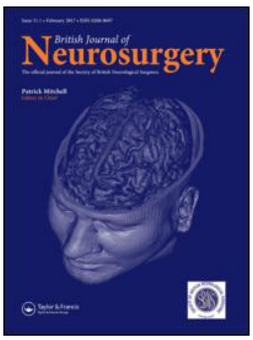

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ORIGINAL ARTICLE

IgG4-related disease presenting as intradural extramedullary lesion: a case report and review of the literature

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ABSTRACT

Object: IgG4-related disease (IgG4-RD) is a fibro-inflammatory disorder affecting various anatomical sites, and only recently was identified to affect the dura of the spine. The authors present the second reported case of an intradural extramedullary lesion consistent with IgG4-related spinal disease.

Methods: A literature review was performed that identified 15 other cases of spinal disease, and common features of all known reported spinal IgG4-RD are discussed.

Results: Spinal IgG4-RD typically affects males of approximately 50 years of age, and often presents as a T1 and T2 hypo- or isointense lesion that homogenously enhances. Surgical intervention typically involves subtotal resection or biopsy, and histopathologic findings include increased IgG4-positive cells or an IgG4:IgG ratio >40%. The disease responds well to steroids early on, and treatment can include adjuvant therapy such as azathioprine.

Conclusions: Systemic involvement is possible, and, early treatment can quickly minimize disease burden. Thus, increased suspicion would result in early diagnosis and improved prognosis.

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KEYWORDS
IgG4-related disease; immunohistochemical staining; spinal cord compression; surgery

Introduction

IgG4-related disease (IgG4-RD) is a fibro-inflammatory disorder involving formation of tumefactive lesions at multiple anatomical sites.[1,2] Lesions contain storiform fibrosis and lymphoplasmacytic infiltrate rich in IgG4.[1] The disease was originally recognized in the pancreas (called autoimmune pancreatitis) and was not recognized as a systemic condition until 2003, when extra-pancreatic manifestations were identified.[2] IgG4-RD can affect several alternative organs as well, often involving the bile ducts, salivary glands, lacrimal gland, mediastinal lymph nodes, retroperitoneum, aorta, lungs, and kidneys.[3–5] Lesions may be synchronous or metachronous.[6] The thoracic paravertebral space is commonly involved, most frequently in elderly males, in whom computed tomography (CT) will show thickening of soft tissue in the thoracic paravertebral space.[7]

In the central nervous system (CNS), IgG4-RD was initially identified in cases of hypophysitis.[4,8] The first case involving the extra-sellar cranial dura was described in 2009, and in the following years, CNS involvement has been reported relatively infrequently.[5,9,10] CNS related IgG4-RD often manifests as a pachymeningitis; an inflammatory disorder causing localized or diffuse thickening of the dura mater.[11,12] The disease most commonly involves the cranial dura, however spinal lesions are starting to be recognized.[9,11,13–15] To date, a total of 15 cases of spinal IgG4-RD have been presented in the literature, most of which are epidural or pachymeningeal in nature (Table 1). To our knowledge, there is currently only one reported case of intradural extramedullary disease manifestation of IgG4-RD.[12] We aim to present a second case of intradural extramedullary involvement while also providing the most up-to-date, and thorough, review of all spinal related IgG4-RD.

Methods

A case report is described from author experience. A PubMed search was conducted for manuscripts pertaining to IgG4 affecting the spine. Initial search items included 'immunoglobulin,' 'G4,' 'IgG4,' 'pachymeningitis,' 'spine,' and 'spinal.' Article reference lists and suggested similar studies through PubMed were utilized to identify additional pertinent studies. Articles pertaining to nonspecific inflammatory pseudotumors or cranial IgG4-RD were excluded. The search was limited to human studies and English-language literature.

Case report

A 68-year-old man presented with a 3-year history of intermittent upper thoracic pain radiating around his trunk to his chest with a 'squeezing sensation' in the T4 and T5 dermatomes. Multiple cardiac investigations were performed without identifiable cardiopulmonary etiologies. In the 6 months before presenting to neurosurgical attention, his pain progressively worsened, and he developed numbness along his trunk as well as his right lower extremity and distal left lower extremity. He additionally experienced dysequilibrium, and noted he could no longer walk independently, prompting him to seek further medical attention.

