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CASE REPORT

C2 Primary leiomyoma in an immunocompetent woman: A case report and review of literature

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ABSTRACT

Clinical case report and review of the literature. This is the first case of primary leiomyoma in an immunocompetent woman without previous history of uterine leiomyoma being reported in the literature to the best of our knowledge.Leiomyoma, a type of smooth muscle cell tumor, involving the vertebra is extremely rare. There were very few primary leiomyoma in patients with AIDS or in the immune-suppressed patients.This 48-year-old female came with H/o neck pain, weakness and bladder retention. On examination, tone increased in all four limbs, power on the right side of the limbs 4/5, power on the left upper limb 0/5, lower limb 3/5, left plantar was up going, decreased sensation over the left second cervical vertebra (C2) dermatome and all modalities decreased below C2. X-ray and magnetic resonance imaging (MRI) of the cervical spine showed kyphosis of the cervical spine with destruction of the C2 vertebral body along with pathological fracture. The patient underwent decompression of the C2 lesion through the C2 right pedicle with occipito-C1-C3 lateral mass screws fixation. Lesion anterior to the cord was reached by a transpedicular approach and decompression was performed.The lesion was pinkish grey, firm and moderately vascular and was destroying the C2 vertebral body. The patient improved symptomatically in power in the left upper limb and lower limb over the next 1 week duration from 0/5 to 4+/5. Histopathology revealed primary leiomyoma. The patient was evaluated with ultrasound abdomen and contrast tomogram of the chest, abdomen and pelvis to rule out other possible lesions in the lung, intestines and uterus.We suggest that leiomyoma should be included in the differential diagnosis of destructive lytic lesions involving the C2 vertebra. Histopathological examination with immunohistochemistry is necessary for the definitive diagnosis. Treatment of choice is surgery with complete removal.

Key words: Acquired immunodeficiency syndrome, benign metastasizing leiomyoma, immunocompetent, primary leiomyoma

Introduction

Primary smooth-muscle tumors of the central nervous system (CNS) are exceedingly rare. We report a primary extradural leiomyoma involving the second cervical vertebra (C2). There are many published reports of benign metastasizing leiomyoma (BML),[1] but primary extradural leiomyoma is rarely reported.[2,3] Primary leiomyoma of the bone is more common in the facial bones, and is less common in the skull bones and vertebra.[4] Our report appears to be the first documentation of primary leiomyoma involving the C2 vertebra in an immunocompetent woman without an uterine leiomyoma. We also discuss the histogenesis and review the literature on smooth muscle tumors of the CNS.

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Case Report

This 48-year-old female came with insidious-onset gradually progressive, continuous, dull aching type of pain on the right side of the neck radiating to the vertex region for 3 months. H/o weakness of all the four limbs along with tingling and numbness (Left >Right) were noted for 1 week, which

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was associated with bladder retention for 3 days. Motor examination showed increased tone in all four limbs. Power on the right upper limb was 4/5, power on the left upper limb was 0/5, lower limbs bilaterally was 3/5, left plantar was up going, sensory examination revealed decreased sensation over the left C2 dermatome and all modalities decreased below C2. X-ray of the cervical spine showed osteolytic destruction of the C2 body with subluxation of the C1 and C2 complex over C3 [Figure 1]. Magnetic resonance imaging (MRI) of the cervical spine showed kyphosis of the cervical spine with destruction of the C2 vertebral body along with pathological fracture. MRI also showed that the C2 vertebral body was showing T1 hypointense, T2 hyperintense lesions with posterior displacement of the severely compressed spinal cord [Figure 2]. Differential diagnosis of the expansile C2 vertebral body in immunocompetent and immunodeficiency patients is enumerated in Table 1. Preoperatively, our presumptive diagnosis was C2 vertebral body tuberculosis. The patient underwent decompression of the C2 lesion through the C2 right pedicle with occipito-C1-C3 lateral mass screws fixation (vertex

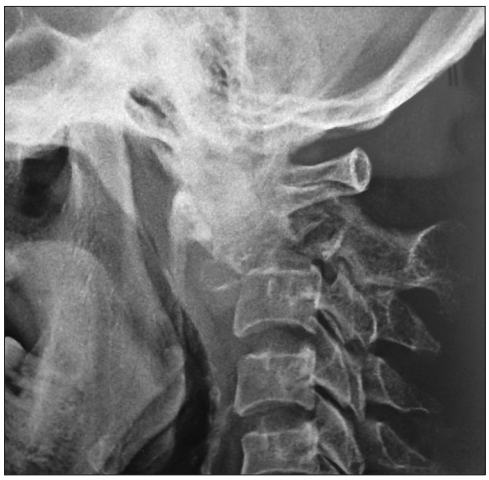

Figure 1: X-ray cervical spine showed osteolytic destruction of C2 body with subluxation of C1 and C2 complex over C3

