

Spinal Amyloidoma Mimic an Intradural Neoplasm. Case report and literature analysis.

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Abstract

Introduction: Amyloidosis represents an alteration secondary to the extracellular deposit of low molecular weight protein with a fibrillar structure. Intradural spinal amyloidoma is unusual, with only two cases previously reported.

Case report: A female 53 years old patient with story of progressive paraparesis and successfully operated of an intradural extramedullary amyloidoma is presented.

Conclusions: Although extremely unusual, the spinal amyloidoma should be considered in the differential diagnosis of any patient with an intradural extramedullary tumor. In this scenario, the laminectomy constitutes a good therapeutic modality which not only afford to stablish the positive diagnosis, also helps to improve the neurological condition of the patient.

Key words: Amyloidosis; spinal amyloidoma; surgical resection

Introduction

Amyloidosis is a disease caused by extracellular deposition of insoluble proteins with a fibrillar structure composed of low molecular weight subunits, most of which are between 5 and 25 Kd. Among the types of amyloidosis, the primary and secondary types are described. In primary amyloidosis, the protein deposit is derived from fragments of immunoglobulin light chains, and is in the group of plasma cell dyscrasias with clonal expansion. The secondary form occurs as a complication of chronic diseases such as rheumatoid arthritis, spongy arthropathies, osteomyelitis and tuberculosis [1].

Among the extramedullary intradural tumor lesions, meningiomas, peripheral nerve sheath tumors and lipomas stand out. However, in certain circumstances they may respond to other non-neoplastic lesions [2]. Within these, intradural spinal amyloidoma is very rare, with only 2 previous cases reported in the literature in the review conducted [3, 4] and within them only one with a localized form ⁴ so the present study constitutes the second international report.

The results of the surgical treatment in a patient with dorsal extramedullary intradural spinal tumor, corresponding to an amyloidoma

in the context of a primary amyloidosis, are presented.

Clinical case

A 53-year-old female patient is presented, of rural origin, white race, with a history of previous good health and a history of progressive decrease in muscle strength in both lower limbs of 3 years of evolution, associated with stiffness.

The neurological examination confirmed a spastic paraparesis with ASIA 3/5 in the lower right limb and 4/5 in the lower left limb, as well as patellar hyperreflexia and bilateral aquilea with increased reflexogenic area and bilateral Babinski sign. Hypoaesthesia was also observed for thermoalgesic sensitivity, apalestesia and abarognosis, with a sensitive level in D4. The rest of the physical exam was negative.

Simple x-rays of the cervical and lumbar spine showed moderate signs of spondylosis without other alterations. In the Magnetic Resonance Imaging (MRI) study, a hypointense posterior intradural lesion was observed in the well-defined sequence in T1 and T2 of well-defined limits that compressed the spinal cord at the level of D4 with a maximum cephalocaudal diameter of 38 mm (image 1A and B).

