

Case Report

Cellular myxoma of the lumbar spine

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Abstract

Background: Cellular myxoma is a histopathologically distinctive benign neoplasm, which has often been categorized among the broad category of benign mesenchymal tumors with myxoid stroma and fibroblast- and/or myofibroblast-like cells. These tumors can arise in any of the large muscles and are usually found in the thigh, shoulder, buttocks, and upper arm, and more rarely in the head and neck or in small muscles of the hand.

Case Description: Here we illustrate the case of a 57-year-old female with a spinal lesion, who initially presented with complaints of vague pelvic discomfort but no focal neurological deficits. Imaging revealed a sharply demarcated paraspinal lesion concerning for a tumorous growth. The lesion was excised *in toto* and a detailed immuno-histopathological analysis was performed revealing the diagnosis of a cellular myxoma. Postoperative imaging showed a gross total resection and the patient is under clinical surveillance since, with no signs of recurrence after 42 months.

Conclusion: Although very rare, this entity should be considered in the differential diagnosis of any spinal and paraspinal mass to allow for adequate treatment, which requires wide excision with clean margins to avoid any local recurrence.

Key Words: Cellular myxoma, intramuscular myxoma, lumbar myxoma, paraspinal myxoma

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Quick Response Code:**INTRODUCTION**

Among the various but scarce benign myxoid soft tissue tumors, intramuscular myxomas stand as one of the more common ones, with an incidence of 0.1 to 0.13 per 100,000.^[13]

Typically a tumor of adulthood,^[5] with virtually no reported examples in children and adolescents,^[7] it is characterized by abundant myxoid matrix, a poorly developed vasculature,^[1] and a small number of inconspicuous stellate- or spindle-shaped fibroblast-like

cells.^[20] Although most tumors appear to be well-circumscribed, it is not uncommon for this tumor to actually infiltrate the adjacent musculature and soft tissue. Occasionally, intramuscular myxomas show focal areas of hypercellularity and hypervascularity. However, these foci show a distinct lack of nuclear atypia, necrosis, and mitotic activity, and hence bear no prognostic significance. The current treatment of choice remains unclear, but so far most of the cases described underwent wide resection only, since local recurrence which is a rare event^[28] in this setting.

CASE REPORT

Here, we report a 57-year-old Caucasian female, with an unremarkable past medical history, who was referred to our neurosurgical department after she presented with an incidentally found lesion on magnetic resonance imaging (MRI) of her lower spine. On physical examination the patient did complain about discomfort in her pelvic area when sitting, left greater than right. However, she did not have any related neurologic complains such as back pain or leg pain, nor did she display any weakness, numbness or tingling sensation or other signs of radiculopathy. The mass was incidentally discovered on lumbar MRI, which was obtained for evaluation of her atypical pelvic pain.

Routine MRI was obtained demonstrating a right-sided sharply demarcated, poorly and heterogeneously contrast enhancing paraspinal lesion at the junction of the lumbar and sacral spine. The mass was localized immediately adjacent to the posterior elements (lamina and spinous process) and was deeply seeded in the right multifidus muscle adjacent to the L5 inferior articular process. It measured approximately $3.0 \times 3.0 \times 3.5$ cm in size on T1 postcontrast images [Figure 1]. Based on radiographic criteria alone, the differential diagnosis included: Schwannoma, lipoma or liposarcoma, fibroma, myoma, sarcoma or metastatic disease among rare other entities. A full systemic work up was performed and included a contrast enhanced computed tomography (CT) of the chest, abdomen, and pelvis as well as a mammogram and were negative for any other focus of systemic disease. As the lesion was concerning for a neoplasm, the patient was counseled and advised to pursue a histopathological diagnosis to direct further treatment options. She opted for

an open surgical approach aiming for an excisional biopsy and she was consented and taken electively to surgery.

Preoperative repeat MRI was performed within a 4 week window after initial presentation and revealed an unchanged right-sided paraspinal lesion at the level of the lower posterior lumbosacral spine. The lesion measured again approximately 3.5 cm in diameter and remained heterogeneously contrast enhancing and appeared somewhat lobulated. It continued to display sharp demarcation to the surrounding soft tissue. It did not show signs of erosion or invasion of the osseous parts of the L5 posterior elements.

