With the ever-increasing challenges, professional and otherwise, a busy schedule, and highly specialized practice of a clinician, diagnosis of certain conditions can be delayed. Among the vascular malformations of the spine, spinal dural arteriovenous fistula (SDAVF) is the most common vascular anomaly, constituting approximately 70% of all vascular anomalies.¹

SDAVF is presumed to be an acquired disease, with its exact cause remaining elusive. It commonly occurs in middle-aged men, predominantly occurring between the D4 and L2 vertebrae. Cervical dural arteriovenous fistula (dAVF) constitutes an extremely small minority and therein lies the significance of the case highlighted by Prof. Thakur et al along with the clinical image of the involvement of the hypoglossal nerve.

Anatomically, an SDAVF is characterized by an abnormal communication between a radiculomeningeal branch of a segmental artery and a draining vein. It lies inside the dura and in proximity to the spinal nerve root. This leads to altered venous flow, causing vascular congestion, which further causes hypoxia of the cord and progressive myelopathy. This explains the worsening of symptoms in these patients after exertion as arterial pressure rises.

The onset is insidious, progression is slow, and its symptomatology is vague starting from nonspecific low back pain without radiculopathy and easy fatiguability. The disease generally runs an indolent course but occasionally may have rapid worsening. However, by the time it is diagnosed advanced deficits in the form of bowel and bladder disturbances and erectile dysfunction set in and the patient may be wheelchair bound with simultaneous presence of mixed upper motor neuron and lower motor neuron signs. Prof. Thakur et al, in their case of SDAVF, displayed a rather unique presentation of dAVF with hypoglossal nerve palsy. A detailed angiographic study would have added more insight into its underlying cause and location.

The diagnosis of dAVF is invariably radiological with a magnetic resonance imaging (MRI) of the spine raising suspicion and MR angiography (MRA) guiding the diagnosis. Dilated and congested venous channels with cord edema with or without myelopathic changes are its hallmark (►Fig. 1). The appearance of dilated and serpentine flow voids on T2 sequences, especially dorsal to the cord, represents abnormal perimedullary veins. Heavy T2-weighted sequences like fast imaging employing steady-state acquisition (FIESTA) and constructive interference in steady state (CISS) help further confirm the diagnosis. In addition to the dilated veins, T2 hyperintensities representing cord edema may be present. The location of the cord edema may be far removed from the

Fig. 1 (A) Sagittal constructive interference in steady state (CISS) sequence showing prominent perimedullary veins. (B) T2-weighted sagittal sequence showing edema of the dorsal cord. (C) Postcontrast image showing enhancement of the edematous cord with perimedullary veins. (D) Digital subtraction angiography (DSA) showing spinal dural arteriovenous fistula (dAVF) with feeder from (Rt) D9 radicular artery and dilated perimedullary veins. (E) Postoperative magnetic resonance imaging (MRI) sagittal T2-weighted sequence showing reduction in the perimedullary flow voids. (Adapted from Sharma et al.⁴)
site of the fistula because of anatomical reasons. Contrast administration improves visualization of the perimedullary veins and may enhance the cord as the blood–brain barrier is broken. The gold standard to understand the anatomy of the fistula and to attain the diagnosis, however, remains a superselective angiography. The problem, however, lies in the fact that each segmental artery needs to be separately cannulated and evaluated after injecting contrast and therefore an MRA prior to invasive angiogram helps in reducing the area requiring evaluation with a super-selective angiography.

DAVs are amenable to surgical intervention. The endovascular option, although less invasive, has its limitations in reaching the terminal point away from collaterals and carries the risk of blocking the useful arteries/veins leading to fresh and permanent neurological deficits. The surgical challenge lies in identifying the exact pathological level of fistula and correlation of the intraoperative anatomy with angiography. To prevent collateral formation and surgical failure, the draining vein needs to be ligated closest to the dura. An endovascular intervention involves super-selective cannulation of the feeder artery, negotiating the nidus and reaching the proximal part of the draining vein. Although minimally invasive, it has a success rate of 25 to 75% as against a surgical success rate of 98%.

SDAVF is a potentially treatable condition but often leads to significant morbidity if left untreated (Fig. 1). A correct and timely diagnosis and intervention can invariably prevent further progression of the disease and lead to reversal of symptoms in a significant number of patients.

Conflict of Interest
None declared.

References