innervation of the sphenoid sinus. This explains the similarities of the sphenoid sinus headache and migraine headache, both involving a trigeminovascular mechanism. A high index of suspicion is required in diagnosis. Sometimes the diagnosis is made incidentally, following an MRI obtained because of chronic headaches. In acute cases, CT scan may be most practical, and prompt treatment with antibiotics is mandatory.

The sphenoid sinus may be the site of an infiltrative tumor (Langerhans' cell histiocytosis) associated with retro-orbital pain and frontal headache (see Headache section below for case-report in current issue of N Engl J Med Feb 14, 2002).

Spontaneous extradural hematoma with sinusitis is an unusual complication of frontal sinusitis, in a case report of a 17 year-old man treated at Princess Royal Hospital, Haywards Heath, UK (Papadopoulos MC et al. IRSM Nov 2001;94:588-589). The patient developed a sudden exacerbation of headache, with nausea and vomiting, 3 days after antibiotic treatment for frontal sinusitis. CT showed a right frontal extradural hematoma which was evacuated surgically, with recovery. Spread of infection beyond the sinus was supported by histological and radiological evidence of inflammation in contiguous bone and dura. Bleeding was explained by weakened dural vessels, secondary to infection. A dissenting opinion was expressed in a letter to the editor (Gordon AG. IRSM February 2002;95:110-111), favoring dural pneumodissection secondary to a traumatic fistula after coughing or sneezing. Gordon cites several examples of acute severe headache with traumatic fistula and extradural hematoma, secondary to barotrauma, violent nose-blowing, or violent wheezing and asthma.

Pott's puffy tumor is another unusual complication of frontal sinusitis, reported in 7 childhood cases, ages 11 to 18 years, and seen at Case Western Reserve University School of Medicine, Cleveland, OH (Babakidis NC, Cohen AR. Pediatr Neurosurg Aug 2001;35:82-89). First described by Sir Percival Pott, surgeon at St Bartholomew's Hospital, London, in 1775, "Pott's puffy tumor" is a localized swelling of the forehead, secondary to subperiosteal or epidural abscess and a complication of trauma or frontal sinusitis. Despite the advent of antibiotics, 21 cases have been reported in children in the last 20 years. Headache, fever, nasal drainage, and frontal sinus tenderness are common presenting complaints. Four of 7 cases had acute neurological complications, including aphasia, hemiparesis, obtundation, and III nerve palsy. Intracranial infections were epidural abscess in 5 patients, subdural empyema in 4, and brain abscess in 1. All underwent craniotomy. The scalp was edematous and purulent. Recovery followed surgery and antibiotic therapy. In adults, another cause of Pott's puffy tumor is aseptic meningeal metastatic carcinomatosis (personal observation).

ATAXIAS

LANGERHANS CELL HISTIOCYTOSIS AND ACUTE ATAXIA

A 3-year-old boy who presented with an acute cerebellar ataxia and x-ray evidence of apparent chest infection, caused by massive infiltration of Langerhans histiocytes, is reported from the Department of Pediatrics, University of Catania, Italy, Northampton General Hospital, and John Radcliffe Hospital, Oxford, UK. Ataxia and slurred speech had been present for 6 weeks before admission. Brain CT and MRI were normal. Chest X-ray showed widespread reticulonodular shadowing, suggestive of mycoplasma pneumonia or metastatic disease. After 2 days treatment with erythromycin for presumed pneumonia, the

patient was readmitted as an emergency because of vomiting, drowsiness, and dyspnea, and he died soon after arrival. Autopsy revealed bilateral tension pneumothorax, diffuse bronchiectatic cavities, and generalized congestion of meningeal vessels. Histologically the lungs and spleen were infiltrated by Langerhans cells. The cerebellum showed focal gliosis associated with Purkinje cell loss. (Polizzi A, Coghill S, McShane MA, Squier W. Acute ataxia complicating Langerhans cell histiocytosis. Arch Dis Child February 2002;86:130-131). (Respond: Dr A Polizzi, Department of Paediatrics, University of Catania, Via S Sofia, 78, I-95123 Catania, Italy).

COMMENT. The diagnosis of Langerhans' cell histiocytosis (LCH) is made by the electron micrograph finding of Birbeck granules, membranous cytoplasmic structures, 200-400 nm in width and shaped like tennis rackets, and CD1a and S-100 protein antigen on the cell surface (Faquin WC. In case-report. Clinicopathological Exercise. N Engl J Med February 14, 2002;346:513-520). The presentation is usually as bone lesions and dermatitis, hepatosplenomegaly, lymphadenopathy, pancytopenia, fever, and weight loss. CNS manifestations are rarely cerebellar, and usually present with diabetes insipidus from hypothalamic or pituitary infiltration. In the rare cases of cerebellar involvement cited in the pediatric literature, symptoms were chronic and progressive and presented years after LCH diagnosis. The absence of cerebellar infiltration supports a paraneoplastic syndrome and immune mediated mechanism. In the above case report, the primary LCH location was lung involvement. In the N Engl J Med report (Feb 14, 2002), the primary location of LCH was a retro-orbital spenoid sinus mass, and the presenting symptom was frontal pain (see below in Headache section).

CEREBELLAR HEMANGIOBLASTOMA AND VON HIPPEL-LINDAU DISEASE

Six pediatric patients with cerebellar hemangioblastoma were screened for germline or somatic mutations of the von Hippel-Landau gene, in a study at Stanford University Medical Center, Palo Alto, CA. Two testing positive for mutations had a history of clinical manifestations of von Hippel-Lindau disease, whereas 4 testing negative had solitary hemangioblastoma and no history of the disease. These solitary cases may involve a molecular process unrelated to the von Hippel-Lindau tumor suppressor pathway. (Fisher PG, Tontiplaphol A, Pearlman EM et al. Childhood cerebellar hemangioblastoma does not predict germline or somatic mutations in the von Hippel-Lindau tumor suppressor gene. Ann Neurol February 2000;51:257-260). (Respond: Dr Paul G Fisher, Room A343, Department of Neurology, Stanford University Medical Center, 300 Pasteur Drive, Palo Alto, CA 94305).

COMMENT. Hemangioblastoma is a rare intracranial tumor, accounting for less than 0.5% of all pediatric brain tumors (Ries et al, 2001). The apparent absence of von Hippel-Lindau (VHL) gene abnormalities in childhood sporadic cerebellar hemangioblastoma indicates an alternative mechanism of tumorigenesis from that in adults. In 3 reports including 50 adult cases of sporadic cerebellar hemangioblastoma cited by the authors, about 50% of patients screened had somatic VHL gene mutations. VHL disease is familial with dominant inheritance and manifested by malignant and benign neoplasms of eyes, kidneys, adrenal glands, spine, and brain. The VHL gene is a tumor suppressor gene located on chromosome 3p25-26.