Surgery was performed under general anesthesia. The patient was positioned prone on a Wilson frame and a standard midline incision was performed after application of local anesthesia. The subcutaneous layers were split and the thoracolumbar fascia was divided. Dissecting into the soft tissue, we immediately identified a well encapsulated lesion deep in the paraspinal muscles, but the mass was adherent to the periosteum of L5 and S1. An unremarkable dissection was performed and the lesion was resected *in toto* with a small soft tissue margin. Intraoperative fresh frozen analysis revealed a myxoid spindle cell tumor without significant atypia. Since the mass was abutting the osseous posterior elements of L5 and S1, we carefully coagulated the surface of the entire bony area that had been contacted to diminish the chances for any local recurrence. The wound was closed in layers and the patient had an unremarkable postoperative course. She was discharged to home the second postoperative day.

Pathology

The entire specimen with a small clean margin was received for pathological examination and measured

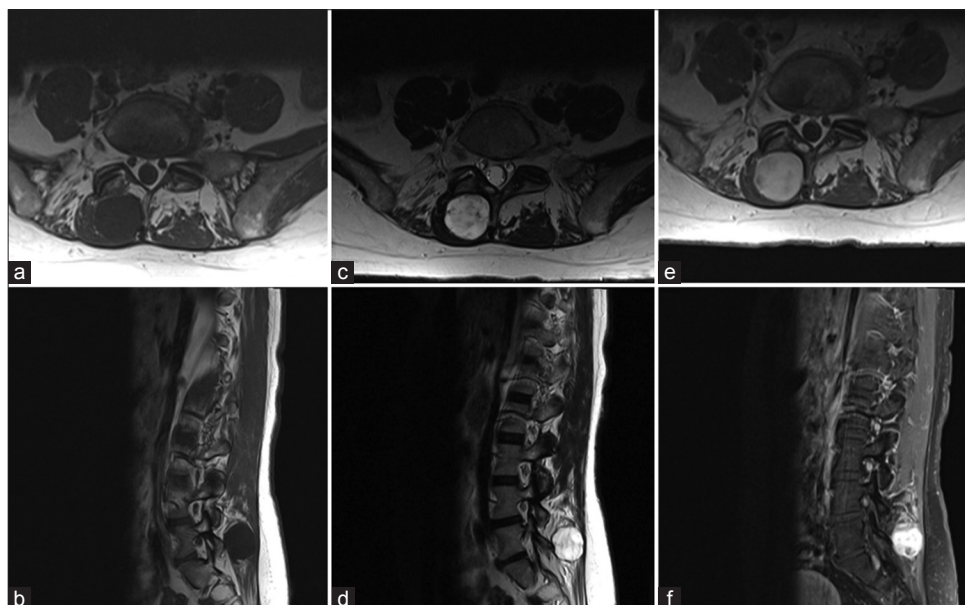


Figure 1: Pre-op MRI scan sequences with and without contrast: (a) axial T1 pre-contrast, (b) sagittal T1 pre-contrast, (c) axial T2, (d) sagittal T2, (e) T1 post-contrast, (f) sagittal post-contrast

ca. 5 cm. Cut surfaces revealed a thinly encapsulated, well-circumscribed lesion with a soft white, gelatinous appearing cut surface. The specimen was entirely submitted for histopathological examination. Routine hematoxylin and eosin stains were performed on formalin-fixed, paraffin-embedded sections and showed a relatively well-circumscribed tumor composed of prominent myxoid stroma with a distinct presence of stellate-shaped bland cells with pyknotic nuclei and virtual absence of blood vessels [Figure 2a]. In approximately 20% of the tumor specimen, areas of increased cellularity and vascularity were noted [Figure 2b]. However, no mitotic activity, necrosis, or increase in nuclear atypia was noted in these foci. The tumor cells stained negatively for S100, actin, desmin, EMA, and cytokeratin cocktail. Based on the histopathological and immunophenotypical profile, a diagnosis of cellular myxoma was made.

