Unusual Radiologic and Clinical Presentations of Posterior Fossa Venous Angiomas

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Venous angiomas are rare congenital vascular malformations of the brain that differ from the more common arteriovenous malformation by the absence of abnormal arterial structures [1]. They are seen most often in the cerebral hemispheres, in the distributions of the anterior and middle cerebral arteries. Less often, they are found in the cerebellum near the vein of Galen [2]. They are often an asymptomatic finding in a patient being investigated by cerebral angiography, but they may become symptomatic due to associated hemorrhage or seizure [3, 4]. The largest autopsy series [4] lists four posterior fossa venous angiomas of a total of nine cases. These were all located in the cerebellar hemispheres in patients who had acute neurologic symptoms secondary to hemorrhage. Our two cases are unusual in several respects. Both patients had longstanding clinical findings localized to the posterior fossa. In both the only abnormality found was a venous angioma. There was no clinical evidence of either recent or old hemorrhage. To our knowledge, this has not been reported before. The location of one venous angioma in the brain stem is also unusual and has not been documented before.

Case Reports

Case

A 34-year-old right-handed man had a 4 year history of sharp. right-sided facial pain. This pain could be triggered by touching the upper lip, brushing the teeth, or eating, and was located in the distribution of the second division of the trigeminal nerve. The pain was believed to represent tic douloureux, and mild relief was obtained with Tegretol. A right-sided intraorbital neurectomy had been performed 1 year before with transient pain relief. Physical examination revealed hypalgesia and hypaesthesia in the right maxillary nerve distribution, related to the previous surgery. The patient was admitted for a Janetta procedure [5] because of his severe pain, and a contrast computed tomographic (CT) scan was obtained preoperatively. This revealed a linear density in the right cerebellopontine angle (figs. 1A and 1B), which was thought to represent a vascular structure; therefore, a right vertebral arteriogram was obtained, which demonstrated a normal arterial phase (fig. 1C). In the venous phase, a network of small veins were seen in the brachium pontis and dentate region draining into several larger veins (figs. 1D–1F). There was no evidence of capillary blush, early venous filling, mass effect, or other vascular abnormality. The appearance was typical for venous angioma. At surgery, two large veins were encountered in the right cerebellopontine angle, one of which was an enlarged petrosal vein compressing the trigeminal nerve. It was coagulated and divided. A second large vein was touching the nerve but not compressing it. A Dacron patch was placed between the nerve and vein. Postoperatively and on discharge, the patient had no facial pain. He was still painfree 6 months after surgery.

Case 2

A 36-year-old woman had gradual onset and progression of unstable gait and staggering over a 6 year period. In the several months before admission, she noted some dysphagia for liquids and solids, with choking and coughing when she swallowed. Neurologic examination revealed several cranial nerve abnormalities. The patient demonstrated mild nystagmus on lateral and upward gaze bilaterally, and there was a slight left upper motor neuron weakness of the facial nerve. The gag reflex was mildly decreased but swallowing was very abnormal with exaggerated pharyngeal motion. Palatal myoclonus was present with a tremulous voice. The general sensory examination was normal, as was motor function with respect to power, tone, and bulk. Cerebellar function was abnormal, with decreased coordination on finger to nose testing and mild left dysdiadochokinesis. The left heel to shin test was slightly abnormal and there was marked gait ataxia. The clinical diagnosis was olivopontocerebellar degeneration. A subsequent CT scan demonstrated several small irregular densities in the brain stem (fig. 2A) that showed contrast enhancement (figs. 2B and 2C), but no mass effect. This was thought to represent a brain stem tumor, and angiography was performed. The arterial phases of bilateral vertebral arteriograms were normal (fig. 2D). In the venous phase, a cluster of small veins at the pontomedullary junction were seen to empty into a large, midline draining vein (fig. 2E). The circulation time was normal, and there was no mass effect or other vascular abnormality. The appearance was that of a venous angioma involving the medulla, pons, and midbrain. A dynamic CT scan was then obtained (fig. 2F), which confirmed the purely vascular nature of the lesion. Due to the location of the venous angioma, surgery and radiotherapy were not indicated, and the patient was discharged with no treatment. No reports of her clinical status have been available since discharge.

