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Case Report

Cervical epidural neurosarcoidosis: A case report and literature review

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ABSTRACT

Background: Neurosarcoidosis is a rare disease. In the spine, it commonly presents as an intramedullary lesion. Epidural spinal lesions are extremely rare.

Case Description: A 29-year-old patient presented with a 22-month history of progressive neck, upper limb pain, and myelopathy. The cervical MRI showed a large epidural mass infiltrating the paraspinal soft tissue. After an open biopsy, the diagnosis of neurosarcoidosis was established and was followed-up by appropriate medical management.

Conclusion: To manage cervical epidural neurosarcoidosis, first, you must obtain a tissue diagnosis and then follow with appropriate medical management.

Keywords: Cervical, Epidural, Neurosarcoidosis

INTRODUCTION

Sarcoidosis is a multisystem disease attributed to an accelerated immune response of unknown etiology.[7] Systemic sarcoidosis occurs in 1–3/100,000 individuals; only 5% of lesions involve the nervous system (e.g., neurosarcoidosis). Just 10% of those with neurosarcoidosis, which have spinal lesions that are typically located within the cord itself (i.e., intramedullary).[7]

Here, we present a 29-year-old female with left-sided C2–C7 epidural cervical neurosarcoidosis diagnosed utilizing an open biopsy, followed by appropriate medical management.

CASE REPORT

A 29-year-old female presented with a 22-month history of progressive dysphagia, neck pain, and cervical myelopathy. Although she had no motor deficit, she exhibited impaired pin appreciation on the left side from C2–C7, bilateral Hoffman's signs, and diffuse upper and lower extremity hyperreflexia.

The computed tomography (CT) and full neuraxis enhanced MR studies showed a left-sided epidural enhanced mass from C2 to C7 compressing the spinal cord and displacing it to the right with a paravertebral extension [Figure 1a-d].

Three days after the presentation, a CT-guided biopsy was performed. However, due to the initiation of steroids (dexamethasone 4 mg every 6 h), the tumor had significantly decreased in size (e.g., repeat MRI showed that the lesion had profoundly regressed), and a biopsy was no longer feasible/reliable [Figure 2a-d].

The next MR performed 2 weeks after cessation of steroids revealed significant tumor recurrence [Figure 3a and b]. The repeated CT-guided biopsy revealed a granuloma in a background of nonneoplastic skeletal muscular and fibrous tissues. This was followed by an open anterior C5–C6 biopsy of the large recurrent extradural lesion; the latter confirmed the diagnosis of neurosarcoidosis (i.e., nonnecrotizing granulomatous lymphadenitis) neurosarcoidosis [Figure 4a and b]. She was later discharged on 60 mg prednisone/day and placed on an 8-week taper. Three months later, she exhibited substantial clinical improvement.

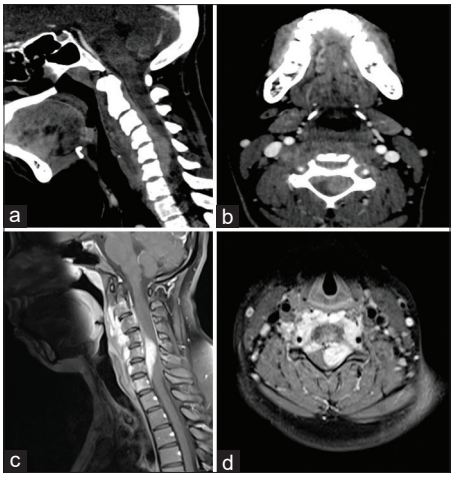

Figure 1: Cervical CT with contrast (a: sagittal and b: axial) and MRI (c: sagittal and d: axial) shows a longitudinally oriented epidural enhancing soft-tissue mass lesion occupying the left side of the spinal canal opposite C2 down to the C7 vertebra. MRI (c: sagittal and d: axial) shows intraspinal epidural and paraspinal avidly enhancing cervical lesions with multiple neural foraminal extensions and cord compression, which was most severe at C5.

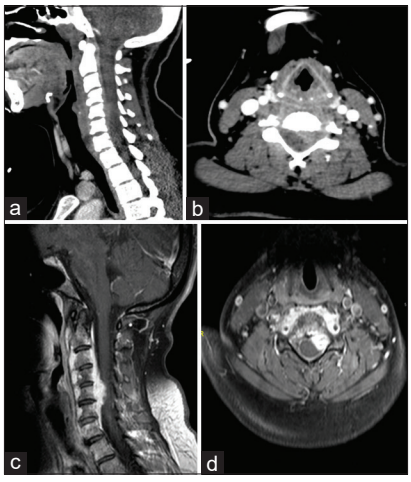

Figure 2: Contrasted cervical CT (a: sagittal and b: axial) during the CT-guided biopsy, preprocedure scan demonstrated improvement of the prevertebral tissue, likely related to steroids treatment. Due to the lack of enough safety margin to perform biopsy, the procedure was cancelled. Repeated cervical MRI (c: sagittal and d: axial), showed the lesion response to the steroids more profoundly.

