14(2):388-95

Case C9.

Maxime Richer[1], John Wong[1], Alden Chesney[2], Mina Jamali[2], Nicholas Phan[3], Julia Keith[1]
[1] Department of Anatomical Pathology, Sunnybrook Health Sciences, University of Toronto, Toronto, Ontario, Canada [2] Department of Clinical Pathology, Sunnybrook Health Sciences, University of Toronto, Toronto, Ontario, Canada
[3] Division of Neurosurgery, Sunnybrook Health Sciences, University of Toronto, Toronto, Ontario, Canada

DIAGNOSIS :
Acute Promyelocytic Leukemia with t(15;17)

REFERENCES:

Seifert RP, Bulkeley W 3rd, Zhang L *et al*. A practical approach to diagnose soft tissue myeloid sarcoma preceding or coinciding with acute myeloid leukemia. *Ann Diagn Pathol* 2014. 8(4):253-260.

Swerdlow *et al*. (2009) WHO Classification of Tumours of Haematopoietic and Lymphoid Tissues. WHO IARC, p439.

Case C10.

S. Magaki[1], E. Chang[2], J. Jen[2], and H. V. Vinters[1,2]
[1]Section of Neuropathology, Department of Pathology and Laboratory Medicine, UCLA Medical Center, Los Angeles, CA; [2]Department of Neurology, UCLA Medical Center, Los Angeles, CA.

DIAGNOSIS :
Rheumatoid meningitis

REFERENCES:

Kim RC, Collins GH. The neuropathology of rheumatoid disease. *Hum Pathol* 12, 5–15 (1981).

Servioli MJ, Chugh C, Lee JM & Biller J. Rheumatoid meningitis. *Front. Neurol*. 2, 84 (2011).

Case C11.

D.W. Ng[1], G. Dalgliesh[2], R.L. Barnhill[1] and H.V. Vinters[1]
Division of Neuropathology, Department of Pathology, University of California Los Angeles, Los Angeles, California[1] Department of Pathology, University of California Los Angeles, Los Angeles, California[2]

DIAGNOSIS :
Intradural atypical melanocytic neoplasm (see comment)

Comment: This tumor has features of a melanocytic neoplasm that is atypical, hypercellular, and proliferative. This shows features in common with atypical variants of cellular blue nevus in the skin and soft tissues and is positive for S100 protein, Mart-1, HMB-45, tyrosinase, MITF, and Sox 10. The lesion has a mitotic rate of 5 per mm^2. Although this tumor appears to lack sufficient criteria for conventional melanoma at this point in its analysis, the features are sufficient to warrant the characterization as a neoplasm with uncertain malignant potential and possibly as a low-grade malignancy.

REFERENCES:

Barnhill RL, *et al*. Atypical cellular blue nevi (cellular blue nevi with atypical features): lack of consensus for diagnosis and distinction from cellular blue nevi and malignant melanoma (‘‘malignant blue nevus’’). *Am J Surg Pathol*. 2008;32(1):36–44.

Bastian, BC. The molecular pathology of melanoma: an integrated taxonomy of melanocytic neoplasia. *Ann Rev Path.* 2014;9: 239–71.

Brat DJ, *et al*. Primary melanocytic neoplasms of the central nervous system. *Am J Surg Path*. 1999;23(7):745-54.

Liubinas SV, Maartens N, Drummond KJ. Primary melanocytic neoplasms of the central nervous system. *J Clin Neurosci*. 2010;13:1227-32.

Wang, H *et al*. Melanocytomas of the central nervous system: a clinicopathological and molecular study. *Eur J Clin Invest*. 2013:43,809–15.

Case C12.

Claire I. Coiré[1] and David Munoz[2]
Departments of Laboratory Medicine [1] Trillium Health Partners and [2] Saint Michael’s Hospital, University of Toronto.

DIAGNOSIS :
Lymphomatoid Granulomatosis presenting as an intramedullary cervical mass

REFERENCES:

AL Katzenstein, E Doxtader, S Narendra. Lymphomatoid Granulomatosis Insights Gained Over 4 Decades. *Am J Surg Pathol* 2010;34:e35-e48.

M Moschewski, WH Wilson. Lymphomatoid Granulomatosis. Review Article.
Cancer J 2012;18:469-474.

S Collins, RD Helme Lymphomatoid Granulomatosis Presenting as a Progressive Cervical Cord Lesion. *Aust NZ J Med* 1989;19:144-146.

C Lucantoni, P De Bonis, F Doglietto, G Esposito, LM Larocca, A Mangiola, *et al*.
Primary Cerebral Lymphomatoid Granulomatosis: report of four cases and literature review.

J Neurooncol 2009;94:235-242.

Case C13.

J. T. Joseph[1], D. M. Pearson[2], A. Khan[3]
[1]Department of Pathology and Laboratory Medicine, [2]Department of Clinical Neuroscience,
[3]Department of Medical Genetics and Pediatrics, University of Calgary, Calgary, Alberta

DIAGNOSIS :
Tauopathy; MAPT gene mutation G389R

Note: This autopsy is interesting because of the young age of the patient and the presentation with dysarthria. The distribution of abnormal tau in the brain is also interesting - predominantly in cortical layer 2 and deep layers, plus in the subcortical white matter.

REFERENCES:

Bermingham N, Cowie TF, Paine M, Storey E, McLean C. Frontotemporal dementia and Parkinsonism linked to chromosome 17 in a young Australian patient with the G389R Tau mutation. *Neuropathol Appl Neurobiol*. 2008 Jun;34(3):366-70

Chaunu MP, Deramecourt V, Buée-Scherrer V, Le Ber I, Brice A, Ehrle N, *et al*. Juvenile frontotemporal dementia with parkinsonism associated with tau mutation G389R. *J Alzheimers Dis*. 2013;37(4):769-76

Murrell JR, Spillantini MG, Zolo P, Guazzelli M, Smith MJ, Hasegawa M, *et al*. Tau gene mutation G389R causes a tauopathy with abundant pick body-like inclusions and axonal deposits. *J Neuropathol Exp Neurol*. 1999 Dec;58(12):1207-26.