



Case Report

Type I AV fistula of the thoracic spinal cord

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ABSTRACT

Type I AV fistulas of the spinal cord are exceedingly rare. The average age at diagnosis is 50. Clinical presentation is often very non-specific, and sensory deficits and sphincter dysfunction may also occur. Neurological deterioration is generally gradual. Thus, failure to diagnosis frequently results in permanent disability.

A 22-year-old female complained of a “muscle spasm” in the midline thoracic area with no history of trauma or prior occurrence. She also experienced bilateral lower extremity weakness/numbness and perianal anesthesia. She is a healthy female with no medical problems. Exam revealed lower extremity motor function of 1/5 as well as diminished sensation. A foley catheter was placed for urinary retention. The remainder of the neurological exam was normal. MRI demonstrated a lesion at the fourth thoracic level with significant cord compression. The patient was taken emergently to the operating room by neurosurgery.

This case demonstrates a rare disorder occurring in a 22-year old female, far younger than the typical 50-year old patient. Moreover, the lesion was located in the thoracic rather than the typical lumbar cord. Symptoms may be misinterpreted as a peripheral nerve lesion and delay time to diagnosis. Early diagnosis remains critical to prevent permanent neurologic sequelae. AV fistula should remain high on the differential of patients presenting with back pain and focal neurologic complaints.

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1. Introduction

Type I AV fistulas of the spinal cord are exceedingly rare. Males are affected 5–6 times more than females. The average age at diagnosis is 50. Less than 1–4% are under age 30 [1,2]. Clinical presentation is often very non-specific, often mimicking DJD, spinal stenosis, peripheral neuropathy, or transverse myelitis [3–5]. Sensory deficits and sphincter dysfunction may also occur [6]. Type I AV fistulas originate dorsally from a radicular artery. Neurological deterioration is gradual; however, sudden deterioration may occur if hemorrhage or thrombosis occurs. This is known as Fiox-Alajenani syndrome [7]. Failure to diagnose frequently results in permanent disability.

2. Case presentation

A 22-year-old white female complained of a “spasm” in the midline interscapular area, bilateral lower extremity weakness and numbness, and perianal anesthesia. She denied any trauma or prior episodes. She is a healthy female with no recent illnesses. She further denied any fever/chills. She has no medical or surgical history and takes no medications. She denies tobacco, alcohol, or recreational drug use. Family

history includes diabetes mellitus. On exam the patient appeared in no acute distress. She had motor strength of 1/5 as well as diminished sensation of both lower extremities. A foley catheter was placed for urinary retention. The remainder of the neurologic exam was normal. The MRI showed a lesion at the T4 level with significant compression of the cord. Typical findings on MRI include cord edema, perimedullary dilated vessels, and cord enhancement [8,9]. The patient was taken emergently to the operating room and recovered uneventfully.

3. Discussion

This case demonstrates a rare disorder occurring in a 22-year-old female, far younger than the typical 50-year-old patient. Moreover, the lesion was located in the thoracic cord rather than the typical lumbar cord. In fact, they are the most common malformation of the spinal cord. There are four types of SDAVF. Type I is located within the dural nerve sheath. Type II occurs within the spinal cord. Type III, occurs both intra and extradurally. Type IV is a perimedullary fistula. The arterial supply typically originates from the anterior spinal artery or posterior artery [10]. Treatment modalities include surgical occlusion of the intradural vein or endovascular therapy [8]. The majority of patients present in the fifth or sixth decade of life, with less than 1% under age 30. Misdiagnosis is frequent, with symptoms often suggesting spinal stenosis, DJD, transverse myelitis, or other pathology. Clinical findings frequently are delayed, resulting in progressive dysfunction. Initial symptoms most commonly include lower extremity weakness and

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sensory disturbance and sphincter dysfunction may be seen also [6]. Symptoms may be misinterpreted as a peripheral nerve lesion [11]. Many cases involved a time to diagnosis of ten months (up to two years) in one series [12]. Early diagnosis remains critical to prevent permanent neurologic sequelae. The pathophysiology is secondary to the common venous arterial flow occurring in the spinal cord. Venous congestion ultimately results in hypoxia with increasing clinical symptoms as the congestion progresses in a caudocranial fashion [12]. Other possible contributing factors include hemorrhage, vascular steal, or mass effect [1]. AV fistula should remain high on the differential of patients presenting with back pain and focal neurologic complaints.

Conflicts of interest

The authors report no conflict of interest.

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