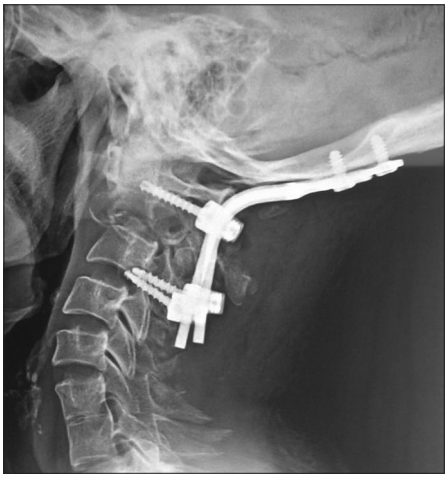

Figure 3: X-ray showing occipito-C1-C3 lateral mass screws fixation with vertex Medtronic system

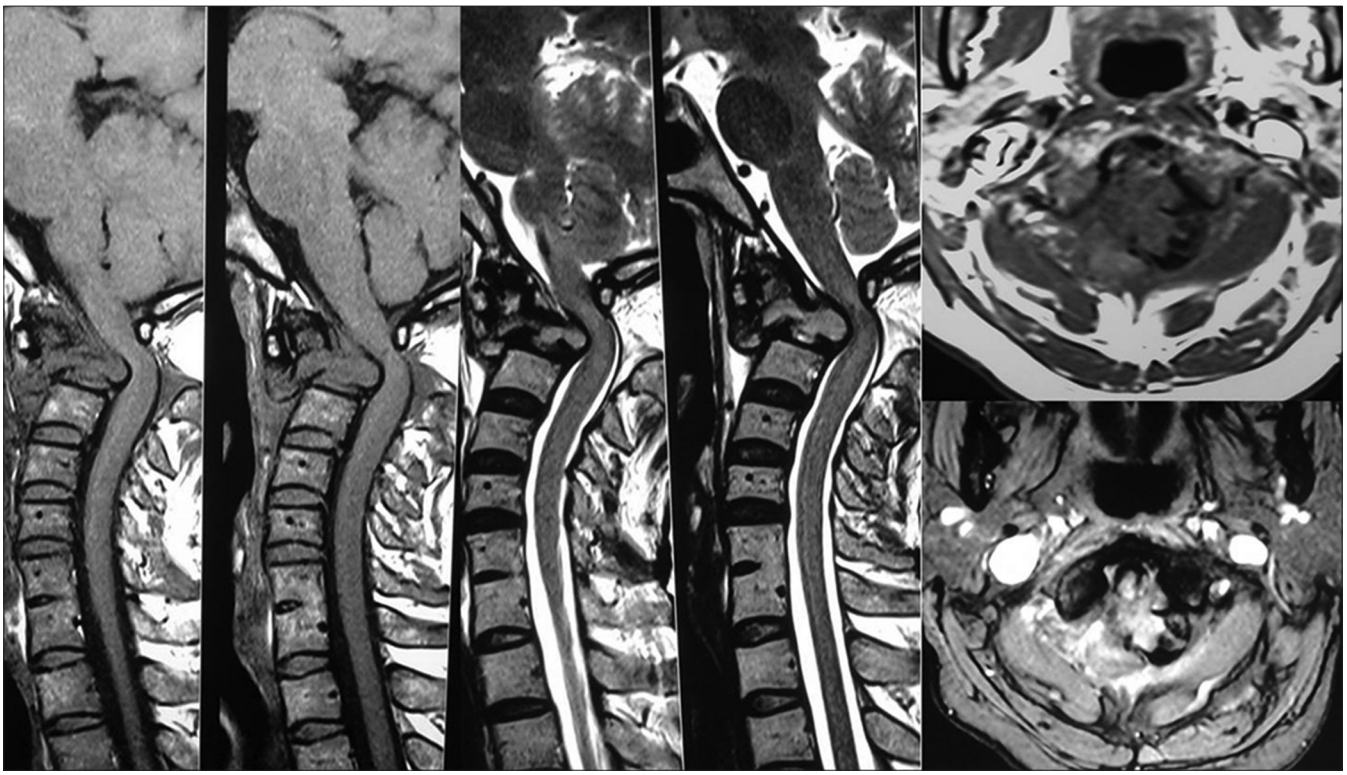

Figure 2: Magnetic resonance imaging cervical spine showed kyphosis of the cervical spine with destruction of C2 vertebral body along with posterior displacement of the severely compressed spinal cord