On examination, he had some mild weakness in his hip flexors bilaterally, and his left gastrocnemius and extensor hallicus longus muscles. He additionally endorsed right hemibody numbness to light touch below the T5 dermatome as well as in the distal left lower extremity. Patellar deep tendon reflexes were hyperactive. He had no ankle clonus. Magnetic resonance imaging (MRI) revealed a T1 and T2 hypointense, enhancing

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Table 1. Case review, clinical presentation and outcome.

Report	Patient age (y), gender	Medical Comorbidities	Clinical presentation	Serum IgG4, mg/dL (reference range)	Presence of extra-spinal disease	Extent of resection	Treatment	Outcome
Chan, 2009	37, M	NR	Bilateral lower extremity weakness, numbness, unsteady gait × 2 weeks	NR	NR	'Excision'	NR	NR
Choi, 2010	46, F	NR	Progressive bilateral lower extremity weakness and numbness × 2 weeks	NR	NR	Resection	Steroids	Improved exam to 'near normal' after initial surgery; progression 2 months post op required second resection (subtotal) with full recovery; no recurrence at 8 months
Lindstrom, 2010	55, M	Chronic Obstructive Pulmonary Disease	NR	NR	NR	NR	Steroids; Radiation Therapy	'doing well' at 15 months
Lindstrom, 2010	63, M	Diabetes Mellitus Type 2, Crohn's Disease, Diverticulitis	Bilateral hand numbness	NR	NR	NR	NR	Lost to follow-up
Tajima, 2012	64, M	Chronic Heart Failure	Gradually progressive dysphagia × 1 month; hoarseness*	221 (<104)	'slight swelling in both kidneys'	Open biopsy	Steroids	Marked improvement of symptoms and MRI findings at 3 weeks
Wallace, 2013	32, M	NR	Weakness	NR	NR	NR	NR	NR
Ezzeldin, 2014	55, M	NR	Diplegia of lower extremities and T4/T5 sensory level × 2 weeks	512 (7–89)	NR	CT-guided Biopsy	Steroids	Able to ambulate at discharge (timing not specified)
Gu, 2016	43, M	None	Neck pain × 15 days, bilateral lower extremity numbness/weakness, bowel/bladder dysfunction × 4 days	NR	Patient refused further testing	NR	NR	Resolution of symptoms at 6 months post-op
Radotra, 2016	50, M	NR	Progressive bilateral lower extremity weakness × 6 months	NR	NR	Subtotal excision	Steroids	Marginal improvement in strength 7 months post-op
Radotra, 2016	19, M	NR	back and left lower extremity pain × 1 year; subtle knee extensor weakness bilaterally	NR	NR	Biopsy	Steroids	Pain free at 6 months; stable exam
Ferreira, 2016	57, F	Graves' Disease, Fibromyalgia	Worsening low back pain × 2.5 years; bilateral lower extremity numbness and weakness	NR	None	Subtotal excision	Steroids	Recurrence and progressive paresis later improved after third resection and long-term steroids
Rumalla, 2017	50, M	None	Rapidly progressive severe back pain at T6 level × 3 months; acute onset paraplegia, T6 sensory level	NR	Right lung associated with adjacent vertebral involvement	Debulking	Steroids	Near normal lower extremity strength 2 months post-op
Lu, 2016	55, M	NR	Diffuse numbness and weakness × 6 months; constipation and dysuria × 7 days	82.2 (3–201)	NR	Subtotal excision	Steroids; cyclophosphamide	Recovered defecation/urination at 20 days; ambulated independently at 5 months
Williams, 2017	46, F	Fibromyalgia, Prior C6/7 anterior cervical discectomy and fusion	Worsening neck pain and bilateral upper extremity paresthesias and weakness × 4 months	NR	NR	Open biopsy	Steroids, azathioprine	Improving strength at 6 month post-op visit
Kim, 2014	52, F		Sudden weakness of bilateral lower extremities × 2 days; paraplegia, urinary retention	NR	None	Complete resection	Steroids	No significant improvement at 2 months post-op
Bridges & Than, 2017	68, M	Hypothyroidism	Intermittent thoracic spine pain × 3 years, progressive trunk numbness and bilateral lower extremity numbness × 6 months, dysequilibrium	NR	None	Subtotal resection	Steroids	Walking independently with resolution of pain at 3 months post-op

