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Case Report

Epiduritis related to IgG4 disease: A very rare cause for spinal cord compression

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ABSTRACT

Background: Inflammatory pseudotumors are rare, and those attributed to immunoglobulin G4 (IgG4) diseases are even less frequently encountered. Here, we reviewed 41 cases from the literature of spinal inflammatory pseudotumors due to IgG4 and have added our single new case.

Case Presentation: A 25-year-old male presented with progressive back pain, bilateral paraparesis, and sphincter dysfunction. His deficit was attributed to MR-documented posterolateral lesion between the T5 and T10 levels for which he levels underwent a T1–T1010 laminectomy. The pathology revealed an immunoglobulin G4-related inflammatory pseudotumor. Postoperatively, the patient additionally required systemic and epidural administration of glucocorticoids.

Conclusion: IgG4-related disease is an emerging clinical condition that rarely involves the central nervous system. Spinal inflammatory pseudotumors, including IgG4 disease, should be more commonly considered among the potential differential diagnoses of lesions compressing the spinal cord.

Keywords: Epiduritis, Inflammation, Neurosurgery, Spinal cord, Spine surgery

INTRODUCTION

Immunoglobulin G4-related disease (IgG4-RD) is a fibroinflammatory disease characterized by sclerosing tumefactions and organ enlargement attributed to increased infiltration of lymphocytes and plasma cells (i.e., mainly into the pancreas, liver, salivary, and lacrimal glands, but rarely the kidneys, bile duct, and thyroid gland).[16,38] The central nervous system (CNS) is a very rare location for IgG4-RD lesions.[14,35] Here, we report a very rare case of radiologically-documented spinal cord compression between the T5 and T10 levels attributed to pachymeningitis caused by IgG4 disease that favorably responded to decompression.

CASE PRESENTATION

A 25-year-old male presented with 2 months of progressive upper back pain radiating around his trunk. However, within the past 7 days, he rapidly developed paraparesis (3/5 level), a relative

T6 pin level and dysuria. The thoracic magnetic resonance imaging (MRI) [Figure 1] showed posterolateral epiduritis between the T5 and T10 levels contributing to marked cord compression; it was isointense on T1, slightly hyperintense on T2-weighted images, and markedly enhanced with contrast. The patient underwent and emergent T5–T10 laminectomy. As surgery, the lesion appeared soft, yellowish, and there was a clear cleavage plane between tumor and the dura mater; gross total resection was readily achieved. The patient's postoperative course was uneventful; he fully recovered motor, sensory, and urological function and was discharged on the 5th postoperative day.

Pathology

The pathological examination [Figure 2] revealed diffuse infiltration of inflammatory cells within the fibrofatty tissue, accompanied by 50 IgG4+ cells/HPF and 50% IgG4+/IgG cell ratio. The serum level of IgG4 was 151 mg/dL. High serum IgG4 levels and immunohistopathological findings fulfilled the criteria for the diagnosis of a IgG4-RD lesion. Thus, the patient was placed on 0.6 mg/kg/d of methylprednisolone for the first 15 days postoperatively, followed later by a tapering dosage.

DISCUSSION

Frequency and location of IgG4-RD

IgG4-RD usually occurs in middle-aged patients (50–60 years) and demonstrates a significant male predominance.[38] It is a rare chronic inflammatory autoimmune disease which may affect almost any organ. It may also present as a single or multiorgan disease. It mostly occurs in the pancreas, salivary glands, and lacrimal glands. Only rarely is IgG4-RD found in the CNS where it mainly presents as a hypertrophic pachymeningitis and/or hypophysitis.[2,13,29,39,47] Notably, spinal pachymeningitis leading to spinal cord compression is extremely rare; we were only able to identify 41such cases in the literature [Table 1].