DISCUSSION

Sarcoidosis is a chronic systemic inflammatory disease characterized by noncaseating granulomas on pathological examination. It most commonly affects the lungs and mediastinal lymph nodes.[7] Only approximately 5% of all sarcoidosis patients have central nervous system involvement; most of these lesions are intracranial.[7,6]

Just 10% of neurosarcoidosis involve the spine. Seven similar cases found in the literature. Of interest, most are intramedullary lesions, and few involve the extradural compartment [Table 1].[2,5,6]

Most cases of neurosarcoidosis have normal levels of ACE, and the histology demonstrates nonnecrotizing granulomas with an inflammatory background [Table 1].[1,7,8]

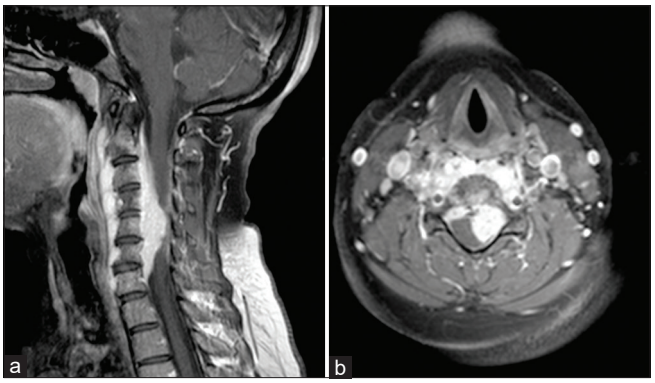

Figure 3: The cervical spine MRI (a: sagittal and b: axial) was repeated after 2 weeks from the previous MRI [Figure 2] while stopping the steroids and showed a significant recurrence.

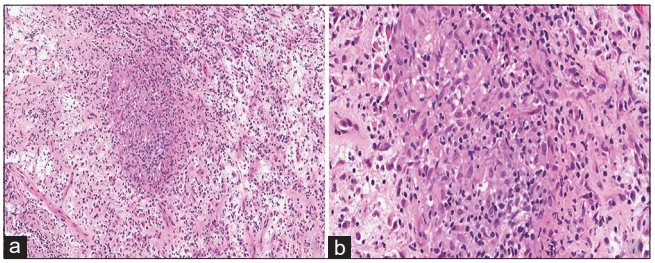

Figure 4: (1) Low magnification – H&E stain (a) shows a well-demarcated granuloma. (2) High magnification – H&E stain (b) the granuloma consists of epithelioid histiocytes and mature lymphocytes.

Table 1: Cases of spinal epidural neurosarcoidosis.

No	Author-year	Presenting symptoms	Number of cases	Level of the mass	Systemic involvement	Serum ACE\CSF ACE	Pathology location and finding	Surgical management if any	Medical management	Patient outcome and duration of follow up	Comments
1	Paglia *et al.*, 2019[6]	Back pain, lower limbs weakness, and paraesthesia	1	T11–L1	Lung nodule, lymph adenopathy of the metastatic lymphnodes and uterine fibromatosis	Not done	Location: T11-L1 finding: fibrous-collagenous tissue referable to yellow ligament infiltrated by a population of bland appearing epithelioid histiocytes aggregated in nonnecrotizing granulomas, together with a number of multinucleated giant cells	T11–L1 bilateral laminectomy and complete macroscopic excision of the lesion	High-dose methyl prednisolone intravenously for 3 days followed by prednisone at the dose of 1 mg/kg body weight daily	After 6 months from the surgical removal of the mass, total body CT scan showed complete diseaseremission	
2	Longo *et al.*, 2019[3]	Back pain, nondermatomal sensory loss, unsteady gait	1	T4-T6 T8 L2-L3	None	Serum ACE=12U/L CSF ACE levels were not done	Location: CT-guided needle-core biopsy of T5 lesion Finding: noncaseating granulomatous inflammation. composed of mononuclear histiocytes and multinucleate giant cells. Plasma cells, and lymphocytes	None	Prednisone 80 mg daily	Follow-up over 18 months after initial presentation, she was neurologically intact	Multiple lesions
3	Munakomi, 2018[4]	Progressive bilateral lower limb weakness and sensory loss below T7	1	T3-T4 T9-T10	Multiple fibrotic changes in the right upper lung with a posteriorly based pleuralnodule	Serum ACE was marginally high (not specified) CSF ACE levels were not done	Histology was negative for any malignant cells. It revealed presence of lymphocytes with few scattered noncaseating granulomas	Thoracic laminectomy and subtotal resection of the lesion	Oral Prednisolone (tapering dose starting from 40mg/day) and weekly methotrexate (7.5 mg) therapy	The patient could walk independently in a follow-up visit at 4 weeks	Multiple lesions. Surgery level was not described
4	Galgano *et al.*, 2017[2]	Thigh pain, lower extremity weakness, and episodic urinary incontinence sensory deficit below T6	1	T3–T6, initial presentation T3–T7, recurrenceafter 3 months T1–T8 recurrence 2 months after 2ndsurgery	CT-PET demonstrated moderate uptake in the axial and appendicular skeleton	Serum ACE=21U/L CSF ACE levels were not done	Initial biopsy at presentation: T3–T4 scant fragments of fibroadipose tissue with focal infiltration by aatypicalhistiocytic cells After recurrence, second biopsy: fibroadipose and elastic tissue with granulomatous inflammation adjacent to necrotic elastic tissue	Initial surgery left T3–T4 hemi-laminotomy and debulking of the epidural mass after recurrence T5–T6: decompressive laminectomy with debulking of the epidural lesion	After the second surgery: methotrexate and steroid taper	1 year after initial presentation, fully ambulatory with a normal neurologic exam.	The patient had poor compliance with medical therapy the second episode of recurrence was managed medically

