Angiographically cryptic AVM presenting as a pontine tumor

Case report

A. LELAND ALBRIGHT, M.D., RYLAND P. BYRD, M.D., AND MARY LEE HARRISON, M.D.

Department of Surgery, Division of Neurological Surgery, and Department of Pathology, University of Louisville School of Medicine, Louisville, Kentucky

A 19-month-old child became disoriented and ataxic 6 days after a diphtheria-pertussis-tetanus (DPT) booster injection. Computerized tomographic scan revealed a hyperdense mesencephalic-pontine mass, and angiography demonstrated an avascular mass. Pontine biopsy revealed no abnormality. Her clinical course fluctuated until her death 15 months later. At autopsy, an angiographically cryptic arteriovenous malformation (AVM) was present in the mesencephalon and pons. Vascular malformations of the brain stem, producing prolonged clinical courses, are reviewed.

KEY WORDS □9 brain stem □9 brain-stem tumor □9 cryptic arteriovenous malformation

The neurological abnormalities of children with brain-stem tumors have been well characterized. The management of those children, however, has been controversial. Some authors have advocated radiotherapy without histological confirmation of the diagnosis, because of the operative risks involved. Other authors have noted that identical clinical findings can result from encephalitis, abscess, or vascular malformations, and have thought that a histological diagnosis allows more rational subsequent therapy.

We report the case of an infant with a pontine arteriovenous malformation (AVM) not apparent on angiography or pontine biopsy. This patient became symptomatic after a diphtheria-pertussis-tetanus (DPT) injection.

Case Report

This 19-month-old girl was seen by her pediatrician because of unsteadiness and alternating spells of rage and sleeping for 3 to 4 days. Six days earlier, she had received a DPT booster injection. At 4 months of age, she had received a DPT injection that was followed 2 weeks later by a right exophoria and limited vertical gaze that persisted for 4 to 5 days.

Examination. She was febrile (39.5°C) and irritable, with a right abducens paresis and marked truncal and appendicular ataxia. Skull x-ray films were normal. Computerized tomography (CT) demonstrated an area of increased pontine density that was enhanced by contrast administration (Fig. 1). Vertebral angiography indicated the presence of an avascular mass. Pneumoencephalography revealed marked enlargement of the pons and posterior displacement of the aqueduct of Sylvius.

Operation. She was treated with dexamethasone, and a right retromastoid craniectomy was performed. The basis pontis appeared enlarged. Biopsy specimens of the basis pontis were normal. Within 2 weeks after operation, her ataxia and listlessness had greatly improved.

Postoperative Course. One month postoperatively, she became more ataxic and irritable, and bilateral abducens paresis was evident. Her cranial sutures were diastatic, and a CT scan revealed enlarged ventricles and subarachnoid spaces. The pontine mass appeared to be smaller. A left ventriculoperitoneal shunt was inserted, and her symptoms and signs improved slowly.

By 23 months of age, the patient was normal except for mild reflex asymmetry and left esophoria. At 24 months, she abruptly developed irritability, ataxia, and worsened abducens paresis, but those symptoms...
Cryptic AVM resulting as pontine tumor

improved within 1 week. Her CT scan was unchanged. At 27 months of age, she had no evident neurological abnormality.

During her 28th month, she became progressively irritable, ataxic, and anorexic. The pontine mass appeared to be larger on CT scanning. Numerous medications were given to lessen her apparent pain, agitation, and extreme wakefulness, but without effect. She then received radiotherapy, 1600 rads, but developed a right facial and left extremity paresis. During the next 2 months, her condition fluctuated and slowly worsened. She was often obtunded and irritable, and experienced shaking in the extremities. At 34 months of age, she developed dysphagia, became comatose, and died.

Postmortem Examination. On gross examination, a moderate amount of blood pigment was evident in the subarachnoid spaces of the base of the brain and ventral surface of the pons. The pons appeared to be smaller than normal. The ependyma of all ventricles was stained with blood pigment. A hemorrhagic mass was present from the upper mesencephalon to the pons, with a globular extension into the fourth ventricle (Fig. 2). Mild cortical atrophy was evident.

A mesencephalic-pontine AVM was found on microscopic examination, with numerous thrombosed vessels and areas of calcification (Fig. 3). There was evidence of intramedullary hemorrhage, with blood pigment in different stages of degeneration.

Discussion

Vascular malformations of the brain stem are characterized by a spectrum of pathological and clinical findings. They include capillary telangiectasias, cavernous hemangiomas, and AVM's. They may produce no neurological deficits or catastrophic hemorrhages. Cavernous hemangiomas and AVM's produce symptoms more commonly than do telangiectasias, and are known to cause progressive or intermittent clinical courses lasting longer than 1 year. In a review of brain-stem vascular malformations producing symptoms for more than 1 year, Stahl, et al., found that two of the 21 patients were less than 12 years old. Those children, aged 3 and 7 years, both had cavernous hemangiomas. Our patient's intermittent signs and fluctuating course were similar to the cases reviewed by Stahl. Relapses and remissions have been reported by Sarkari and Bickerstaff in three cases of brain-stem tumors and one of a para-stem vascular tumor.

Nearly all vascular malformations are evident on good-quality cerebral angiograms. However, Shuey, et al., reported three adults with angiographically cryptic vascular malformations that caused neurological signs for 0.5 to 4 years. All had an avascular mass evident on angiography, and CT scans that showed hyperdense areas enhanced by contrast administration.

Our patient was thought for several months to have brain-stem encephalitis. She had become symptomatic after DPT immunizations at 4 and 19 months, but had seemed to improve after steroid administration, and no neoplasm was evident on biopsy specimens. The enlargement of the pons seen on pneumoencephalography has been reported with brain-stem encephalitis. It has subsequently been our experience that regions affected by brain-stem encephalitis are usually hypodense on CT scans.

Although the DPT immunization and the development of this patient's symptoms were temporally related at both 4 and 19 months of age, the causal relationship is uncertain. One of the three patients reported by Stahl, et al., developed symptoms 2 weeks after immunization for smallpox, tetanus, and typhus, and was considered to have postvaccinal
encephalomyelitis. Although the autopsy on our patient did not reveal a hypersensitivity reaction, even if there had been one, such reactions usually resolve within a few weeks after antigen administration.

The pontine biopsy was performed in the basis pontis because that area appeared on the CT scan to be affected, and because the biopsy of pontine tumors through the floor of the fourth ventricle has been complicated by cranial nerve deficits. Reigel, et al., have emphasized the need for biopsy of pontine lesions prior to radiotherapy or chemotherapy. They showed that pontine astrocytomas are either low-grade and amenable to radiotherapy, or malignant and refractory to radiochemotherapy. Our experience with this patient emphasizes the risk of radiotherapy without histological confirmation of neoplasia. She received a limited course of radiotherapy in spite of a negative pontine biopsy because a CT scan showed apparent enlargement of her mass. She was not helped by radiotherapy.

References