Spontaneous extracranial vertebral arteriovenous fistula with fibromuscular dysplasia

Case report

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A case of spontaneous vertebral arteriovenous fistula in association with fibromuscular dysplasia is reported. The patient presented with progressive cervical myelopathy and cervical bruit. The pathogenesis of the fistula development and the spinal cord symptoms is discussed. Symptoms subsided after obliteration of the fistula.

KEY WORDS • vertebral artery • arteriovenous fistula • fibromuscular dysplasia • spinal cord

Arteriovenous (AV) fistulas of the extracranial vertebral artery are uncommon. Until 1977, only 81 cases had been reported in the literature. Most of the reported cases developed secondary to trauma, either accidental or iatrogenic, and a few cases were congenital. Most of the iatrogenic cases were the result of percutaneous vertebral or carotid angiography, but occasionally fistulas developed following accidental trauma to the vertebral artery during cervical spine surgery. Very few cases of spontaneous vertebral AV fistulas have been reported, and in most of these reports the etiological factors leading to the formation of the fistulas were uncertain. One of the patients reported had neurofibriomatosis.

Fibromuscular dysplasia (FMD) of the extracranial vertebral artery is rare. To our knowledge, there has been only one previous case of a vertebral AV fistula in association with FMD of the extracranial vertebral artery, but that patient had a previous history of trauma to the neck, and therefore the etiological relationship of FMD to the fistula was only presumptive. We are reporting a case of a spontaneous vertebral AV fistula in a patient with histologically confirmed FMD who presented with symptomatology of progressing cervical cord dysfunction.

Case Report

In the spring of 1979, this 65-year-old hypertensive woman awoke one morning with paresthesia of the left hand. This dysfunction gradually progressed over the next few days to involve the entire left upper extremity. Within 2 months the paresthesia involved the entire left upper extremity and both feet. She also noticed a burning sensation in both hands, arms, both shoulders, the anterior portion of the chest, and the upper part of the back. She experienced stiffness of both hands, worse on the left than the right, and mild weakness of all the extremities, the left side worse than the right. The weakness was more conspicuous after physical exertion. Her gait became progressively worse, with a tendency to lean to the left while walking. She had no sphincter dysfunction.

In September, 1979, she developed some suboccipital and upper cervical pain. She was investigated elsewhere and was treated for “arthritis.” Her condition continued to deteriorate and she was hospitalized.

FIG. 1. Sequential angiograms, lateral view, of the right carotid and the vertebral arteries demonstrating extensive fibromuscular dysplasia of the vertebral artery. A large vertebral arteriovenous fistula located at the C-2 vertebral level, and rapid opacification of the cervical venous plexus, can be seen. The right carotid artery shows no evidence of fibromuscular dysplasia.

An extensive evaluation included electromyography, brain scan, and electroencephalography. A bruit developed in the suboccipital region in April, 1980, and the patient was referred to the Mayo Clinic.

Examination. Neurological examination at the time of admission revealed spasticity and mild weakness of the left upper extremity and mild spasticity of both lower extremities. There was bilateral hyperreflexia, greater on the left than the right, with bilateral ankle clonus and an extensor plantar response on the left. The gait was spastic. There was some decrease in two-point discrimination and vibratory sense in both hands, but greater on the left. A loud continuous bruit was heard over the suboccipital region and over the upper cervical spine, with the maximum intensity toward the right side.

The initial clinical impression was an AV malformation or a highly vascular neoplasm. X-ray films of the skull and cervical spine revealed basilar invagination. An electrocardiogram revealed sinus tachycardia and marked left axis deviation. Retrograde femoral angiography of both the vertebral arteries revealed changes typical of FMD affecting both vertebral arteries (Figs. 1 and 3). There was a fistulous communication between the right vertebral artery and the epidural venous plexus at the level of the C-2 vertebra (Figs. 1 and 2). The left vertebral injection revealed no evidence of steal to the right, which suggested that the right vertebral artery distal to the fistulous site was probably occluded (Fig. 3). A small aneurysm was found incidentally, involving the pre-cavernous segment of the left internal carotid artery.