As recurrence is very uncommon for such tumors after complete excision, no other therapeutic intervention was recommended. Due to the rather rare diagnosis of

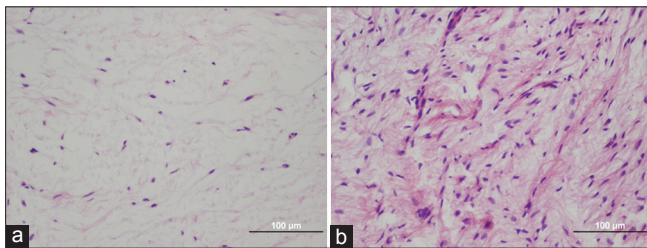


Figure 2: (a) In many areas, the tumor showed the classic appearance of intramuscular myxoma, characterized by a hypocellular tumor with prominent myxoid background, stellate-shaped cells and poorly developed vascularity. (b) In contrast, many foci of increased cellularity and vascularity (arrows) were identified. Note the lack of nuclear atypia, necrosis, and mitotic activity. (H and E, ×20 objective)

cellular myxoma in this locale – with overall dearth of evidence-based clinical recommendations – a close and frequent follow-up surveillance imaging schedule was established for this patient in spite of this entity being described as a benign tumor. To date, 42 months after the surgery, the patient remains neurologically intact, feels well and no recurrence has been observed on sequential imaging [Figure 3a-f].

DISCUSSION

Most intramuscular myxomas present as a solitary painless mass that typically occur in the large muscles of the thigh, shoulder, buttocks, and upper arm.^[1,3,4,18]

Clinical

Unusual sites include the muscles of the head and neck, the forearm, and small muscles of the hand.^[22] So far, only a few cases of classic intramuscular myxomas have been reported^[8,12,27] [Table 1] and to the best of our knowledge, only one other case of cellular myxoma has been reported in this paraspinal lumbar region (Falavigna *et al.*^[8]).

These tumors are usually observed in adults at about 40–70 years of age, with a female predominance.^[18] A history of trauma to the region is rarely given. A rare association with a variety of other pathologies has been described and includes Fibrous dysplasia (e.g., Aoki *et al.*^[2], 1995), Albright's Syndrome,^[17] and Mazabraud's Syndrome.^[11] The average size at the time of diagnosis varies greatly, the majority measuring between 5 and 10 cm. The tumor is usually well-circumscribed grossly, but infiltration into adjacent skeletal muscle and/or soft tissue is not uncommon.^[22] Characteristic MRI features include rather homogenous low signal intensity on T1-weighted MR sequences and high signal intensity

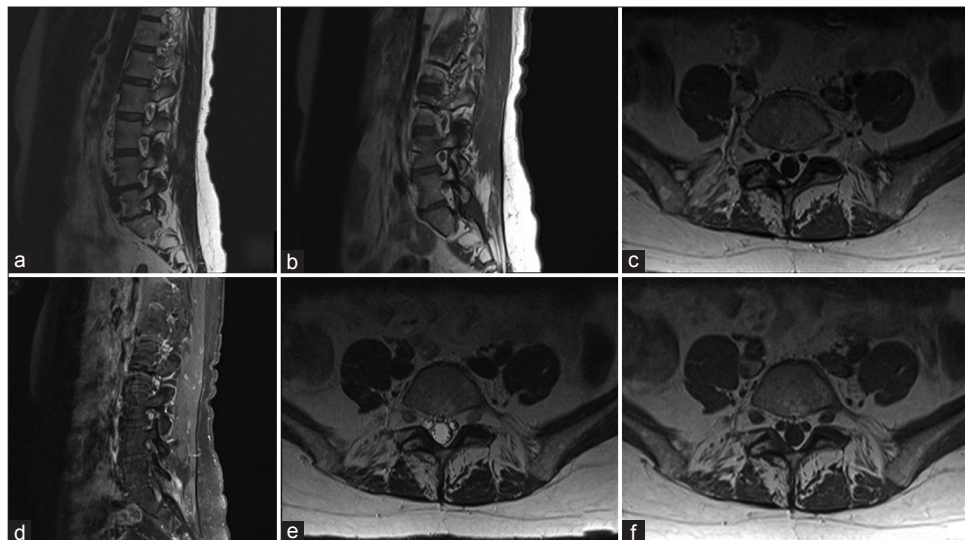


Figure 3: (a-f) MRI in T2 and T1 sequences in sagittal and axial planes with and without intravenous contrast application showing no tumor recurrence on subsequent films taken every 6 months