(*Contd...*)

Table 1: *Continued.*

No	Author-year	Presenting symptoms	Number of cases	Level of the mass	Systemic involvement	Serum ACE\CSF ACE	Pathology location and finding	Surgical management if any	Medical management	Patient outcome and duration of follow up	Comments
5	Barazi *et al.*, 2008[1]	Back pain, altered sensation in lower limbs	1	T11–L1	None	Serum ACE=25 U/L CSF ACE levels were not done	T11–L1 extensive granulomatous inflammation with nonnecrotising epitheloid granulomata and langhans type giant cells	Bilateral T11, T12, and L1 laminectomies and complete macroscopic excision of the lesion	Not described	Four months postoperatively was found to be neurologically intact, with no radiological evidence of recurrence.	
6	Nardone *et al.*, 2005[5]	Back pain, numbness and weakness in lower limbs	1	T2–T6	None	Serum ACE=22 IU/l CSF ACE=4.8 mol/l/min	Noncaseating granulomas consistent with sarcoidosis	Laminectomy and biopsy of the spinal pathology	Methyl prednisolone, followed by daily dose of 60 mg prednisone for 7 days, then reduced to 20 mg daily	Follow-up MRI after 1 year did not show any abnormalities	
7	Weissman *et al.*, 1996[8]	Weakness, radicular pain and paresthesias with difficulty voiding	1	L1–S1	Para-tracheal adenopathy	Not done	L1-S1 noncaseat inggranulomas associated with typical and well-developed Langhans' giant cells without asteroid bodies. Abundant lymphocytes were present without anytrue lymphoid architecture	A multilevel bilateral lumbar laminectomy down to the sacrum with gross total resection of the epidural mass	Oral prednisone for a 4-month period and was gradually weaned off	At 7 months postoperatively the patient was asymptomatic. MRI revealed a well-decompressed dural tube without residual mass	
8	Our case	Progressive dysphagia, neck pain radiating to both upper limbs and cervical myelopathy	1	C2–C7	None	Normal	Well-demarcated granulomaconsists of epithelioid histiocytes and mature lymphocytes	A CT-guided biopsy was inconclusive so anterior approach at the level of C5–C6 extra vertebrally for open biopsy	Initially was started on dexamethasone and after diagnosis she was discharged with 60 mg prednisolone daily, with tapering of the dose after 8 weeks	The 3-month follow-up results of the clinical patient showed improvement in all her symptoms	

Although medical management is the standard of care for neurosarcoidosis, most patients initially undergo biopsy\ surgical decompression or debulking to obtain pathology [Table 1]. Medical management usually includes steroids (e.g., in some cases, require a prolonged course of steroid utilization) along with immunosuppressive medications(e.g., methotrexate).[3,4]

CONCLUSION

Epidural spinal neurosarcoidosis is very rare. Establishing a pathological diagnosis warrants cessation of steroids (i.e., if previously administered), and the performance of a biopsy\ or open surgical debulking. Once pathology has been established, these lesions can be typically managed with short-\long-term steroids and/or other medical regimens.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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