medtronic system) [Figure 3]. Lesion anterior to the cord was reached by a transpedicular approach and decompression was performed. The lesion was pinkish grey, firm and moderately vascular and was destroying the C2 vertebral body. Preoperative and intraoperative opinion was tuberculoma; therefore, safe maximal resection was performed instead of complete removal. The patient improved symptomatically in power in the left upper limb and lower limb over the next 1 week duration from 0/5 to 4+/5. Histopathological examination of the tumor tissue showed spindle cells arranged in whorls and fascicles with proliferation of smooth muscle cells surrounding the blood vessels. Spindle cells showed eosinophilic cytoplasm with elongated nuclei with blunt ends. There was moderate cellularity, minimal atypia, inconspicuous mitosis and no evidence of necrosis [Figure 4a and b]. The tumor cells stained positively for smooth muscle actin, desmin and vimentin and negative for epithelial membrane antigen, S-100, Cluster of differentiation 34, estrogen and progesterone receptors, confirming the diagnosis of the leiomyoma [Figure 4c and d] and excluding the other possible lesions like meningioma and

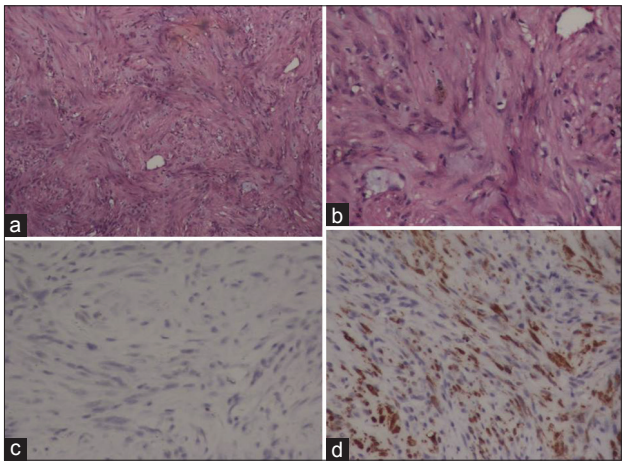

Figure 4: (a and b) Histopathological examination showed spindle cells arranged in whorls and fascicles with proliferation of smooth muscle cells surrounding the blood vessels. Spindle cells showed eosinophilic cytoplasm with elongated nuclei with blunt ends. There is moderate cellularity, minimal atypia, inconspicuous mitosis, and no evidence of necrosis; (c and d) Immunohistochemistry of the tumor cells stained positively for smooth muscle actin, and negative for S-100 protein

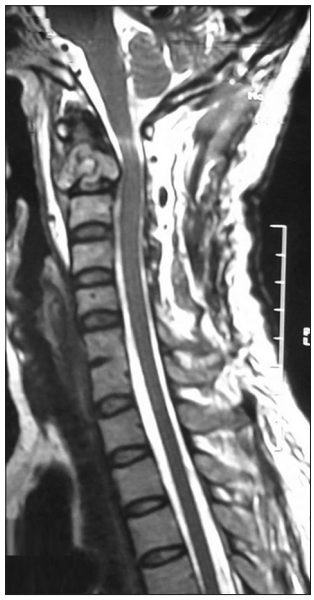

Figure 5: Post operative magnetic resonance imaging at 1 year showed small residual tumor at C2 body

Table 1: Differential diagnosis of the C2 vertebral body expansile lesion

Lesion	Age	Histology	Imaging
Hemangioma	any age; peak fourth decade	vascular spaces lined by endothelial cells	vertical parallel densities spotted appearance on CT high signal on T1W and T2W images; involvement of posterior elements
Langerhans cell histiocytosis	first, second decades	sheets of Langerhans cells, lymphocytes, and eosinophils	lytic lesion of the vertebral body leading to collapse
Aneurysmal bone cyst	young patients upto 20 years vertebral body 40%	cystic spaces containing blood products	lytic expansile lesion with fluid-filled levels
Osteosarcoma	Fourth decade	Osteoid within sarcomatous tissue	Osteosclerotic and osteolytic areas with soft tissue component
Chondrosarcoma	Fifth decade Predilection for vertebral body	Hyaline cartilage with increased cellularity within myxoid matrix	Bone destruction with characteristic punctuate calcifications
Plasmocytoma	>40 years old	Sheets of plasma cells on a delicate reticular stroma	Radiolucent areas or reduction in bone density hypointense on T1W and hyperintense on T2W images
Giant cell tumor	Third decade	Osteoclastic giant cells intermixed with spindle cells	Osteolytic geographic area with soft tissue component
Ewing's sarcoma	Second to third decades	Sheets of small round blue cells	Lytic lesion, associated soft tissue mass
Chordoma	Middle-aged patients Exclusively affects vertebral body; most often sacrum, rarely mobile spine	Lobulated mass with mucinous containing cells	Destructive midline expansile lesion with associated soft tissue mass; extension into adjacent vertebra
Tuberculosis and vertebral Osteomyelitis	Younger age with or without Immunosuppression	Giant cells, lymphocytes, epithelioid cells	T1 Hypo, T2 hyper and enhances with contrast along with periepidural soft tissue

CT – Computed tomography

schwannoma. The patient was evaluated with ultrasound abdomen and contrast tomogram of the chest, abdomen and pelvis to rule out other possible lesions in the lung, intestines and uterus. Second surgery (trans-oral) was planned based on histopathology for complete removal of the tumor, which was postponed because of drastic improvement in myelopathy features. Postoperative MRI at 1 year showed residual tumor without cord compression [Figure 5], and she was asked for regular follow-up for any recurrence of symptoms.

