NEUROMUSCULAR DISORDERS

NEOPLASTIC CAUSE OF CONGENITAL BRACHIAL PLEXUS PALSY

Two patients with a neonatal onset of arm weakness and neoplastic involvement of the brachial plexus are reported from the Miami Children's Hospital, FL. The initial diagnosis was an obstetric brachial plexus palsy. 1). A 7-day-old female presented with a left supraclavicular malignant rhabdoid tumor and left proximal arm weakness, first noted at 2 days of age. Weakness quickly progressed to involve the whole arm within 3 days. MRI showed involvement of the brachial plexus, 4th and 5th cervical roots, and the spinal canal. 2). A 28-month-old male presented with scratch marks over the right upper extremity, beginning at 4 months of age, and a slowly progressive right arm weakness, first noted at 3 weeks of age. MRI revealed a plexiform neurofibroma of the right brachial plexus, extending from all cervical to the 5th thoracic spinal roots. (Alfonso I, Papazian O, Prieto G, Alfonso DT, Melnick SJ. Neoplasm as a cause of brachial plexus palsy in neonates. Pediatr Neurol April 2000;22:309-311). (Respond: Dr Israel Alfonso, Department of Neurology, Miami Children's Hospital, 3100 SW 62nd Ave, Miami, FL 33155).

COMMENT. In obstetric brachial plexus palsy, the weakness is apparent at birth and is non-progressive, and the birth process is usually complicated by shoulder dystocia. In infants with brachial plexus palsy caused by neoplasm, the onset of weakness is after the first day, the course is progressive, and the delivery is usually normal and free from traumatic injury. The diagnosis is confirmed by MRI.

SPINAL LESIONS

NEONATAL SPINAL NEUROENTERIC CYST

A neonatal spinal neuroenteric cyst (NC), presenting with persistent fever and acute myelopathy at the age of 10 days, is reported from the University Catholique de Louvain, Brussels, Belgium. Chest X-ray showed enlargement of interpeduncular spaces, vertebral body clefts, and a paravertebral mass. Ultrasonography of the spinal canal revealed an anterior cystic mass. MRI confirmed the spinal malformation and its connection through the vertebral defect with the prevertebral, retro-aortic cystic mass. At 8 weeks of age, the infant developed acute hypertonia, ankle clonus, and paresis of the lower limbs and left arm, and urinary retention. Thoracic laminectomy at T6 to T9 for partial resection of the intradural cyst was followed, at age 8 months, by excision of the mediastinal component of the cyst. At 18 months, fever, paraplegia, and neurologic bladder persisted while motor function of the left arm improved. The cyst consisted of smooth muscle entrapping cavities lined by digestive and respiratory epithelia, and chronic inflammatory cell infiltration. (Kadhim H, Proano PG, Saint Martin C et al. Spinal neuroenteric cyst presenting in infancy with chronic fever and acute myelopathy. Neurology May 2000;54:2011-2015). (Reprints: Dt Guillaume Sebire, Service de Neurologie Pediatrique, Cliniques Universitaires Saint Luc, Avenue Hippocrate 10/1068, 1200 Brussels, Belgium).

COMMENT. The authors refer to 13 additional cases of infantile neuroenteric cyst reported in the literature. A sudden spinal cord compression is precipitated by an increase in size of the cyst due to inflammation and retention of mucous

secretion. Diagnosis is by Chest X-ray, ultrasonography, and MRI. The value of ultrasound in diagnosis is stressed. Recurrences of the cyst after surgery are reported, even in adulthood.

INFECTIOUS DISORDERS

CLINICAL MANIFESTATIONS OF NEUROCYSTICERCOSIS

A retrospective study of 25 children with neurocysticercosis is reported from the University of the State of Sao Paulo School of Medicine, Brazil. The age range was 1 to 11 years (mean, 8 years 6 months), and 68% resided in urban areas. Clinical manifestations included seizures, mainly partial in type (72%), headache (60%), learning disability (24%), behavioral changes (12%), developmental regression (8%), and intracranial hypertension (4%). The neurologic exam was normal in 80%. Eosinophilia occurred in 36%, and the CSF showed increased protein in 37%. Neuroimaging studies showed calcifications in 60%, cysts (12%), calcifications and cysts (16%), and lateral ventricle asymmetry (4%). The majority received only symptomatic drugs in treatment, and remission occurred in 43%, while 48% improved. Cysticidil drugs were given in only 3 (praziquantel in 1 and albendazole in 2), with complete remission in 1 and improvement in 2. (Morales NMO, Agapejev S, Morales RR et al. Clinical aspects of neurocysticercosis in children. Pediatr Neurol April 2000;22:287-291). (Respond: Dr Agapejev, Department of Neurology and Psychiatry, University of the State of Sao Paulo School of Medicine, PO Box 540, Botucatu, Sao Paulo 18618-000 Brazil).

COMMENT. In endemic areas, neurocysticercosis should be considered in children presenting with learning disabilities, behavioral abnormalities, and developmental regression. The course is often benign, many recovering spontaneously without cysticidal therapy. The disease is an indication of poverty and poor hygiene. Improved sanitation and health care are important in prevention.

NEUROLOGICAL COMPLICATIONS OF SINUS INFECTION

The neurological manifestations of deep sinus infection in 6 children are reported from the University of Crete, Heraklion, Greece. Ages ranged from 8 to 14 years. Infection of sphenoid and posterior ethmoid sinuses was confirmed by CT. The admitting clinical diagnosis was epilepsy in 2 patients, migraine in 2, CNS tumor in 1, and CNS infection in 1. Presenting neurologic signs included focal convulsions in 1, blurred vision and scotomata in 2, vertigo and ataxia in 1, and nuchal rigidity in 1. Complete recovery occurred after treatment with intravenous antibiotics and resolution of the sinusitis. (Bitsori M, Galanakis E, Kokori H, Sbyrakis S. Neurologic manifestations of deep sinus infection. Acta Paediatr April 2000;89:490-491). (Respond: Maria Bitsori MD, Department fo Paediatrics, University Hospital of Heraklion, PO Box 1352, 711 10 Heraklion, Greece).

COMMENT. Incidental reports of sinusitis are common in children undergoing neuroimaging studies for suspected intracranial disease. Sphenoidal and posterior ethmoidal sinuses are involved infrequently, and deep sinus infection should be considered in the differential diagnosis of some epilepsies, headache, ataxia, and meningeal irritation.