M: Male, F: Female, Y: years; IgG4: Immunoglobulin G4; HPF: High Power Field; ESR: Erythrocyte Sedimentation Rate (mm/hr); CRP: C-Reactive Protein (mg/L); Alk Phos: Alkaline Phosphatase; NR: Not Reported; post-op: post-operatively.

*Patient had 2nd lesion along the right foramen magnum.

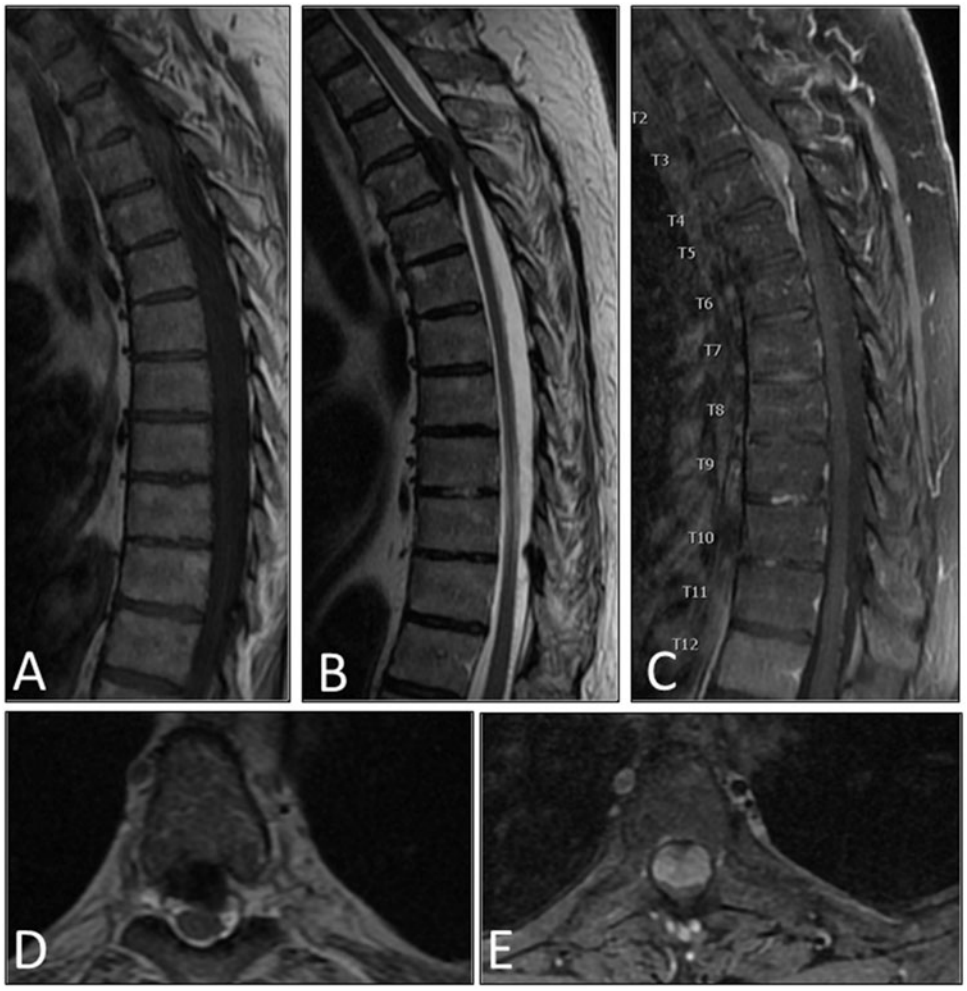

Figure 1. Thoracic spine MRI demonstrates an intradural extramedullary lesion along the ventral spinal canal extending from T3-T5 with what appears to be a dural tail. There is associated spinal cord compression most pronounced at T4. (A) Sagittal T1, (B) Sagittal T2, (C) Sagittal T1 post contrast, (D) Axial T2, (E) Axial T1 post contrast.

intradural extramedullary lesion extending along the ventral dura at T3-T5 with a dural tail (Figure 1). There was additionally subtle T2 hyperintensity within the adjacent spinal cord. Computed tomography showed no evidence of soft tissue involvement in the paraspinal tissue adjacent to the thoracic vertebra, and no calcification in the lesion (Figure 2).

