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Intractable Hiccups and a Posterior Fossa Arteriovenous Malformation: A Case Report

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A 29-year-old man is presented who developed intractable hiccups following a subarachnoid hemorrhage from a posterior fossa dural arteriovenous malformation (AVM). The hiccups persisted even after various treatments had been attempted, and the AVM had been excised surgically. The hiccups stopped after a bilateral phrenic nerve section was performed. This report reviews the central causes and mechanisms of hiccups and emphasizes the importance of selective vertebral angiography in demonstrating posterior fossa AVM.

A hiccup is a clonic spasm of the diaphragm which results in a sudden inspiration that ends abruptly with an audible closure of the glottis. While hiccups are a universal human phenomenon, persistent hiccups are rare and are more likely to be caused by central nervous system lesions. Numerous diseases involving the brainstem have been associated with intractable hiccups, including thrombosis of the posterior inferior cerebellar and vertebral arteries (1), multiple sclerosis (2), intramedullary glioma (3), and medullary ependymoma (4).

In our case, intractable hiccups began after the patient suffered an acute subarachnoid hemorrhage from a previously silent posterior fossa dural arteriovenous malformation (AVM). A survey of the literature indicates that this case may be unique.

Case Report

A 29-year-old man was admitted to Henry Ford Hospital with the chief complaint of persistent hiccups of six months' duration. These began two days after the acute onset of severe headache, paresthesias and weakness over the entire right side. At that time, he was admitted to another hospital, where a lumbar puncture was consistent with subarachnoid hemorrhage. Bilateral carotid and vertebral angiograms were interpreted as normal. For the first two months the hiccups were nearly continuous, except for a few hours of remission. During the next four months, the remissions became longer and more frequent. Although somewhat variable, his condition consisted of four-to-six day periods of continuous hiccups interspersed by two-to-three day intervals without hiccups. Treatment with chlorpromazine, carbamazepine, and baclofen had been ineffectual.

When he was admitted to our hospital, his physical examination was essentially normal. Routine laboratory studies revealed a hematocrit of 26.9, and the stool guaiac was strongly positive. Subsequent gastroscopy showed esophagitis and erosive gastritis. Upper GI, scan of liver and spleen, abdominal ultrasound, and an oral cholecystogram were all within normal limits.

Bilateral carotid and left vertebral angiograms were normal. Computed tomography (CT) with double dose contrast infusion showed an enhanced density on the right side of the lower brainstem just above the level of the foramen magnum. When the right vertebral artery was injected, a large AVM near the foramen magnum on the right side of the brainstem was revealed (Fig. 1). Multiple arterial feeders arose from the right vertebral artery proximal to the origin...
of the right posterior inferior cerebellar artery. Selective injection of the right external carotid artery identified the lesion as dural (Fig. 2).

Three weeks after admission, a suboccipital craniectomy was performed. A large AVM was seen involving the dura over the right anterolateral aspect of the medulla extending from the cervicomedullary junction to the level of the jugular foramen. After surgical resection, it was noticed that the right side of the medulla was indented. Postoperative CT with double dose contrast together with selective right external carotid artery and right vertebral artery angiograms indicated that the AVM had been completely removed. Two days postoperatively, hiccups began again and followed the same pattern as before. The patient's recovery was otherwise uneventful. He was followed as an outpatient for three months but was then readmitted when the hiccups failed to improve.

Symptomatic treatment was begun with pharyngeal stimulation using a nasogastric tube, and chlorpromazine, haloperidol, and diphenylhydantoin were tried in dosages suggested by Williamson, et al (6). When these treatments were ineffectual in controlling the hiccups, bilateral phrenic nerve section was then considered. Preoperative evaluation included chest fluoroscopy and pulmonary function tests. Left and right phrenic nerve block by injection of a local anesthetic were performed with temporary obliteration of the hiccups. Bilateral phrenic nerve sections were then carried out. Postoperative fluoroscopy showed that the right diaphragm was free of hiccups, while the left side was still affected. Re-exploration revealed an accessory left phrenic nerve, which was then sectioned. Fluoroscopy at this time showed a bilaterally quiescent diaphragm. Pulmonary function tests were repeated and were still within a normal range.

The patient has remained free of hiccups for 10 months and has experienced no subjectively appreciable loss of respiratory function.

**Discussion**

Ordinary hiccups, as described by Charles Mayo in 1932 (7), nearly always have an obvious stimulus, which is usually gustatory. Hiccups are thus characterized as a reflex mechanism (Fig. 3). It is generally accepted that sensory fibers of the vagus and phrenic nerves comprise the afferent limb, while motor fibers of the phrenic nerve make up the efferent limb (8).

The central connections which mediate hiccups are less well defined. The functional association between the phrenic nerve nuclei and the respiratory centers, as well as the inspiratory component of hiccups, leads to the postulate...
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that a center for hiccups exists in close proximity to the respiratory nuclei. However, Davis (9) in an electrophysiological study concluded that "hiccup is served by a supraspinal mechanism largely distinct from that generating rhythmic breathing, and that the principal site of interaction of the hiccup discharge with other descending drives to the respiratory motoneuron is at the spinal level."

Our case involved a subarachnoid hemorrhage from an AVM supplied by branches from the right vertebral artery proximal to the origin of the posterior inferior cerebellar artery and from the right external carotid artery. This is the only case in the literature of a posterior fossa AVM and intractable hiccups. The AVM produced an indentation along the right anterolateral aspect of the brainstem just superior to the cervicomedullary junction. It is, therefore, reasonable to assume that neuronal damage might have occurred in the region of the respiratory nuclei.

Several cases have been reported in the literature of intractable hiccups associated with brainstem lesions. In 1923, Rosenow (1) described a case of persistent hiccups associated with thrombosis of the posterior inferior cerebellar and vertebral arteries. Autopsy revealed lesions at the level of the nucleus of the vagus nerve and in the region of the olive. McFarling, et al (2) reported three cases of multiple sclerosis associated with intermittent hiccups that spanned seven days in two of the cases and several years in the third. Kozik and Owsianowska (4) discussed the case of a medullary ependymoma with involvement from C-1 to the pontine-medullary junction. Hiccups were the presenting complaint and persisted intermittently until the patient died six weeks later. Stotka, et al (3) presented a case of an intramedullary glioma found at operation to be in the midline of the lower one half of the medulla and extending into the right inferior cerebellar peduncle. Hiccups were again the presenting complaint. In these cases, as well as in ours, the hiccups were intermittent, with the cycle usually being measured in days.

Bilateral section of the phrenic nerves is a well-recognized treatment for intractable hiccups (6), and in our case the patient has been free of hiccups for 10 months. Campbell (1) cautions, however, about the danger of bilateral phrenectomy in the elderly because of possible ensuing dyspnea and chronic congestion of the base of the lungs due to the coughing difficulties. In these cases, pulmonary function should be carefully assessed before surgery is performed.

References