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Case Report

Late diagnosis of spinal dural arteriovenous fistulas resulting in severe lower-extremity weakness: a case series

Ivelin Iovtchev, MD[a], Nurith Hiller, MD[b], Yona Ofran, MD[a], Isabella Schwartz, MD[a], Jose Cohen, MD[c], Stuart A. Rubin, MD[a], Zeev Meiner, MD[a,*]

[a]*Department of Physical Medicine and Rehabilitation, PO Box 24035, Hadassah Hebrew-University Medical Center, Jerusalem, Israel*
[b]*Department of Radiology, PO Box 24035, Hadassah Hebrew-University Medical Center, Jerusalem, Israel*
[c]*Department of Neurosurgery, PO Box 24035, Hadassah Hebrew-University Medical Center, Jerusalem, Israel*

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Abstract

BACKGROUND CONTEXT: Spinal dural arteriovenous fistula (SDAVF) is a slow-flow extramedullary vascular lesion affecting primarily the lower thoracic and lumbar spine. The clinical sequela of these vascular changes is progressive myelopathy and severe lower-extremity weakness. Although surgical or embolic treatment of SDAVFs has improved significantly in the last years, the ambiguity of the symptoms may complicate and delay the diagnosis. The influence of the postponed diagnosis on the functional outcome of patients with SDAVF is unknown.

PURPOSE: To describe a case series of patients with SDAVF that illustrates that delayed diagnosis leads to grave neurologic and functional prognosis.

STUDY DESIGN: A case series.

METHODS: We present a series of seven patients, treated in a tertiary university rehabilitation center over 20 years. Clinical, radiologic, and functional outcomes were evaluated by retrospective chart review. Neurologic and functional evaluation at the end of rehabilitation was evaluated with the lower extremities motor score and the Aminoff-Logue scale, respectively.

RESULTS: All our patients were men with a mean age of 60.3±16 years (30–72 years), mean time until the diagnosis of SDAVF was 302.8±239 days (60–730 days), and mean overall length of stay in acute department and rehabilitation unit was 88.6±34 days (46–149 days). At the end of rehabilitation period, four patients remained at wheelchair level with an Aminoff-Logue scale grading of five whereas other functional scales showed also low levels of recovery.

CONCLUSIONS: Our series showed that the potential for functional ambulation was poor despite prolonged rehabilitation treatment in late diagnosis SDAVF. Awareness of the early symptoms of SDAVF and immediate intervention may help reduce impairment in such patients.

Keywords: Spinal dural arteriovenous fistula; Rehabilitation; Myelopathy; Functional outcomes; Foix-Alajouanine syndrome; Aminoff-Logue scale

Introduction

Spinal dural arteriovenous fistulas (SDAVFs), also known as Foix-Alajouanine syndrome, are slow-flow extramedullary vascular lesions affecting mostly the lower thoracic and lumbar spinal levels [1]. SDAVFs represent approximately 75% to 80% of all spinal vascular malformations, and the majority of affected patients are men older than 50 years of age [2]. Progression to severe myelopathy or paraplegia is slow. Patients initially may present with acute lower-extremity dysesthesias and intermittent radicular pain mimicking peripheral nerve lesions. There may be also bowel or bladder incontinence and impotence. These

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* Corresponding author. Department of Physical Medicine and Rehabilitation, PO Box 24035, Hadassah University Hospital, Jerusalem, 91240 Israel. Tel.: +972 2 5844474; fax: +972 2 5829113.

E-mail address: meiner@hadassah.org.il (Z. Meiner)

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vague symptoms make the diagnosis complicated and delayed [3]. Many of these patients have already suffered from their symptoms for several months and have undergone unnecessary procedures and surgery before diagnosis [4]. The recommended treatment is either endovascular embolization of the fistula or surgical removal [5]. The influence of the postponed diagnosis and therefore delayed interventions on the functional outcome of SDAVF patients is unknown. In this article we describe a case series comprising seven patients treated in a rehabilitation unit in which the diagnosis of SDAVFs was delayed and were left with severe lower extremity weakness.

