Inflammatory myofibroblastic tumor in the intradural extramedullary space of the lumbar spine with spondylolisthesis: case report and review of the literature

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Abstract An inflammatory myofibroblastic tumor (IMT) is a rare disease entity reported to arise in various organs but still thought to be a neoplastic or reactive inflammatory condition controversially. The author reports an extremely rare case of intradural extramedullary IMT of lumbar spine which was presenting radiculopathy and neurogenic intermittent claudication due to concomitant spondylolisthesis.

Keywords Inflammatory myofibroblastic tumor · Lumbar spine · Spondylolisthesis · Claudication

Introduction

Inflammatory myofibroblastic tumor (IMT), synonymous with inflammatory pseudotumor, had been known to involve the lung or pleura (or both), until the cases described in the abdomen, skin and soft tissues, spleen, heart, bladder, upper respiratory tract, mediastinum, and kidney [9, 22]. Numerous nomenclatures were designated for this unusual pathological entity such as “inflammatory pseudotumor”, “plasma cell granuloma”,

“pseudosarcomatous myofibroblastic proliferation”, “inflammatory myofibrohistiocytic proliferation”, etc. In the 2002 World Health Organization classification of soft tissue tumors, these lesions were renamed [16].

Although the central nervous system is the rare site affected by IMTs, more over 100 sporadic cases have been reported in the literatures [11, 26], but intraspinal IMTs were extremely rare. Except our own case, 13 cases of intraspinal IMT have been reported in the literatures. Among them only one case in the lumbar spine has been reported but this case was not described well in the literature. This is the first case of intradural extramedullary (IDEM) IMT of lumbar spine who had radiculopathy and neurogenic intermittent claudication due to concomitant spondylolisthesis.

Case report

A 56-year-old woman was admitted complaining of low back pain and radiating pain on right posterolateral thigh. She had had low back pain for about 4 years and it was so worsening that she could not stand and walk over 10 min without back pain. On neurological examination, neither motor deficit nor changing in deep tendon reflex could be elicited. Plain radiographs showed spondylolisthesis of L5 on S1. Magnetic resonance imaging (MRI) showed a well-circumscribed and round IDEM mass compressing dural sac at L5 body level (Fig. 1) Her MRI showed high signal intensity on T2-weighted images and strong homogeneous enhancement. All laboratory findings were normal. We performed L5 total laminectomy. After dural incision, there was a soft and whitish color intradural mass displacing thecal sac from right to left and extended to right S1 root sleeve. It was separated and removed totally with some

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Fig. 1 A plane radiograph with flexion showed anterior slippage of L5 on S1 (a). T2-weighted magnetic resonance imaging (b) and T1-weighted imaging (c) showed iso-signal intense mass lesion

comparing with cauda equina. After gadolinium contrast enhancement (d, e), T1-weighted imaging with gadolinium enhanced MRI showed strong enhancement compressing right S1 nerve root sleeve

difficulty from nerve root. L5-S1 posterior lumbar interbody fusion was performed.

The histological diagnosis was compatible with IMT. Histopathologic examination revealed nodular proliferation of spindle or oval cells with dense infiltration of lymphoplasma cells in hyalinized stroma. Dense infiltration of plasma cells was noted with lymphoid follicles. Some plasma cells contained Russell bodies (globular plasma cell inclusions), representing aggregates of immunoglobulins synthesized by the plasma cells (Fig. 2). Immunohistochemical

examination showed expression of vimentin in the proliferated stromal cells. Possibilities of neurogenic tumor and solitary fibrous tumor were ruled out because no immunoreaction of S100 protein and CD34 was noted in the proliferated cells. There was no overexpression of p53 and the Ki-67 proliferative index was less than 2% (Fig. 2).

Postoperatively her intermittent claudication was gone and only paresthesia on her right toes remained. There is no evidence of recurrence of tumor after surgery for 24 months without any adjuvant therapy.

Fig. 2 a Photomicrographs of the tumor showed nodular proliferation of spindle or oval cells (asterisk) with dense infiltration of lymphoplasma cells in hyalinized stroma (H&E $\times 200$). b Note dense infiltration of plasma cells (black arrows) with Russell bodies (white arrowhead), representing aggregates of immunoglobulins synthesized by the plasma cells (H&E $\times 400$)

Discussion

Inflammatory myofibroblastic tumor is a recognized spindle cell neoplasm composed of lymphocytes, plasma cells, histiocytes, macrophages, and foam cells within a spindle-shaped stroma and its nature and origin are still controversial. After initial publications on “IMT” considered this entity synonymous with the inflammatory pseudotumor [20], some neuropathological studies used the terms IMT and inflammatory pseudotumor interchangeably [6, 7, 16]. But the recent report about IMT of the central nervous system by Swain et al. [24] argued that it had distinct neoplastic characteristics compared with those of inflammatory pseudotumor. After IMT was defined as “a distinctive lesion composed of myofibroblastic spindle cells accompanied by an inflammatory infiltrate of plasma cells, lymphocytes, and eosinophils” in the 2002 World Health Organization [5], it has been reported involving various organs increasingly but intraspinal involvement of IMT is still rare. A total of 12 spinal lesions have been reported and only one of them at the level of lumbar lesion (Table 1). There is no age, sex, or geographic predilection to their incidence. They are important because of the difficulty in differentiating them from true other neoplasms clinically, radiologically, and intraoperatively.

