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Original article

Neurosurgical aspects of dialysis-related spinal amyloidosis: Report of three cases and a review of the literature

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ABSTRACT

Background and purpose. – Osteoarticular manifestations of beta-2 microglobulin amyloidosis are often diagnosed in long-term dialyzed patients. However, spinal involvement is rare (10–25% of patients), and generally not associated with neurological deterioration. Compression of the spinal cord or roots is extremely rare, and probably under-recognized.
Methods. – The authors describe three cases of spinal stenosis presenting with neurological signs in long-term dialyzed patients, prospectively collected over 2 years in two different institutions and treated by surgical decompression. In all three cases, the main cause of neural compression was amyloid deposition in the spine, either extradurally in the ligamentum flavum or intradurally.
Results. – All patients improved after surgery and did not present any postoperative complications. However, two out of three patients with amyloid in the cervical spine required surgical revision to obtain a satisfactory decompression of the spinal cord.
Discussion. – The authors discuss spinal amyloidosis which is a well-known complication of long-term dialysis. However, neurological complications such as spinal cord or radicular symptoms have been rarely reported and, when present in dialyzed patients, are symptoms that are often attributed to other causes. To our knowledge, this is the first case series that demonstrates the relationship between neurological deterioration and amyloid depositions in the spinal canal that occur in long-term dialyzed patients. The prevalence of spinal stenosis related to the presence of amyloid in this specific subgroup of patients is probably underestimated.

1. Introduction

It is well known that patients with end-stage kidney disease (ESKD) treated with long-term hemodialysis may develop dialysis-related amyloidosis (DRA).

DRA is a dramatic painful complication of ESKD. Clinical diagnosis is generally made in patients treated by dialysis for more than 6–10 years. The main complications are carpal tunnel syndrome and chronic arthralgia, with the eventual occurrence of destructive arthropathies and spondyloarthropathies [1].

The main pathologic feature affecting the spine in patients with DRA is destructive spondyloarthropathy (DSA), which was first described as a consequence of DRA by Kunz et al. in 1984 [2]. It is characterized by vertebral body collapses, subluxation or spondylolisthesis and spinal stenosis, caused by amyloid deposition in vertebral bodies, disks and ligaments.

Patients may develop myelopathy and/or radiculopathy and sometimes surgery with decompression combined or not with arthrodesis is needed. Urgent surgery may also be required in rare cases, due to a rapid and very destructive evolution [3,4]. In these cases, the most appropriate surgical intervention must be carefully chosen. The use of instrumentation should be cautiously considered, because of bone fragility reported in these patients [5].

In this report, we present a series of patients affected by chronic renal failure surgically treated in two different institutions between 2009 and 2011.

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Table 1
Clinical, radiological and surgical characteristics of the patients.

Patient no.	Sex/Age (years)	Preoperative duration of hemodialysis (years)	Presenting symptoms	Lesion site	Type of surgery	Postoperative outcome
1	F/53	5	Progressive upper and lower limb motor deficit, urinary retention	Cervical, extradural (flavum)	Decompressive C3-C7 laminectomy	Amelioration of urinary symptoms, slight improvement of upper motor limb deficit, no recovery of walk
2	M/63	3	Severe tetraparesis, urinary retention	Cervical intra- and extradural	Decompressive C4-C7 laminectomy, with revision	Upper motor limb improvement, no recovery of walk and urinary symptoms
3	M/80	28	Back pain and neurogenic claudication	Lumbar, extradural (flavum)	L3-L5 laminectomy	Improvement of claudication

All cases presented with progressive myelopathy or radiculopathy, were related to amyloid depositions that caused compression of neural structures, and confirmed by MRI.

2. Materials and methods

Three cases were collected. Baseline clinical and radiological findings are summarized in Table 1. All patients were referred to the neurosurgical clinic after appearance of neurological signs and symptoms that were related to a compression of the spinal cord or roots. Decompressive surgery without arthrodesis was performed in all cases. The main cause of compression was a thickened ligamentum flavum. In one case, a cervical intradural extramedullary amyloidoma was also removed.

The diagnosis of DRA was confirmed by pathological examination of surgical samples of bones and ligamentum flavum, performed with Congo red stain and then assessed under polarized light.

2.1. Patients

2.1.1. Patient 1

A 53-year-old female patient with chronic renal failure was diagnosed with DRA after the discovery of carpal tunnel syndrome. She has been diagnosed with cervical spinal stenosis and treated conservatively years previously.

The patient was hospitalized at our institution for pyelonephritis of the transplanted kidney. During the hospitalization she began having severe pain in the four limbs that required morphine treatment, and developed several episodes of acute urinary retention. A subsequent MRI was performed, showing severe cervical stenosis at C3-C6 levels (Fig. 1). At that time, patient was affected by a severe upper and lower limb deficit, with complete inability to walk. The patient was then admitted to our department where a decompressive C3-C7 laminectomy was performed. Intra-operatively, there was evidence of severe stenosis with an abnormal thickened ligamentum flavum over 4 levels. Surprisingly, few arthritic modifications of posterior elements were present. Histology showed amyloid deposits in the ligamentum flavum and laminae.

Postoperatively, the patient showed a slow but progressive improvement with complete resolution of pain and urinary retention, but never regained the ability to walk.

2.1.2. Patient 2

A 63-year-old male patient had a 3-year history of hemodialysis for chronic renal insufficiency caused by IgA glomerulonephritis. He presented to our emergency department after the appearance of sensory disturbances to the four limbs associated with acute urine retention. He was then transferred to nephrology department where, the day after, he developed severe tetraparesis.

The neurological examination showed lower limb paraplegia and upper limb paresis (grade 2/5), which was more severe distally. There were no clinical signs of infection.

Spinal MRI revealed the presence of an intradural extramedullary lesion, extending from C4 to C7 and compressing the spinal cord, located mainly in the anterior part of the spinal canal (Fig. 2). Diffuse enhancement of the dural sac after gadolinium injection was also observed. A decompressive laminectomy was then urgently performed. The main goal of the planned procedure was to decompress the cord on the site of maximal external compression, and to obtain a diagnosis. For that purpose, a limited laminectomy involving C6 and C7 was performed.

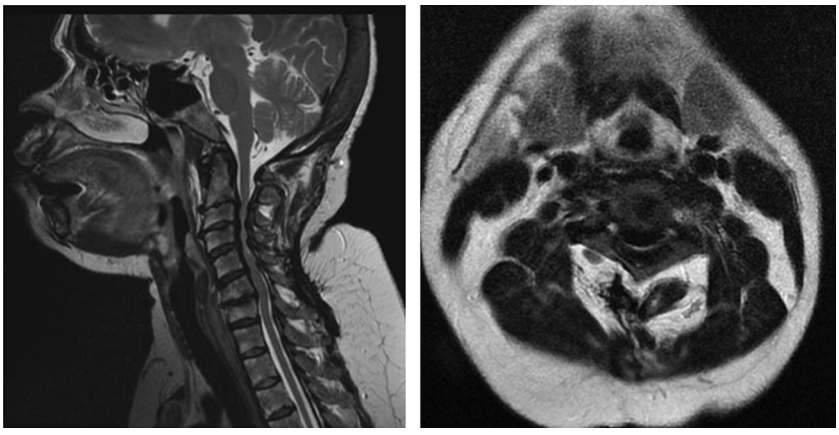

Fig. 1. Preoperative sagittal (left) and axial (right) T2-weighted MRI showing severe cervical stenosis from C3 to C6 due to hypertrophic ligamentum flavum in a 53-year-old female patient with chronic renal failure and progressive motor deficit in the lower limbs.

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