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The Spine Journal 15 (2015) e1–e4

THE SPINE JOURNAL

Case Report

Infiltrative intramuscular myxoma of the cervical spine: a case report

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Received 1 April 2014; revised 28 July 2014; accepted 15 September 2014

Abstract

BACKGROUND CONTEXT: Myxomas are benign tumors of mesenchymal cell origin that usually present as solitary lesions. They are infrequently associated with fibrous dysplasia, as in McCune-Albright or Mazabraud syndrome. Myxomas can develop in a variety of locations, although the most frequent sites are the thigh, buttocks, shoulder, and upper arm. Intramuscular myxomas (IMs) refer to lesions that occur within muscle compartments. They have been infrequently reported in the neck musculature.

PURPOSE: To date, only five cases have been reported within the posterior neck muscles without associated intraspinal extension. To our knowledge, this is the first case of an IM presenting with extension into the spinal canal.

STUDY DESIGN: We report a case of posterior cervical IM with intraspinal extension presenting in a 63-year-old woman as a palpable mass.

METHODS: Complete intralesional resection of the tumor was achieved by standard midline posterior approach. Meticulous resection of the entire capsule was achieved and all margins were confirmed to be free of neoplasm.

RESULTS: A diagnosis of myxoma was provided on pathologic evaluation. Follow-up at 1.5 years confirmed maintained complete resolution of the preoperative symptoms, with no evidence of local recurrence on imaging.

CONCLUSIONS: Intramuscular myxomas should be included in the differential diagnosis of cervical paraspinal tumors. Furthermore, we suggest that masses involving the axial muscles should be closely monitored and the patient counseled regarding potential neurologic sequelae.

Keywords: Intramuscular myxoma; Cervical spine myxoma; Infiltrative myxoma; Spinal tumor; Cord compression; Lytic spine lesion

Introduction

Although myxomas in general are common and have been known to occur in a variety of locations [1,2], intramuscular myxomas (IMs) are infrequently reported in the scientific literature, with an incidence of one in 1 million population per year [3]. Intramuscular myxomas affecting the cervical paraspinal musculature are rare [4], and despite an extensive search of the literature, we could not find any reports of extension into the spinal canal. Most IMs are slow-growing but nevertheless can be infiltrative, requiring prompt intervention to minimize potential morbidity.

Case report

A 63-year-old woman presented with complaints of neck and radicular right arm pain, with associated numbness and tingling, starting 2 years before presentation. Over time, her pain improved except for a palpable,

FDA device/drug status: Not applicable.

Author disclosures: ***SRRM:*** Nothing to disclose. ***ABS:*** Nothing to disclose. ***CAA:*** Nothing to disclose. ***HFF:*** Speaking and/or Teaching Arrangements: Depuy Synthes (B).

The disclosure key can be found on the Table of Contents and at www.TheSpineJournalOnline.com.

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http://dx.doi.org/10.1016/j.spinee.2014.09.008
1529-9430/

mostly nontender mass over the posterior aspect of her neck on the right. She remained neurologically intact. X-ray imaging showed a vague lucency involving the posterior elements at the C5 level, which was confirmed in the computed tomography imaging to be extending into the right pedicle (Fig. 1). Magnetic resonance imaging reveals an enhancing lesion within the right paraspinal soft tissues at the level of C4–C5 measuring 27×16×38 mm, involving the right C5 pedicle and hemilamina, and causing mild mass effect on the thecal sac (Fig. 2). Magnetic resonance imaging angiography confirmed that the vertebral artery was not involved. At 1.5 years after gross total resection of the tumor, the patient was symptom-free with return to normal activities. Follow-up contrast-enhanced computed tomography and magnetic resonance imaging of the cervical spine showed no evidence of tumor recurrence (Fig. 3).

Surgical technique

The patient was positioned prone with midline exposure from C3 to C6. On the right side, the plane surrounding the tumor was carefully identified starting superficially, then extending mediolaterally, and finally rostrocaudally. The tumor itself appeared to be primarily well circumscribed, encapsulated, and firm. Given this overall benign intraoperative appearance, we elected to proceed with an attempt at gross total resection of the tumor. On reaching the lateral aspect of the tumor, large feeding vessels were identified, bipolared, and divided. The extradural portion was resected meticulously by removing the remaining portion attached to the C4–C5 right-sided facet joint. Further inspection revealed that the C4 and C5 right hemilaminae were grossly resorbed and infiltrated with tumor and that there was soft residual epidural tumor remaining. Right-sided C4 and C5 hemilaminectomies were fashioned and the epidural component was dissected free. Finally, residual tumor within the right-sided C4–C5 facet joint was scraped off as well to ensure a gross total intralesional resection.