He underwent a T3-T5 instrumented laminoplasty (i.e. the laminae were reaffixed with titanium cranial plates and screws) for debulking of the lesion. Ultrasound was utilized to plan the dural exposure and opening, and the lesion was noted to be hyperechoic. The T3 and T4 nerve roots were sacrificed bilaterally to gain circumferential access to the tumor, and the dentate ligaments were also sectioned. The lesion was quite firm, making it difficult to debulk with various modalities including ultrasonic aspiration. As such, a gross total resection could not be achieved, albeit a substantial debulking was accomplished. On review of the frozen specimen, the neuropathologist reported the lesion was collagenous in nature, but no further differentiation was possible. Final pathology revealed fibro-connective tissue with mixed inflammation containing predominantly plasma cells and focally increased eosinophils with less prominent populations of lymphocytes, macrophages, and neutrophils (Figure 3). IgG4 immunostaining showed an increased number of IgG4 plasma cells (with over 100 cells in some high-power fields), and the ratio of IgG4-positive plasma cells to IgG-positive plasma cells was significantly

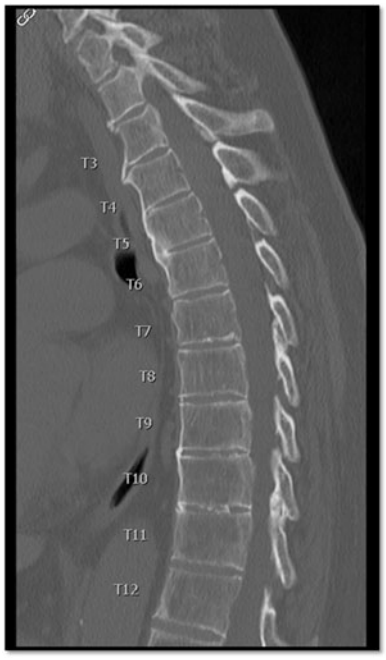

Figure 2. Thoracic spine non contrast CT shows no evidence of bone erosion or sclerosis.