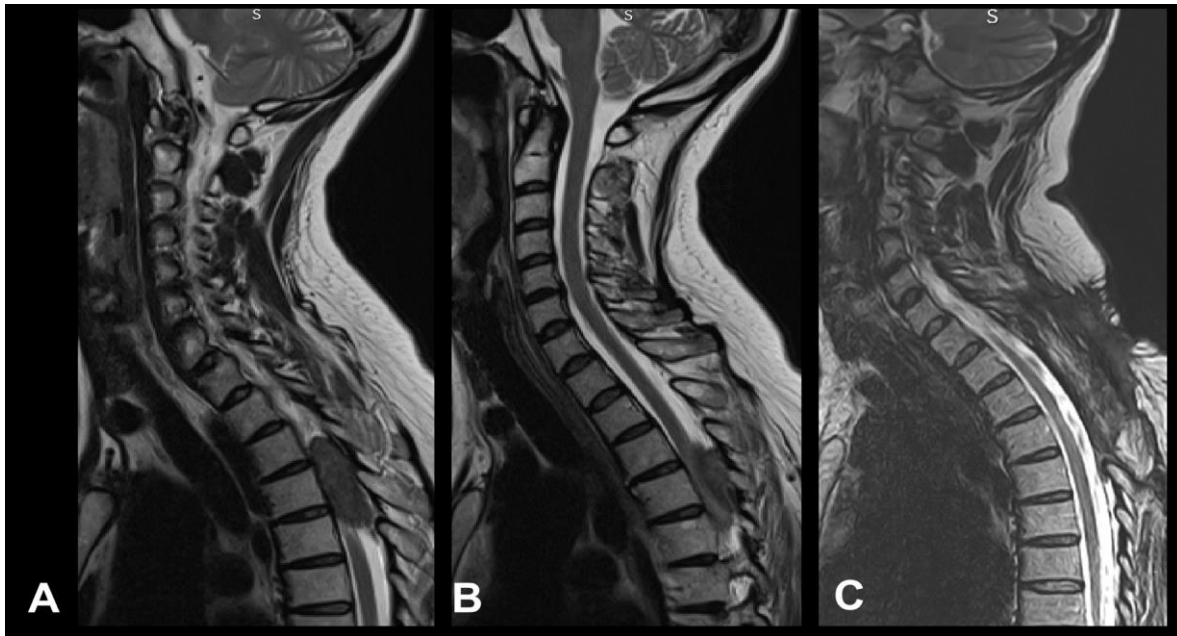


Figure 1: Comparison between sagittal sections of MRI T2 weighted preoperatively (A and B) and postoperative (C) where the total resection hypointense lesion T2 spinal posterior (A and B) is confirmed.

prior asepsis and antisepsis and was performed under general anesthesia orotracheal a posterior approach to the thoracic spine by complete laminectomy D4, and D3 and right interlaminectomy D5 observed dura thickened at that level. After durotomy on the midline point placement dural traction solid nodular lesion, aspect gray-I pink and limits defined well vascularized, on the posterior aspect of the spinal cord was observed.

microsurgical dissection was performed and lesion was resected en bloc. Subsequently, hermetic duroplasty was performed with continuous suture with polyester 4.0 and closure was performed by soft tissue planes (image 2, A and B). The operating time was 3:30 hours and intraoperative bleeding 150 ml. There were no complications during the procedure.

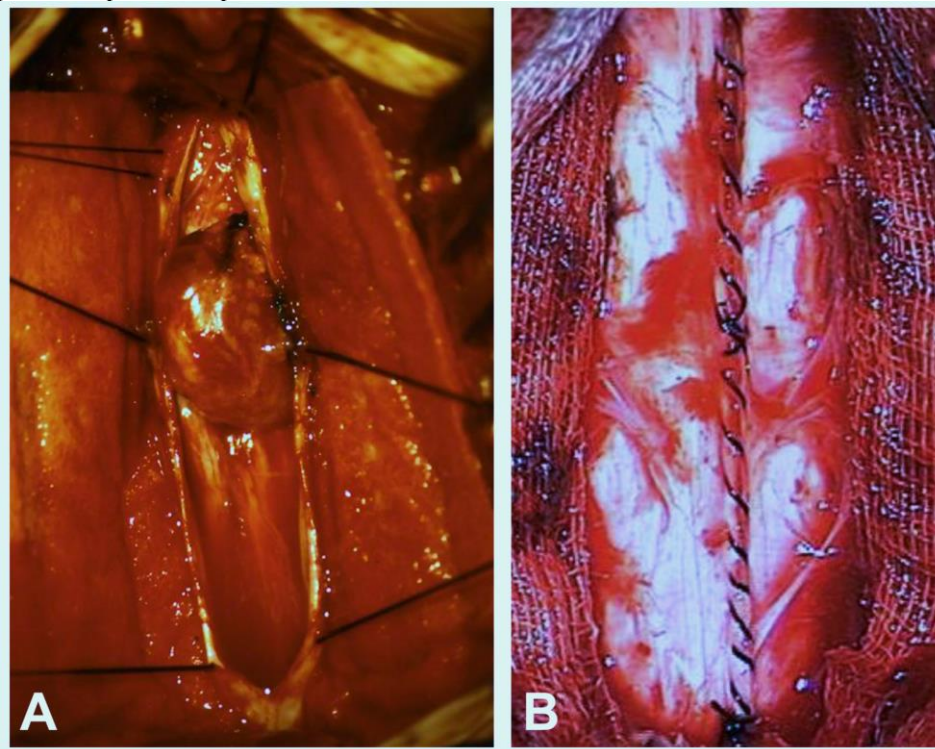


Figure 2: Intraoperative pictures of the case. A: After the laminectomy and durotomy, spinal injury nodular well-defined limits are observed. B: At the conclusion of the tumor hysteresis and perform the durotomy.