Pathology

At low power, the lesion was paucicellular with a prominent myxoid background (Fig. 4, Left). At highest power, cytologically bland fibroblasts are seen suspended within the myxoid matrix among scattered eosinophilic collagen fibers (Fig. 4, Right). The lesional cells also displayed reactivity with vimentin, CD34 (focal), and S-100 protein (data not shown).

Discussion

Although very infrequently reported in the neck muscles, myxomas have been known to man since Rudolf Virchow [1,4] coined the term in the eighteenth century to describe a benign tumor that resembled mucinous umbilical cord tissue. These tumors may occur in various locations. Intramuscular myxomas were described as a distinct subtype of myxoma in 1965 by Enzinger [5], and comprised only 17% of all soft-tissue myxomas in this series. The majority of IMs present in the elderly female population [3,5]. They have been reported in the neck region in children as young as 22 months. The incidence appears to follow a bimodal distribution with a peak in the first to second decades and then again from the fifth to seventh decades. A majority of IMs arise in the extremities, particularly the thigh muscles [6]. Intramuscular myxomas rarely occur in the neck, with only four cases reported to date in the cervical paraspinal muscles [1,4,7,8]. The clinical manifestations of neck IMs are nonspecific, and they remain difficult to diagnose without pathologic evaluation.

Intramuscular myxomas typically present as slow-growing painless masses that may produce symptoms because of compression of surrounding structures [1,7,8]. In pediatric patients, the time to presentation is often within a year, whereas adults tend to present after several years. It remains unclear if this relates to differential growth rates in these age groups. Only approximately 20% are painful [5].

In the case described, the pain evolved for over 2 years and the patient noted an enlarging mass over the

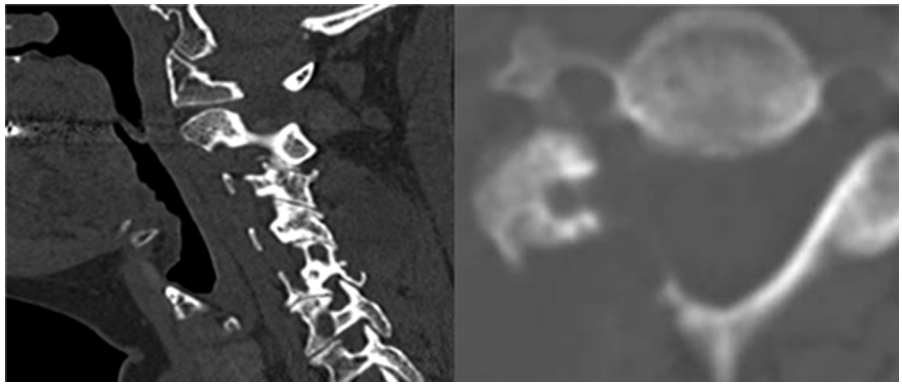

Fig. 1. Computed tomography imaging (sagittal and axial views) reveals a lytic defect over the lamina of C5 and involving the right pedicle of C5.

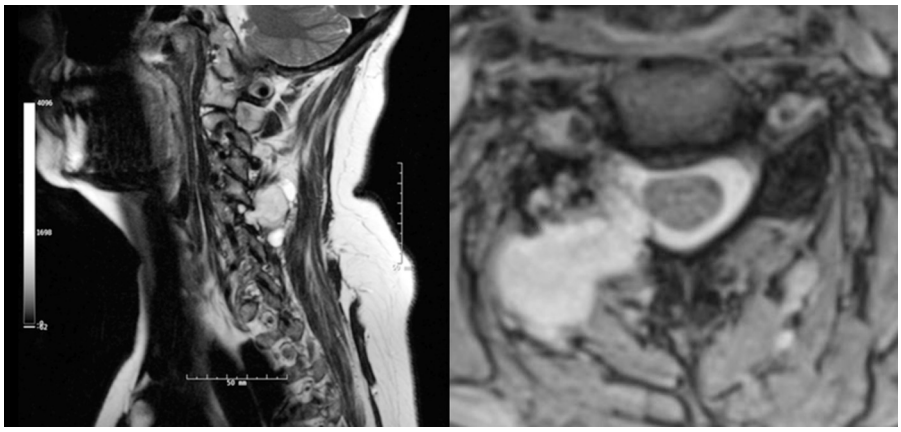

Fig. 2. Magnetic resonance imaging (sagittal and axial views) revealing a 27×16×38 mm enhancing lesion at C4–C5, involving the right paraspinal soft tissues with extension into the spinal canal.

preceding year. Although this clinical presentation was consistent with those of previously reported neck IMs [1,4,8], and suggestive of a benign course, the tumor in this case was infiltrative, resulting in erosion of the bony laminae, epidural extension, and cord compression. These features are atypical and appear unique to this case. The imaging findings of IMs are otherwise nonspecific with respect to other fluid-filled soft-tissue masses. Radiologic differential diagnosis for IMs includes fluid-filled collections such as cystic hygroma, cystic teratoma, lipoma, hematoma, and abscess [4]. Histologic differential diagnosis includes juxtaarticular myxoma, myxofibrosarcoma, and low-grade fibromyxoid sarcoma. Juxtaarticular myxomas are more fibroblastic and cystic and less homogenous as compared with IMs. Myxofibrosarcomas and low-grade fibromyxoid sarcomas are distinguished by pleomorphism, atypical cells, and signet ring lipoblasts [6].

