Canadian Association of Neuropathologists

L'Association Canadienne des Neuropathologistes

CASE No 2

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This 17 yo male was diagnosed with NF1 several years before his last admission. Numerous café au lait spots and plexiform neurofibromas at various anatomical sites had been well documented by MRI. Some were biopsied and diagnosed as consistent with benign plexiform neurofibromas. A neurofibrosarcoma of the pelvis had been diagnosed before, but appeared to be rather slowly growing since many years, without further evidence of malignant behavior. He presented last with nausea, vomiting and obtundation due to a block of a ventriculo-peritoneal shunt for communicating hydrocephalus. An MRI revealed a diffuse leptomeningeal enhancement of supra and infratentorial compartments, extending around most of the spinal cord. A biopsy of the thoracic extension of the enhancing perimedullary lesion was submitted.

Materials submitted: One representative HPS digital slide

Questions: Diagnosis?
Significance in the context of NF1