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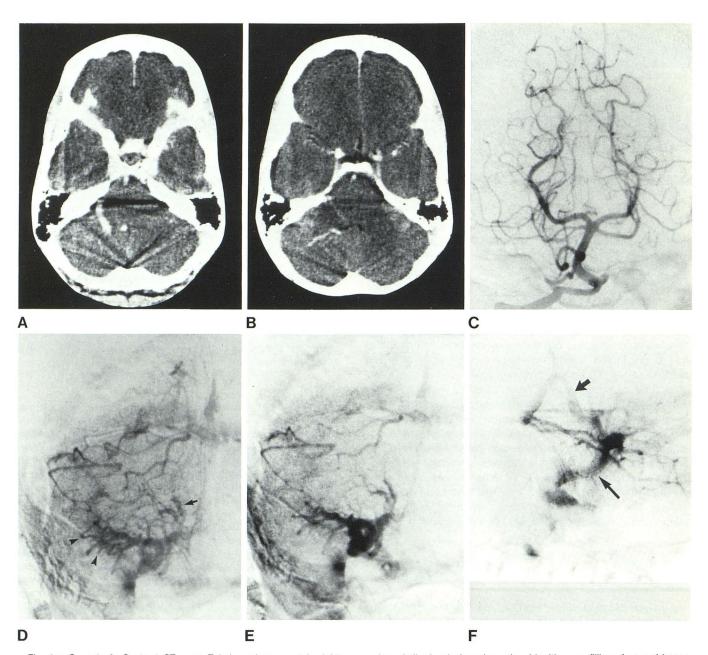


Fig. 1.—Case 1. A, Contrast CT scan. Tubular enhancement in right cerebellopontine angle extends into brachium pontis. B, Linear density extends into right cerebellar hemisphere, which was subsequently proven to be enlarged draining vein. C, Right vertebral arteriogram, anteroposterior (AP) projection. Normal arterial phase. D, Right vertebral arteriogram, AP projection. Early venous phase. Numerous small veins in brachium pontis (arrow)

and cerebellar hemisphere (arrowheads) with some filling of several larger draining veins in cerebellopontine angle. E, Right vertebral arteriogram, AP projection. Later venous phase. Two anomalously enlarged veins now well seen. F, Right vertebral arteriogram, lateral projection, venous phase. One large vein (short arrow) drains into superior petrosal sinus and another (long arrow) drains inferiorly into jugular bulb.

Discussion

Venous angiomas are the rarest of the four basic types of brain vascular malformations [1]. Pathologically, they are characterized by large numbers of irregularly dilated venous channels, the walls of which may show connective tissue thickening but no elastic tissue. Intervening neural tissue is normal [3]. Angiographically, a myriad of small veins are typically arranged in a radial fashion around a large vein that drains superficially. The arterial phase is usually normal

with no arteriovenous shunting or capillary blush [3]. No mass effect is present unless there is associated hematoma.

The lesion is known to become symptomatic due to hemorrhage or seizures. Wendling et al. [3] described two patients with intracerebral venous angiomas who had 1 and 4 year histories of seizures. The lesions were unassociated with significant hemorrhage. Fierstien et al. [6] reported seven cases of supratentorial venous angioma, all of which originally had sudden onset of neurologic symptoms. Wolf et al. [4] collected nine autopsy-proven cases from the