The gross appearance of myxomas is typically grey-white or white depending on the relative amounts of collagen and myxoid material. Although usually grossly circumscribed, microscopic infiltration into the surrounding soft tissues is not uncommon. The lesion consists of lobules of a myxoid matrix rich in glycosaminoglycans and hyaluronic acid. Cyst-like spaces with abundant pools of myxoid material are common and the scattered, bland fibroblasts are intermixed with variable collagen fibers. No prominent or characteristic vascular pattern defines this lesion and features of malignancy are consistently absent. Although these lesions are characteristically vimentin reactive and nonreactive for desmin, S-100 protein, and CD34, our case displayed focal S-100 protein and CD34 immunolabeling of unclear significance [3,6].

Surgical excision is the treatment of choice for IMs. Initial needle biopsy or open biopsy should be considered in case of any suspicion for malignancy. Although there are no reports of metastasis or malignant change, myxomas have been reported to recur when the surgical procedure has exclusively involved enucleation or incomplete resection [9,10]. Most authors, therefore, recommend complete resection with a normal tissue margin to minimize the chance of recurrence. In this case, gross total intralesional tumor removal was achieved with no recurrence evident at 1.5 years of follow-up.

In conclusion, IMs are infrequent in the neck. Although typically benign, this case underlines the fact that IMs can be infiltrative. Given the proximity to critical structures including the spinal cord, timely intervention for cervical paraspinal IMs appears warranted to minimize potential neurologic complications. Complete resection in these

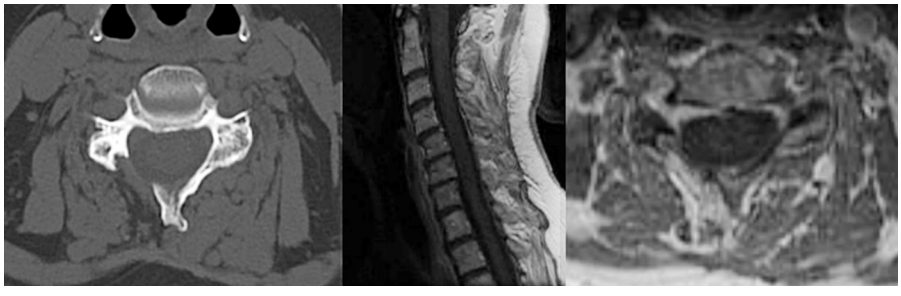

Fig. 3. (Left) Contrast-enhanced cervical computed tomography at 18 months follow-up (axial view) does not show any evidence of tumor recurrence. (Middle and Right) Contrast-enhanced cervical magnetic resonance imaging at 18 months follow-up (sagittal and axial views) does not show any evidence of tumor recurrence.

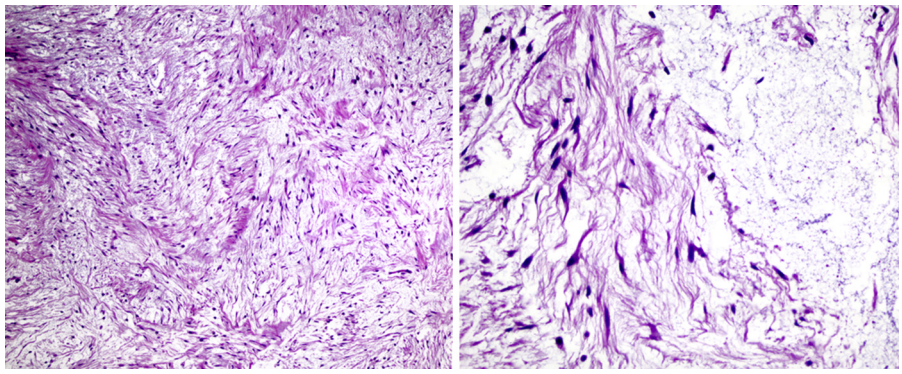

Fig. 4. (Left) At low power, the lesion was paucicellular with a prominent myxoid background. (Right) On highest power, cytologically bland fibroblasts were seen suspended within the myxoid matrix among scattered eosinophilic collagen fibers. No features of malignancy were seen; no mitoses, necrosis, or pleomorphism was identified.

cases also will minimize the chance of recurrence and the need for further surgical intervention.

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