The location of involvement of these spinal tumor could be anywhere such as extradural space, IDEM space, and intramedullary space. The most reported case revealed some difficulties in differentiating them from true neoplasms clinically, radiologically, and intraoperatively. But neither accurate pathogenesis nor causative events is known. Some cases have been associated with malignancy or tuberculosis as satellite lesions. It is also suggested to be

associated to cocaine consumption [4, 15, 17] and viral infection such as Epstein–Barr virus [3, 4]. Therefore, IMT can be diagnosed after elimination of any causative organisms such as fungi and bacteria.

No inciting factor could be determined in this case. It is usually possible to present nonspecific clinical features including fever, weight loss, and pain. Sometimes laboratory findings elicited anemia, thrombocytopenia, polyclonal hypergammaglobulinemia and elevated erythrocyte sedimentation rate, etc. But these findings could not help clinician to make a diagnosis.

Radiologically, IMT appear as solid intraspinal tumor. Common MRI findings in the most documented cases of IMT are low or iso-signal intensity comparing with conus medullaris in T2-weighted images and homogeneous contrast enhancement. A low signal intensity on T2-weighted images, indicating either elevated cellularity or a high nuclear-to-cytoplasmic ratio, is also typical of malignant neoplasms [9]. In our case, MRI showed a well-circumscribed round IDEM mass compressing thecal sac from ventral side of spinal canal at the level of L5 body (Fig. 1) The lesion gave us so strong evidences that this lesion was considered as IDEM tumor such as schwannoma or meningioma because it revealed relatively iso-signal intensity on T2-weighted images and showed strong homogeneous enhancement as IDEM tumor.

In our case, there was no bony involvement causing structural instability. We consider spondylolisthesis is not concerned with this tumorous condition but it might contribute to her symptom. Clinically, IMT showed diverse symptom and sign because it could arise in any vertebral canal. In this case, IMT might worsen neurogenic claudication caused by spondylolisthesis. Especially spinal

Table 1 Characteristics of previous reported cases intraspinal inflammatory myofibroblastic tumors

References	Age	Sex	Intraspinal location	Level of vertebra	Treatment	Recur	FU (month)
Roberts et al. [22]	58	F	ED	T9-T11	Resection	N	5
Aizawa et al. [1]	46	M	IM	C7-T1	Resection	N	24
Jeon et al. [16]	60	F	IDEM	Lumbar	Resection	N	NA
Hsieh and Lin [14]	37	M	IDEM	T5, T12-L1	Resection	N	14
Eimoto et al. [8]	37	M	IDEM	C3-4	Resection + RTx	N	9
Gilliard et al. [10]	45	M	ED	C3-T2	Resection	N	2
Hsiang et al. [13]	57	M	ED and IDEM	T12-L1	Resection + reop	Y	67
Roberts et al. [21]	39	F	ED	T5-T6	Resection	N	6
Lee et al. [19]	3	F	IM	C2-T10	Resection	N	12
Kilinc et al. [18]	34	M	IM	T11-12	Resection	N	3
Despeyroux-Ewers et al. [7]	22	F	IDEM	T9	Resection, reop + CTx + RTx	Y	11
Seol et al. [23]	44	M	ED	T1-T7	Resection	N	12
Boutarouch et al. [4]	30	F	IDEM	C4-T2	Resection	N	6

ED epidural, IDEM intradural extramedullary, IM intramedullary, RTx radiotherapy, CTx chemotherapy, reop reoperation, NA not available

pathology such as schwannoma of cauda equina can mimic common complaints of low back pain seen in clinical practice. But there were few reports that tumor located in lower lumbar spine was exacerbating neurogenic claudication.

Some authors recommended radiation therapy despite uncertainty about its efficiency in case of involving the nervous system [12, 25]. Although systemic steroids and immunosuppressive drugs are also given in IMTs in other organs and lead to mass volume decreasing effectively, surgical excision is usually needed in IMT of the nervous system because of the emergent requirement of relieving the mass effect; it is generally curative when total removal is performed [2]. We did not perform the adjuvant therapy because the mass was completely resected. Bony fusion and rigid fixation were performed for relief of her symptoms and instability. For 2-year follow-up period, there was no evidence of recurrence. Although there were no large-scaled reports about the treatment of IMTs because of the rare incidence involving the nervous system, there were only two recurred cases (14.2%) of 14 IMT cases (including 13 cases reported in the literatures and this case). In a solid intra- or extradural tumor of the spine IMT is a rare differential diagnosis without any specific imaging characteristics. Recurrence rate is about 15%, and radiation or chemotherapy is not required.

Conclusion

In neurosurgical field, the degenerative spinal disorder usually presented neurogenic claudication and back pain. But it may be exacerbated by a rare tumorous condition due to compressing cauda equine and it may be curable only by resection. It is the first case of IMT in the IDEM space of the lumbar spine presenting neurogenic claudication.

Conflict of interest None of the authors has any potential conflict of interest.

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