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Spine

Spontaneous spinal epidural arteriovenous fistulae in neurofibromatosis type-1

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Abstract

Background: NF-1 is one of the most common autosomal-dominantly inherited genetic disorders with an incidence of approximately 1:3500. We report a case and review the literature to characterize spontaneous spinal AVF that occur in neurofibromatosis (NF-1).

Case Report: A 51-year-old woman presented with NF-1 and progressive radiculomyelopathy. Angiography revealed an AVF terminating in a giant intraspinal epidural varix extending paraspinally through the C3/4 neural foramen. Trapping of the AVF attempted 18 years earlier prevented endovascular access for embolization, and vigorous bleeding made direct surgical resection impossible. Therefore, as palliation, arterial feeding collaterals were occluded, and surgically exposed tortuous veins were packed with coils. Laminectomies and partial resection of the epidural varix resulted in subtotal occlusion with clinical improvement.

Conclusion: The spinal AVF associated with NF-1 appears to show dominant venous drainage to the intraspinal extradural and paraspinal venous plexus without evidence of intradural drainage. The vertebral artery is typically the origin of the fistula. A giant venous varix and numerous collateral feeders to the vertebral artery may give an AVM-like appearance. Clinically, the fistulae produce a syndromic triad including symptoms of NF-1, progressive radiculomyelopathy, and a bruit. Treatment is direct attack on the fistula by either surgery or embolization. If, however, a direct approach cannot be chosen, occlusion of feeding vessels combined with laminectomies can result in long-term symptomatic improvement.

Keywords: Spinal arteriovenous malformation; Epidural fistula; Vertebral artery; Neurofibromatosis

1. Introduction

NF-1 is one of the most common autosomal-dominantly inherited genetic disorders with an incidence of approximately 1:3500 [67]. The genetic mutation localized to the long arm of chromosome 17 causes a dysplastic disorder also affecting the vasculature [6,51,60,67]. Patients with NF-1 may present with a spectrum of vascular lesions ranging from occlusive disease to those of acute hemorrhage [1,13,14,43,46,69,71]. The pathognomonic cutaneous neurofibroma itself is associated with an abnormal vascular supply that may cause pronounced bleeding on resection [34]. Vascular lesions affecting the central nervous system of patients with NF-1 have been described as occlusion or hypoplasia of intracranial arteries [5,14,46,56], moyamoya vessels [3,12,26,32,46,56], and aneurysms [3,19,40,43,58,59]. However, little is known about the spontaneous spinal AVF associated with NF-1. We report a case and review the literature to characterize this complex type of fistula.

2. Case report

2.1. History and examination

This 51-year-old lady presented with intractable pain in her left rib cage, difficulty in walking, weakness/numbness in her left hand, and a pulsatile left-sided tinnitus. Eighteen years earlier, she underwent attempted trapping of a cervical AVF involving the vertebral arteries. Examination at the

Abbreviations: AVF, arteriovenous fistula; AVM, arteriovenous malformation; CT, computed tomography; NF-1, neurofibromatosis type-1; SSEP, somatosensory-evoked potentials; MRI, magnetic resonance imaging.