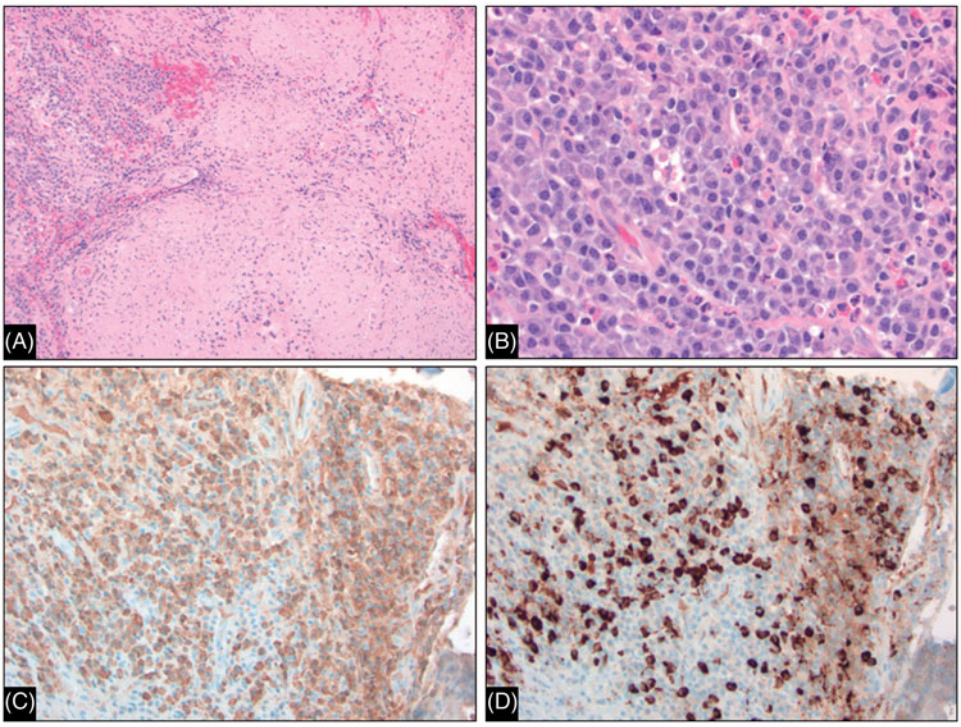

Figure 3. (A) 100x magnification view of a Hematoxylin and Eosin (H&E) stain shows extensive sclerotic/fibroconnective tissue deposition (on the right) with mixed inflammation (upper left). (B) 400× magnification shows most of the inflammatory cells to be plasma cells. (C) 200× magnification view of IgG immunostaining demonstrates most of the plasma cells are positively staining. (D) 200× magnification view of IgG4 immunostaining (from the same area as shown in (C), shows that a large percentage (estimated at up to 40% in some fields) of the IgG positive cells are IgG4 positive, supporting the diagnosis of IgG4 sclerosing disease. Individual high power fields showed up to 100 IgG4 positive cells.

increased, to approximately 40% in some areas. These findings were suggestive of an IgG4-related autoimmune/sclerosing disease. A positron emission tomography/CT showed no evidence of systemic IgG4 disease.

He was discharged from the hospital 10 days after surgery with continually improving strength and resolving paresthesias. He has been subsequently followed by rheumatology department and is treated with prednisone 10 mg daily. By 3 months postoperatively, he was walking independently and had resolution of his back pain. An MRI at that time showed significant decompression of the spinal cord without much residual lesion (Figure 4).

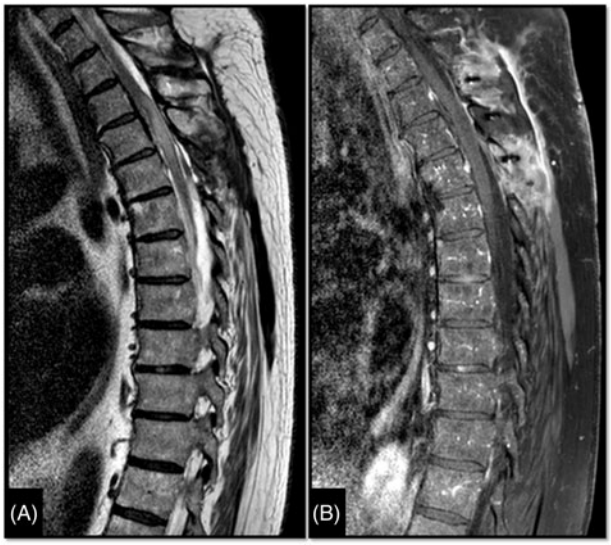

Figure 4. Thoracic spine MRI at 3 months following surgery reveals significant decompression of the spinal cord without much residual lesion. (A) Sagittal T2, (B) Sagittal T1 post contrast.

Discussion

IgG4-RD is a relatively new disease, first identified in the spine in 2009, and the clinical picture continues to evolve in published reports.[5] A total of 15 cases of spinal IgG4-RD to our knowledge have been reported, with this case totaling the number to 16 (Table 1). Among these 16 patients, 12 (75%) were male, and 4 (25%) were female and average age was 49.5 years. Patients commonly presented with progressive pain, paresthesias, and weakness, typically over the course of weeks to months. Lesions were localized to the cervical spine in 2/16 (12%) cases, thoracic spine in 7/16 (44%) cases, and lumbar spine in 3/16 (19%) cases. Lesions spanned the cervicothoracic junction in 4/16 (25%) cases. The location of the lesion relative to the dura was reported in 14 of the cases, with 12/14 (86%) lesions involving the dura and epidural space. One of these cases specifically involved the L5 nerve root.[16] Two (14%) patients, one of which is the case we report, had intradural extramedullary lesions, both with mass effect on the upper thoracic spine, resulting in bilateral lower extremity symptomatology. Magnetic resonance imaging results were reported in the majority of cases (10/16), and all lesions demonstrated homogenous enhancement (Table 2). When reported, lesions were either hypo- or isointense on T1 and mostly hypo- or isointense on T2 (with exception of one lesion reported to be hyperintense on T2).[17] Ten authors clarified the extent of resection, with subtotal resections accounting for 5/10 (50%) surgeries,