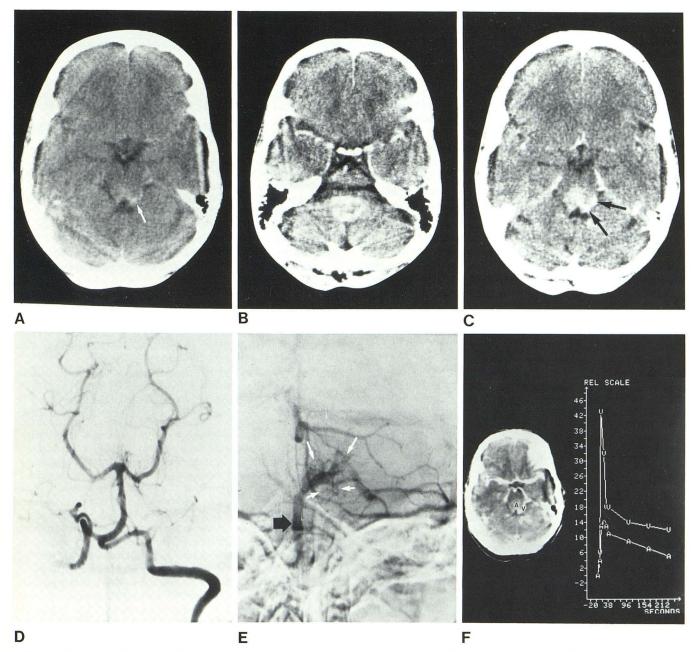


Fig. 2.—Case 2. A, Noncontrast CT scan. Small punctate densities in upper pons (*arrow*) and midbrain extend into lower pons and medulla on other slices. B, Contrast CT scan. Diffuse enhancement in medulla. C, Contrast scan. Diffuse central enhancement within pons and linear density posterolaterally (*arrows*) was subsequently shown to represent draining vein. D, Left vertebral arteriogram, AP projection, normal arterial phase. E, Venous phase. Network of small veins (*white arrows*) near pontomedullary junction

empty into abnormally enlarged posterior medullary vein (black arrow). F, Dynamic CT scan. Cursor A was placed over diffuse central brain stem enhancement and cursor V was placed over known draining vein, as shown at angiography. Density vs. time tracings of two structures are in graph. Density peaks and drainage phases are simultaneous and congruent, indicating absence of extravascular component within central brain stem lesion.

literature and added one case of their own. Six cases of intracerebral and four cases of posterior fossa venous angioma were reviewed. Each of these cases presented acutely over several hours or days with neurologic deficits. In one patient with an intracerebral lesion, transient headaches and vomiting had occurred 3 years before. In one patient with a cerebellar lesion, headaches, vomiting, and vertigo had occurred transiently about 6 years prior to the

final episode. At postmortem, these two cases showed evidence of significant prior hemorrhage. Saito and Kobayashi [7] described three cases of venous angioma in the cerebellar hemispheres. Two of these patients had been symptomatic for over 1 year. In both of these cases, alternate diagnoses were subsequently found to explain the neurologic findings. Michels et al. [8] reported two cases of posterior fossa venous angioma, one of which was located

in the right cerebellopontine angle. Both of these patients presented acutely with symptoms that were attributed to a variant of Guillain-Barré syndrome in one case and postconcussion-syndrome in the other. The venous angiomas were believed to be incidental findings.

Our two cases are unusual in that posterior fossa neurologic symptoms had been present for several years, with no clinical or radiologic evidence of significant associated hemorrhage. In case 1, symptoms had been present for 4 years and were likely related to mechanical pressure on the trigeminal nerve by anomalously enlarged veins. In case 2, brain stem symptoms were present for 6 years with no evidence of significant hemorrhage. The cause of the clinical findings in this case is uncertain, but may relate to altered regional hemodynamics, microhemorrhage, or some type of degenerative disorder unrelated to the venous angioma.

These cases also demonstrate unusual locations of venous angiomas. To our knowledge, these lesions have not been previously reported within the brain stem and are rare in the brachium pontis region. Case 2 illustrates the continuing need to perform angiography, even when a lesion is well demonstrated on CT. The lesion looks very much like an infiltrating brain stem tumor, and may have been irradiated if angiography had not been performed. This case also demonstrates an interesting application of the dynamic CT scan to differentiate between parenchymal and purely vascular lesions.

Venous angiomas are rare lesions, which are often asymptomatic. However, they may be the cause of unusual neu-

rologic symptoms in the posterior fossa, as shown in these two case reports.

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