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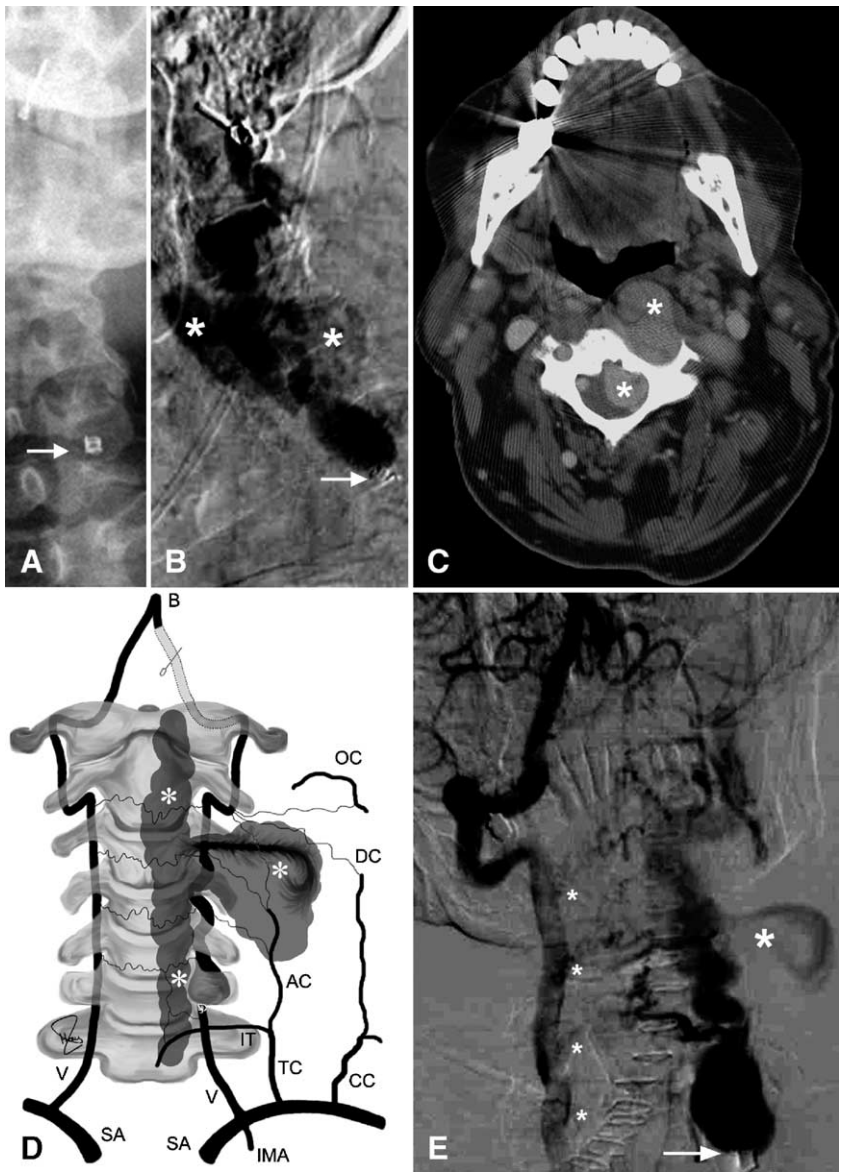

Fig. 1. Imaging of a spontaneous vertebral AVF in NF-1. A: Ap plain x-ray demonstrating a clip placed at the foramen magnum level and a coil package at the C6/7 neural foramen (white arrow) in an attempt to trap a left vertebral artery (VA) fistula 18 years earlier. B: Digital subtraction angiogram, lateral view, shows maintained flow in the isolated segment of the left VA. Note the tortuous course of the VA ending in an aneurysmatic dilatation proximally just above the coil package (white arrow). The asterisks (*) mark parts of the intra- and paraspinal varix. C: Contrast-enhanced CT of the C-spine at C3, the level of the fistula. Note bone scalloping at the left foramen transversarium and the large epidural intra- and paraspinal expansion of the varix (*). Also note the mass effect with shift of the cord to the right. D: Artistic sketch summarizing the findings of superselective angiography and contrast-enhanced CT. The arteries are abbreviated as AC, ascending cervical; B, basilar; CC, costocervical trunk; DC, deep cervical; IMA, internal mammary; IT, inferior thyroid; OC, occipital; SA, subclavian; TC, thyrocervical trunk; and V, vertebral. Clip placement at the distal VA resulted in thrombus formation proximal and distal to the clip. The coil package occludes the left VA at C6/7. Several collaterals maintain flow in the "trapped" segment of the left VA and give the appearance of an AVM. However, the VA is the only direct feeder to the AVF. The varix extends intra- and paraspinally (*). E: Ap view, digital subtraction angiogram. Maintained flow in the trapped left VA segment with partial thrombosis distally. Note the jet stream into the fistula at C3/4 (large asterisk). The vertebro-vertebral collaterals (small asterisk) indicate the spinal levels.

current admission showed disseminated cutaneous neurofibromata, café-au-lait spots, a Lisch nodule, a palpable thrill, and a machine-like neck bruit at the left side of her neck. Neurological findings included left C4 to C7 radiculopathy, marked gait unsteadiness, and hyperalgesia in the thorax. Plain x-ray (Fig. 1) showed a clip placement at the foramen magnum level and a coil package next to the C6/7 neural foramen consistent with the earlier trapping of the intervening left vertebral artery. However, angiography (Fig. 1) demonstrated maintained blood flow in the "trapped" left vertebral artery through multiple engorged, tortuous collaterals including branches of the thyrocervical and costocervical trunk, the contralateral vertebral, ipsilateral subclavian, occipital, ascending cervical, and deep