Discussion

Leiomyomas are benign smooth muscle tumors most commonly found in the gastrointestinal and genitourinary tracts and in the skin.[3] They are much less frequently found in the deep muscles of the extremities or within the abdominal cavity or retroperitoneum.[3] Rarely, leiomyomas have been found in the nasal cavity, peripheral nerve, mediastinum and lung, thyroid gland and liver.[3] Primary smooth muscle tumors of the CNS are exceedingly rare. There were 76 reports regarding the BML,[1] but reports of primary extradural leiomyoma are scarce.[2,3] Primary leiomyoma of the bone is common in the facial bones, and is very rarely reported in the skull bones and vertebra.[4] Primary lesions in CNS were identified in the sellar and parasellar areas.[5] Leiomyoma of the bone is extremely rare due to a paucity of smooth muscle cells in that location. The lesion has a female preponderance (1.7:1) It is presumed that smooth muscle tumors in the CNS may arise from embryonic rests, pluripotent mesenchymal cells, perivascular connective tissue, leptomeninges or smooth muscle cells of the blood vessels.[5]

There is an association between human immunodeficiency virus infection and smooth muscle tumors like leiomyoma, particularly in the pediatric age group and in unusual anatomical locations like the spine.[2] This was supported by other reports as smooth muscle tumors are rare in children, and it has been suggested that this association may not be random. Recently, the causality of Epstein-Barr virus is implicated in the etiology of the leiomyoma, especially in immunodeficiency states.[6] Some of these tumors are seen to occur in uncommon locations, further strengthening the association of immunodeficiency and the neoplasm.[3]

BML of the uterus is a rare, although well recognized, entity that was reported in the literature up to 76 cases. BML is common in women aged 35-55 years old, occurring after hysterectomy for leiomyoma.[1] The most common site of presentation is lung, although other sites involved are lymph nodes, heart, skull, spine and retroperitoneum.[1] Tumor metastasizes to lungs and other tissues via hematogenous spread.[1] Careful follow-up of BML patients is recommended because of its low-grade clinical malignant behavior.[1] Our case is not BML as there is no uterine leiomyoma or hysterectomy.

Leiomyoma usually presents with myelopathy as in our case due to compression over the cord or radiculopathy due to compression of the nerve root. On X-ray or computerized tomogram (CT) imaging, leiomyoma of the vertebral body presents with osteolytic lesion with destruction of the vertebral body. On MRI, the tumor appears T1 isointense to hypointense and T2 hyperintense and enhances with contrast, as in our case.[5] Presence of spindle cells with intense positivity for smooth muscle actin confirmed the diagnosis of leiomyoma.

The confusion in the diagnosis of the leiomyosarcoma and BML has been present for years; our tumor differs from leiomyosarcoma in view of absence of the mitoses, cellular atypia and necrosis. Therefore, it is suggested that the meticulous sampling of the pathology specimen, which indicates benign tendency, should not be easily diagnosed as leiomyosarcoma in spite of an aggressive course. The final diagnosis requires immunohistochemical staining and careful histological examination. The histological characteristics of this pathological entity differentiate it from other commonly occurring lesions like meningioma, hemangioma and metastasis. Thus, leiomyoma is one of the differential diagnoses for extradural lesions of the spine.[1] The present neoplasm, which was diagnosed in C2 vertebra, had disclosed little histological signs of malignant character.

Treatment of leiomyoma in immunodeficiency states is same as in traditional primary leiomyomas.[2] In case of the BML, they may be estrogen and progesterone receptor positive, and they have a tendency to recur after treatment.[1] Therefore, medical treatment with leuteinizing hormone releasing hormone analogues and tamoxifen, or surgical treatment with a bilateral oophorectomy, may be useful.[1] Shrinkage of the tumor after menopause and pregnancy, and even spontaneously, are reported with BML.[1] These tumors are benign but have the potential to metastasize; therefore, complete removal of the tumor is needed.

Conclusion

Primary leiomyoma in the spine is very rare, and all previous reports are in immune-deficient patients. This is the first case report of the leiomyoma in the immunocompetent patient to the best of our knowledge. We recommend this diagnosis in differential diagnosis whenever an osteolytic spine lesion is noted. The treatment of choice is surgery with complete removal.

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Conflicts of interest
There are no conflicts of interest.

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