Table 1: Location of resected paraspinal intramuscular myxomas reported in literature

Case	Reference	Age/sex	Site	Size (cm)	Presentation	Follow-up
1	Liguomo ^[16]	63 F	L5 with 1 cm intracanalicular extension	8	Lower back pain and sciatica	2 years with no recurrence
2	Taggarse ^[26]	52 F	Thoracic spinal m.	12.5	Intermittent back pain	3 months with no recurrence
3	Falavigna ^[8]	64 F	Cervical paraspinal m. (C2-C5)	12	Occipital and neck pain	12 months with no recurrence
4	Guppy ^[12]	80 F	Lumbar paraspinal m. (L5-S1)	7	Low-back pain for over 20 years with development of L-5 radiculopathy over a 3-month period	3 months with no recurrence
5	Feldman ^[9]	62 F	Cervical paraspinal m.	3	-	-
6	Shugar ^[24]	68 F	Cervical paraspinal m.	4	No neurological symptoms	12 months with no recurrence
7	Crankson ^[7]	5 F	Cervical paraspinal m.	4	No neurological symptoms	12 months with no recurrence
8	Tahmouresie ^[27]	50 M	Thoracolumbar paraspinal m. (T11-12)	12	Severe back pain and progressive gait disturbance	-
9	Rashid ^[23]	70 F	Paravertebral m. (L2-S3)	15	Back pain, asymmetry of the waist and a mass in right paravertebral region	12 months with no recurrence
10	Stinchcombe ^[25]	80 F	Paravertebral m. (L2 to S1)	-	Lower back pain, no focal neurological deficits	-
11	Kamoun ^[15]	54 F	Paravertebral m. (L5-S1)	-	Claudication, lumbar weakness and stiffness	3 years with no recurrence
12	Ishoo ^[14]	1.10 F	Cervical paraspinal m.	4.9	Decreased cervical mobility	2 years with no recurrence
13	Ohla	57 F	Lumbar paraspinal m.	3.5	No related neurological symptoms	3.5 years with no recurrence

on T2-weighted or fluid sensitive MR sequences. The majority of lesions display well-defined borders and a peritumoral fat ring. Heterogeneous contrast enhancement was observed in many of the studied lesions (Bancroft *et al.*^[4]; Nishimoto *et al.*^[19], 2004).

On histological examination, a unique hypocellular tumor with striking myxoid background and almost absent vasculature is very characteristic. The tumor lacks nuclear atypia, necrosis, and prominent mitotic activity.^[1,20,21]

Differential diagnosis include low-grade myxofibrosarcoma, low-grade fibromyxoid sarcoma, myxoid liposarcoma, and myxoid neurotheomas.^[6,20] The absence of nuclear hyperchromasia, prominent curvilinear vasculature, perivascular tumoral condensation, collagenous matrix, well-formed lipoblasts, and well-developed lobules with characteristic fibrous bands and S100-positivity are used to exclude these other considerations.^[10,12]

Intramuscular myxoma is a benign lesion, and surgery is considered curative, as seems to be the case with our patient, with rare reported cases of local recurrence. Although part of the general “myxoid” family of tumors, these lesions are not associated with the Carney complex,^[1] classically characterized by a triad of cutaneous and cardiac myxomas, spotty pigmentation, and endocrine hyperactivity.

CONCLUSION

Although cellular myxomas are only rarely encountered

in the practice of neurosurgery, neurosurgeons should be aware of this entity. Despite the fact that the increase in cellularity may raise concerns about potential aggressiveness of such lesions, previous cases show that complete surgical excision of these lesions is apparently curative and should be seen as the treatment of choice until longer follow up data are available examining outcomes of a larger cohort.

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