The patient had a favorable evolution, discharge 7 days of the proceeding with a complete neurological and functional recovery. Postoperative MRI confirmed total resection of lesion (Figure 1 C).

The pathology report confirmed dorsal spinal amyloidoma (Figure 3 A and B). It was observed in the study with blocks hematoxylin and eosin amorphous cellular substance and positivity was shown to Congo red (characteristic of amyloidosis) and negativity for trichrome Gomori

which rules out the possibility of collagen and elastic fibers, still contributing the diagnosis.

After diagnosis, the patient was studied looking for other locations of the disease through abdominal ultrasound which showed no renal alterations and the echocardiogram which was negative. Renal function on the other hand remained normal.

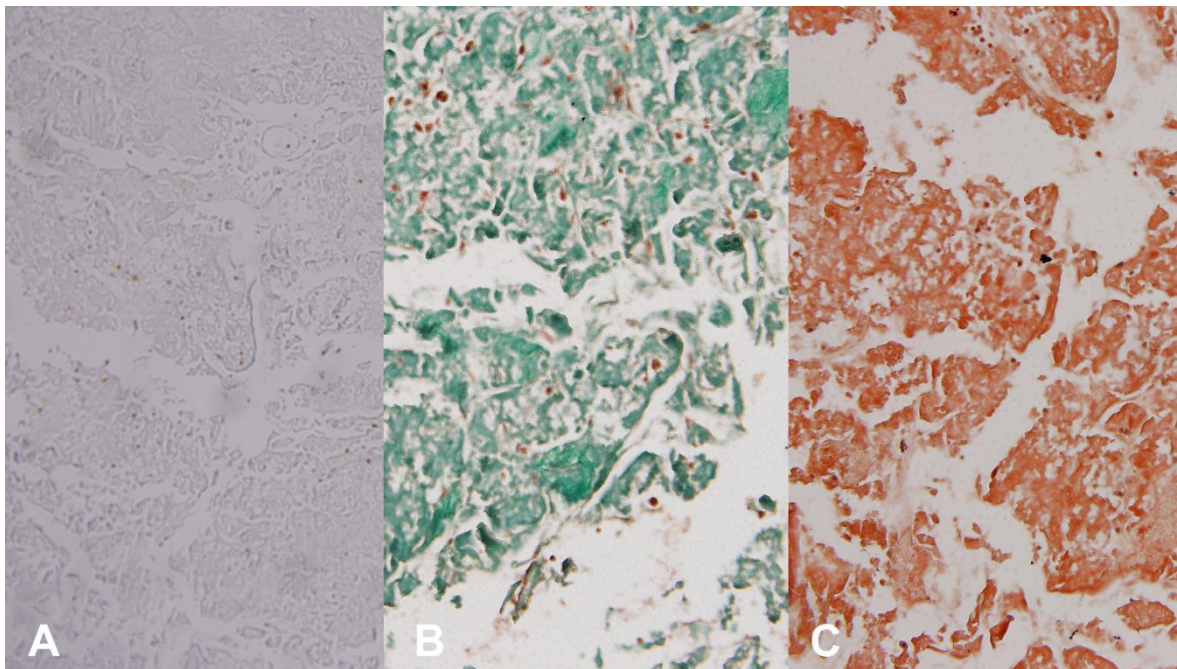


Figure 3: Pathological photographs. A: Study of hematoxylin and eosin. S and observe the presence of an amorphous tissue. B: Study applying the Gomori Trichrome technique where the negativity is observed confirming the presence of elastic and collagen fibers. C: Congo red technique that shows an apple-green birefringent intercellular substance corresponding to the amyloid substance.

Discussion

In 1863 Rudolf Virchow observed abnormal deposits of a substance he called "amyloid." This disease has systemic (the most frequent) and localized forms (unusual intracellular or extracellular deposits) [5].

Among the clinical manifestations of amyloidosis, the most frequent is renal, with patients with renal insufficiency. It has been suggested that there is an increase in renal size in amyloidosis, as indeed occurs by amyloid deposition, however, it is reported that in most patients the renal size is normal as was the case with this patient [1].