Table 2. Lesion location, pathology, and imaging characteristics.

			MRI findings				
Report	Location of lesion	Level of lesion	T1	T2	Contrast	Intraoperative description of specimen	Pathology/Histology findings
Chan, 2009	Epidural	T5-T10	NR	NR	Homogenous enhancement	Firm yellow mass arising from the dorsal aspect of the dura	Sclerosis, phlebitis, IgG4-positive cells 310/HPF, IgG4:IgG 70%
Choi, 2010	Epidural	T9-T11	NR	Isointense	Homogenous enhancement	Firm epidural mass compressing the spinal cord	IgG4-positive cells >20/HPF
Lindstrom, 2010	NR	\C3-C7	NR	NR	NR	NR	Severe fibrosis, severe lymphoplasmacytic infiltration; IgG4-positive cells 46/HPF, IgG4:IgG 60%
Lindstrom, 2010	NR	C2-C3	NR	NR	NR	NR	Moderate fibrosis, severe lymphobplasmacytic infiltration; IgG4-positive cells 11/HPF, IgG4:IgG 30%
Tajima, 2012	Epidural	T3-T11	NR	NR	Homogenous enhancement	NR	IgG4-positive plasma cells
Wallace, 2013	Nerve root dura	L5 nerve root	NR	NR	NR	NR	Lymphoplasmacytic infiltrate, storiform fibrosis; IgG4-positive cells 11/HPF, IgG4:IgG 35%
Ezzeldin, 2014	Epidural	T2/3	NR	Isointense	Homogenous enhancement	Not applicable	Fibrotic and myxoid change; no storiform fibrosis nor obliterative phlebitis; IgG4 positive cells 2/HPF
Gu, 2016	Epidural	C4-T2	Hypointense	Hyperintense	NR	'fish-meat like rubbery lesion' tightly adherent to the dura	IgG4:IgG >40%; IgG4 positive cells >50/HPF
Radotra, 2016	Intradural/ Meningeal	L2/3	Isointense	Hypointense	Homogenous enhancement	Firmly adherent to the dura (no differentiation from normal dura)	Storiform fibrosis, plasma-rich infiltrate, strong IgG4 positivity, IgG4:IgG 50%
Radotra, 2016	Dural	L2/3	Isointense	Hypointense	Homogenous enhancement	Dura was diffusely thickened and hypertrophic, moderate vascularity	IgG4:1gG 40%
Ferreira, 2016	Epidural	T10-T12	Isointense	Hypointense	Unknown	NR	Lymphoplasmacytic infiltrate with fibrosis; no phlebitis; IgG4:IgG >50%
Rumalla, 2017	Vertebral bodies, Epidural extension	T4-6	NR	NR	Homogenous enhancement	NR	Fibroinflammatory consolidation; 'significant number' of IgG4-positive plasma cells
Lu, 2016	Dural	C2-T9	NR	NR	Homogenous enhancement	'tawny and closely connected to the dura'	sclerosis; predominance of plasma cells; IgG4:IgG >40%; IgG4 positive cells >10/HPF
Williams, 2017	Paraspinal and epidural	C4-T1	NR	NR	NR	Firm tissue with no discernable borders from paraspinal musculature	Dense fibrosis, lymphoplasmacytic inflammation, IgG4-positive cells 10/HPF
Kim, 2014	Intradural, extramedullary	C7-T5	NR	Hypointense	Homogenous enhancement	Moderate adhesion to anterior dura	Sclerotic fibrosis, lymphoplasmacytic infiltration, 'many' IgG4-positive plasma cells
Bridges & Than, 2017	Intradural, extramedullary	T3-T5	Hypointense	Hypointense	Homogenous enhancement	Firm, difficult to debulk	Fibroconnective tissue, mixed inflammation, plasma cell predominance, IgG4:IgG 40%, IgG4 positive cells >100/HPF in some slides

and biopsies (CT-guided or open) accounting for 4/10 (40%) of surgeries. Complete resection was reported in only one report.[12] In open biopsies and surgical excisions, the lesions were almost universally described as firm and adherent to the dura.