Operation. On April 4, 1980, through a transverse supraclavicular incision, the right vertebral artery was exposed after the scalenus anticus muscle and suprapleural membrane had been divided. The distal right vertebral artery was then exposed through a curvilinear incision at the base of the occiput. There was profuse venous bleeding in this area, and it was difficult to separate the right vertebral artery from the surrounding plexus of dilated venous channels. Ultimately, the vertebral artery was identified between the arch of the atlas and the occiput, and was ligated and divided. A segment of the artery was excised for histological confirmation of the diagnosis. There was no back bleeding from this vessel, presumably because of distal thrombosis, and this accounted for the absence of angiographic evidence of steal. The proximal vertebral artery was then doubly ligated at the level of the C-6 transverse process, and divided distal to the ligatures. There was a small amount of back bleeding from the vessel. This was thought to be coming from the muscular collateral vessels, which presumably anastomosed with the distal vertebral artery. In order to promote intraluminal thrombosis, a Portnoy catheter was passed distally into the distal vertebral artery beyond the site of the fistula, and was ligated in place.
Spontaneous vertebral arteriovenous fistula

FIG. 3. Angiography, anteroposterior and lateral views, of the left vertebral artery showing typical changes of fibromuscular dysplasia involving the entire extracranial segment of this vessel.

Postoperative Course. The patient had an uneventful recovery. The bruit, the spasticity, and the burning sensation in the extremities subsided, and her gait improved markedly. Sensation in the left upper extremity increased noticeably, although she had minimal residual paresthesia of the left hand at the time of discharge. The patient refused postoperative angiography. Histological examination of the biopsy specimen of the vertebral artery confirmed the angiographic diagnosis of fibromuscular dysplasia.

Discussion

Fibromuscular dysplasia (FMD) of a cervical vertebral artery is rare, but can affect any part of the vertebral artery. The etiology is not known, although the female predilection of this disease suggests the possibility of a hormonal factor. Most cases of extracranial vertebral artery FMD are asymptomatic, but symptoms of vertebrobasilar insufficiency have been reported. Aneurysms and intramural dissections of extracranial vertebral arteries have been observed in association with FMD, which suggests that rupture of such an aneurysm into the adjacent venous plexus was the most likely cause for the fistula reported here. It is interesting to note that our patient also had an intracranial saccular aneurysm of the internal carotid artery.

The clinical features of a vertebral AV fistula depend upon the location and size of the fistula. In children, some cases have been discovered as an incidental finding in an otherwise asymptomatic patient with a cervical bruit. Although spontaneous closure has been reported occasionally, most of these fistulas tend to increase in size as extensive collateral channels develop. Cardiac enlargement and cardiac failure are uncommon but have been reported. Many adult patients present with incapacitating tinnitus, occipital headaches, and a progressing neurological deficit. Fistulas located in the suboccipital region may cause reversal of blood flow in the distal vertebral artery with development of symptoms of vertebrobasilar steal.

The cause of cervical cord dysfunction in these cases is intriguing. Compression of the markedly engorged epidural venous plexus and a steal phenomenon due to shunting of arterial blood with resultant local cord ischemia may both play a role in the development of the spinal cord symptoms.

The treatment of the vertebral AV fistulas is complicated by the relatively inaccessible location of the vertebral artery in the transverse foramina, and by the presence of multiple secondary arterial feeders from the muscular branches of thyrocervical trunk and occipital arteries. Simple proximal ligation of the vertebral artery alone is ineffective because of these anastomotic channels along the course of the vertebral artery and retrograde flow through the distal vertebral artery.

Although direct obliteration of a fistula with proximal and distal ligation has been reported, this procedure is hazardous and sometimes may result in major intraoperative hemorrhage. Good results have been achieved with proximal and distal ligation and intra-arterial balloon occlusion of the fistula. In the case presented here, the vertebral artery was ligated proximally and distally, and a Portnoy catheter was placed in the second part of the vertebral artery to act as a nidus for thrombosis and thus prevent the development of collateral flow along the course of the vertebral artery.

References


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