The most frequent extrarenal manifestations are cardiovascular events, which are associated with cardiomyopathy with diastolic dysfunction. On the other hand, manifestations of primary amyloidosis have been described in other organs and systems such as the digestive system, ocular, among others [1]. Within the manifestations of the nervous system associated with amyloidosis, carpal tunnel syndrome occurs in about half of the patients, and a symmetrical distal to proximal sensory neuropathy is sometimes described associated sometimes with dysesthesias [1]. On the other hand, the presence of amyloidomas in the central nervous system is very unusual, with few previous reports, which makes it difficult to diagnosis [2]. Another usual presentation is cerebrovascular amyloidosis, which constitutes the first cause of spontaneous intraparenchymal hemorrhage in older adults [6]. On the other hand, destructive spondyloarthropathy has been reported in patients with amyloidosis

secondary to renal dialysis, which causes amyloid deposits in the joints, intervertebral discs, ligaments and vertebral bodies along with the consequent myelopathy or radiculopathy [7]. In 2015 Liu et al. [8] reported a case with circumferential intrathecal ossification secondary to oculoleptomeningeal amyloidosis and as recently as 2019 Rotter et al [5]. reported the first case of focal intradural primary amyloidoma of the cervical spine.

In 1998 Horowitz S et al [3]. reported a case with primary spinal leptomeningeal amyloidosis and in the posterior fossa. However, the imaging findings were diffuse and were far from the nodular focal presentation of the case presented. So far, according to the review by the authors, 40 cases of spinal amyloidoma have been reported, including 16 cervical, 1 cervical, 19 thoracic, 3 lumbar and one thoracolumbar [9-25]. However, strikingly all of them extradural except for a patient reported by Smitherman et al [4]. in 2015 with cervical intradural amyloidoma, which to date was the first report of an intramedullary localized amyloidoma. This study is about the second report, taking into account the review carried out.

In Cuba, Alemán Rivera et al [26] reported in 2012 a case of lumbar extradural amyloidoma simulating a lumbar disc herniation.

The following 4 criteria are required for the diagnosis of primary amyloidosis:

1. Presence of a systemic syndrome related to amyloid.

2. Congo red positive staining in samples such as fatty aspirate, bone marrow or organ biopsy (in this case of the intracranial lesion).

3. Direct examination (immunoperoxidase staining, sequencing) that reveals amyloid related to light chains.

4. Evidence of cellular monoclonal expansion (such as serum or urinary monoclonal protein, plasma cells in bone marrow). After the pathological study, all these criteria for the diagnosis of the disease were confirmed. Biopsy is very important in the diagnosis of amyloidosis. It is suggested that this disease has been known since the 1700s, but it is not until 1800, with the appearance of methyl violet stains and in 1900 the technique of Congo red, which was done easier identification [26]. Regarding the pathological characteristics, amyloid deposits (amyloidomas) are extracellular, eosinophilic and chromatic meta microscopic [22]. The staining of Congo red is necessary for diagnosis and shows the characteristic of appearing as apple-green birefringent images, which is necessary for the typing of amyloidosis 1 (as in the case presented) and the use of antibodies that are directed against amyloidogenic proteins known in immunohistochemical and immunofluorescence studies.

In this patient, the uniqueness of the diagnosis of amyloidosis is presented at in this patient, the uniqueness of the diagnosis of amyloidosis is presented through the clinical and imaging suspicion of a dorsal extramedullary intradural tumor and it is not until the surgical approach is made that the true diagnosis is made, after the biopsy of the tissue removed. Although there were no elements in the striking personal pathological history, clinical suspicion of amyloidosis is essential in subjects with an underlying disease potential to cause amyloidosis as well as in the presence of proteinuria or auto-inflammatory syndromes, they must also be considered, in the differential diagnosis of cardiomyopathies, hepatomegalies, gastrointestinal disorders and peripheral neuropathies; Biopsy is an irreplaceable tool for the diagnosis of this disease.

Conclusions

Although extremely unusual, the presence of a spinal amyloidoma should be taken into account in the differential diagnosis in patients with extramedullary intradural tumors. In this context, laminectomy with total resection of the lesion and spinal decompression constitutes a therapeutic modality that not only allows the diagnosis to be established, but also improves the patient's symptoms.

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