The diagnosis of IgG4-RD is made by histopathological findings, requiring the presence of at least 2 criteria including lymphoplasmacytic infiltrate, fibrosis in a storiform pattern, and obliterative phlebitis.[1,15,18] In the literature regarding spinal IgG4-RD, most authors ($n = 10/14$) specified the number of IgG4 cells per high power field (HPF) and/or IgG4:IgG ratios (Table 2). The IgG4:IgG ratio was >40% in 8/10 (80%) cases and between 30–39% in 2/10 (20%) cases. The number of IgG4-positive cells ranged from 2-310/HPF. In spinal IgG4-RD, 3 authors documented the serum IgG4 levels, 2 of which (66.7%) were elevated.[11,19] The presence of extra-spinal disease was rarely reported. However, early diagnosis of IgG4-RD has been encouraged, given its known multisystem involvement, which is chronically progressive.[13] This condition, which has now been described in most organ systems, is considered analogous to sarcoidosis, another systemic disease with varied organ manifestations linked by histological appearance.[1]

Rapid disease progression and mass effect necessitates prompt spinal decompression, optimizing prognosis while also providing adequate tissue for histopathological examination.[3,13,17] Adjuvant treatment is with glucocorticoids, and relapses are common following steroid taper or discontinuation.[3,13] Alternative options have been utilized for remission-maintenance therapy, including methotrexate, azathioprine, and mycophenolate mofetil.[3] Of the 16 cases of spinal IgG4-RD, all patients underwent initial surgery for biopsy and/or debulking. Twelve authors reported their treatment regimen, and all 12 (100%) utilized glucocorticoids (Table 1). One patient also received radiation, another patient received adjuvant cyclophosphamide, and a third patient received azathioprine in addition to the steroids.[4,9,15]

Following the initial response to glucocorticoids, the disease course is progressive in nature.[13] In this literature review, 13 authors reported their outcomes, with follow-up time ranging from 3 weeks to 7 months 9 (Table 1). Of these patients, 11/13 (85%) demonstrated some improvement, while 2/13 (15%) showed no improvement. Tajima, et al reported improved imaging findings after 3 weeks of treatment following biopsy.[19] Meanwhile, Choi et al and Ferreira et al encountered recurrence, necessitating further surgical intervention.[20,21]

The case presented in this manuscript represents the second case of intradural extramedullary IgG4-RD, bringing to light the increasing prevalence of this disorder and its varied manifestations. It would be useful for spine surgeons and pathologists to consider this disorder when reviewing imaging so that it can be included on the differential diagnosis, a workup for systemic disease can be performed, and treatment can be rendered in a timely fashion.

Conclusion

The authors describe a rare case of spinal IgG4-RD presenting as an intradural extramedullary mass with compression of the upper thoracic spinal cord. A literature review identified 15 cases of spinal IgG4-RD published in the English literature, this case being the 16th, but only the second presenting as an intradural extramedullary lesion. Spinal IgG4-RD typically affects males of approximately 50 years of age, and often presents as a T1 and T2 hypo- or isointense lesion that homogenously enhances. Surgical intervention typically involves subtotal resection or biopsy, and histopathologic findings include increased IgG4-positive cells or IgG4:IgG ratio >40%. The disease responds well to steroids early on, and treatment can include adjuvant therapy such as azathioprine. Systemic involvement is possible, and, early treatment can quickly minimize disease burden. Thus, increased suspicion would result in early diagnosis and improved prognosis.

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All authors certify that this manuscript is a unique submission.

Disclosures statement

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