Case series presentation

In the last 20 years, seven patients with delayed-diagnosis SDAVF were treated in our inpatient rehabilitation unit. Clinical, demographic, radiologic, and functional outcomes for these seven patients are summarized in Table and presented in Figs. 1–3. All were men with mean age of 60.3±16 years, with a range from 30 to 72. The mean time until the diagnosis of SDAVF was 302.8±239 days, with a range from 60 to 730. The overall mean length of stay was 88.6±34 days (46–149 days), including 26.1 ± 9 days (14–36 days) in the acute department and 62.4±29 days (32–113 days) in the rehabilitation unit.

The diagnosis of SDAVF was delayed in most of the patients in our series; requiring an average of 10 months and reaching almost 2 years in one patient. Many alternative diagnoses were proposed such as spinal stenosis, myeloradiculitis, peripheral neuropathy and more. In two of our patients, axonal motor neuropathy was found by nerve conduction studies. Cerebrospinal studies were performed in four of six patients. The main findings were elevated red blood cells in two and high protein levels in three. Positive oligoclonal bands typical for inflammatory conditions were found in two patients. Some of the patients were treated because of these false diagnoses with surgical procedures, epidural injections, intravenous and oral steroids, and plasmapheresis. Three patients were treated by embolization, two patients were treated by surgery alone, and two patients were treated by combined approach. Because of the small sample, no definite conclusion regarding the correlation between type of therapy and prognosis could be obtained.

The neurologic function of patients with SDAVF at the end of rehabilitation were evaluated using the lower extremities motor score according to the International Standards for the Neurological Classification of Spinal Cord Injury [6]. As can be seen in Table, lower extremities motor score remained less than 30 in 5 of 7 patients. This reflected the severe lower extremity weakness of these patients at the end of rehabilitation. The functional level of patients with SDAVF at the end of rehabilitation were evaluated using the Aminoff-Logue scale (ALS) [7]. This scale consists of six grades of gait, between 0 normal and 5 confined to a wheelchair, and 4 grades of micturition between 0 normal and 3 total incontinence or persistent retention. At the end of rehabilitation period, four patients remained at wheelchair level with an ALS grading of 5, and two patients had ALS grading of 4, indicating that they required two crutches, or walker. One patient was able to ambulate with a cane (ALS of 3).

Discussion

In this article we described a case series of seven patients suffering from SDAVF treated in one rehabilitation facility over 20 years. Despite prolonged rehabilitation treatment, four of seven patients remained at wheelchair level and their level of independence was low. The diagnosis of SDAVF was delayed in all 7 patients contributing to the low level of independence at the end of rehabilitation.

SDAVFs are the most prevalent type of spinal vascular lesions and have been classified into four types: type I, SDAVFs located within the dura of the nerve root sleeve, connecting a dural branch with an intradural medullary vein; type II, congenital intramedullary AVM fed by spinal artery branches; type III, juvenile AVM, a very rare lesion comprising an intramedullary nidus with extramedullary/extraspinal extension; and type IV, intradural extramedullary AVF, also known as perimedullary fistula [8].

Foix-Alajouanine syndrome was first described in two young men (ages 31 and 37 years) in 1926 [9]. Until the middle of the 20th century, this pathologic entity was only made at autopsy. With the advent of high-resolution magnetic resonance imaging (MRI) and modern spinal angiography, it became possible to diagnose the syndrome ante-mortem and to offer an effective treatment. However, even today, the true incidence of SDAVF is unknown, and often there is a significant delay in the diagnosis, as was demonstrated in our cases. In a series of 49 patients with SDAVF admitted to one unit over 15 years, the time interval between the initial symptoms and diagnosis was a median of 10.5 months (range, 1 day to 20.8 years) [10] and in another large series of 66 patients, the average time from symptoms onset to diagnosis was reported to be 27 months [4].

Because of the low incidence of SDAVF and its puzzling symptomatology, an appropriate diagnostic workup is often not performed, and the time to diagnosis is prolonged [11]. Most of the patients complain of leg weakness or paraparesis, sensory numbness or paresthesias, urinary or bowel incontinence or retention, and pain. Pain was a concern in approximately half of the patients, including back pain and/or radiculopathy [4]. At the time of diagnosis, patients with SDAVF have signs of both myelopathy and lower motor neuron findings emanating from lower thoracic spinal cord and conus medularis involvement of anterior horn nuclei. In our patients several alternative diagnoses were made before the right diagnosis of SDAVF was reached. These possibilities included benign hypertrophy of the prostate in patients 1 and 4; myeloradiculitis in patients 2 and 5;