Pathological diagnosis of CNS IgG4-RD

Although immunohistochemistry is the gold standard for establishing the accurate diagnosis of IgG4 cells ratios, other major histopathological confirmatory features include lymphoplasmacytic infiltrates, fibrosis, and obliterative phlebitis.[10]

Radiological features for diagnosing CNS IgG4-RD

The radiological features for diagnosing CNS IgG4-RD are nonspecific. In fact, pathology may mistakenly lead to the diagnosis of other granulomatous diseases, such as lymphoma or even metastatic epiduritis.[2,20]

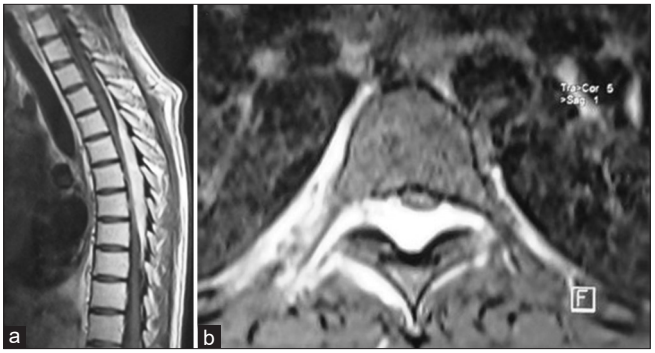

Figure 1: Sagittal (a) and axial (b) sections of a spinal cord magnetic resonance imaging on T1-weighted image with injection of Gadolinium showing a posterior epiduritis ranging from T5 to T10 responsible for compression of the spinal cord.

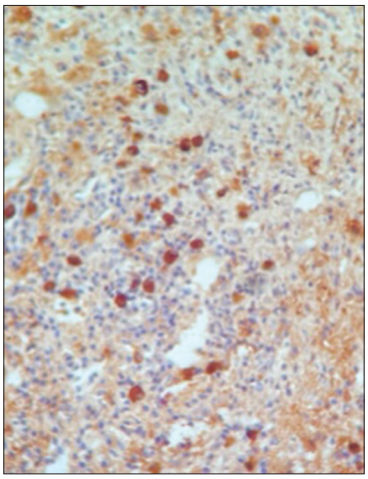

Figure 2: Pathological examination showing a diffuse infiltration of inflammatory cells within the fibrofatty tissue, with presence of 50 immunoglobulin G4 (IgG4)+ cells/high-powered field and 50% IgG4+/IgG cell ratio.

Surgical indications with the onset of neurological impairment

Emergent surgical decompression may be warranted when spinal pachymeningitis related to IgG4 disease causes neurologic impairment. Certainly, the spinal cord compression from IgG4 can be readily treated with emergent decompressive laminectomy with or without fusion. Although gross total excision is optimal, more restricted tumor removal may be dictated by critical adjacent structures.

No strict protocols to treat IgG4-RD lesions

As spinal pachymeningitis related to IgG4 disease is rare, there are no specific therapeutic protocols available.[14,47] Typically

Table 1: Table summarizing all previously reported cases of spinal epiduritis related to an IgG4 disease.[1,3-9,11-13,15,19-28,30-34,37,40-49]

Parameters	Number of patients
Age	
<40	12
40–60	23
>60	7
Sex	
Male	28
Female	14
Clinical features	
Motor deficit	17
Sensitive deficit	12
Cranial nerve palsies	5
Pain	15
Urinary dysfunction	5
MRI	
Spinal level	
Cervico-occipital hinge	2
Cervical	12
Thoracic	18
Lumbar	4
Multilevel	6
Spinal location	
Epidural	19
Intradural	21
Epidural and intradural	2
Cord compression	
Yes	30
No	7
Not specified	5
Extraspinal disease	
Submandibular gland	1
Cranial	8
Orbit	3
Lungs and pleura	3
Pituitary gland	1
None	30
Not specified	1
Surgery	
Laminectomy	25
Spinal biopsy	8
Other organs biopsy	3
Unspecified	7
Evolution	
Improved	35
Died	2
NA	5

NA: Not available, MRI: Magnetic resonance imaging

patients are first treated with systemic corticosteroids for at least 1 year.[17,36] Although lesions may recur following incomplete resection, they are more frequently encountered inpatients who did not receive adjuvant postoperative steroids. Further, immunosuppressive therapy may be warranted in patients showing systemic involvement.[17,18]

CONCLUSION

Inflammatory CNS IgG-RD lesions should be considered among the differential diagnoses for patients presenting with spinal cord compression.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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