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Intramuscular myxoma of the paraspinal muscles: A case report and systematic review of the literature

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Abstract. Intramuscular myxoma (IM) is a rare mesenchymal tumor of the head and neck region. The current study reports a case of a 45-year-old man who presented with a painless neck mass. Imaging showed involvement of the levator scapulae and scalene muscles. Core needle biopsy was consistent with intramuscular myxoma. Surgical excision was performed and follow-up for 30 months showed no recurrence. The present study includes a systematic review of head and neck IMs, with a summary of the clinical and demographic parameters of all reported cases in the head and neck region. Surgery was curative in 28 of the 29 published cases, as well as in the current case (96.7%), with the lone recurrent tumor cured following re-resection. Females constituted 57% of the cases and the mean age was 49.7±20.4 years. Although uncommon, IM should be considered in the differential diagnosis of deep neck masses, and surgical excision is the treatment of choice with a low risk of recurrence.

Introduction

In 1863, Virchow first described myxoma as a tumor anatomically resembling the umbilical cord (1). Myxomas (from the Greek word 'muxa' meaning mucus) are rare, benign connective tissue tumors arising from stellate mesenchymal cells (2), comprising entities such as fibromyxoma, cardiac myxoma and intramuscular myxoma (IM).

IM is an uncommon variant of the disease that typically presents between the fourth and seventh decades, with a slight female predilection (3). The majority of IMs present as slow-growing, painless masses within the thigh muscles and lower limb girdle (4,5). By contrast, IMs are rarely found in the head and neck region (4,5).

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Histopathologically, the lesions are usually recognized by their paucicellularity and minimal vascularity. Similar to other myxomas, IMs consist of fibroblasts and an abundant myxoid stroma (2), primarily composed of glycosaminoglycans and fibrous structural proteins (6). However, cases of IM displaying hypercellularity and abundant vascularity have been reported, often incorrectly leading towards a diagnosis of myxoid sarcoma (7). IMs are characterized by small stellate or spindle cells without features of atypia, mitosis and necrosis (8). Tumor cells possess small, hyperchromatic nuclei and inconspicuous cytoplasm. Immunohistochemically, they generally stain positively for vimentin and cluster of differentiation 34 (9). Mutations activating Gs (α) have been suggested to show correlation with this disease process (10).

Imaging modalities, including magnetic resonance imaging (MRI), computed tomography (CT) and ultrasonography, are useful for diagnosis, but the definitive diagnosis is histopathological. IMs display low signal intensity on T1-weighted MRI images and high intensity on T2-weighted images, with peripheral or patchy enhancement following gadolinium injection (3). CT scan evaluation typically reveals a hypodense mass in comparison to adjacent musculature, without contrast enhancement (3). Consistent with CT and MRI, ultrasonography reveals a hypoechoic lesion with a partial or complete capsule (3).

Other conditions that should be considered in the differential diagnosis include aggressive angiomyxoma, myxoid neurofibroma, low-grade fibromyxoid sarcoma, myxoid liposarcoma, low grade myxofibrosarcoma, cellular myxoma, juxta-articular myxoma and nodular fasciitis (11). IMs are located entirely inside the skeletal muscle, in contrast to myxoid liposarcomas, which are intermuscular.

Histopathologically, the absence of vascularity decreases the likelihood of sarcoma, and S-100 protein negativity excludes myxoid neurofibromas and low-grade malignant peripheral nerve sheath tumors (11). Once the diagnosis of IM is confirmed through biopsy, the treatment of choice is surgical excision (5,7).

The current study reports a new case of IM involving the levator scapulae and scalene muscles, and presents a systematic review of head and neck IMs, with a summary of the clinical and demographic parameters of all reported cases in the head and neck region.

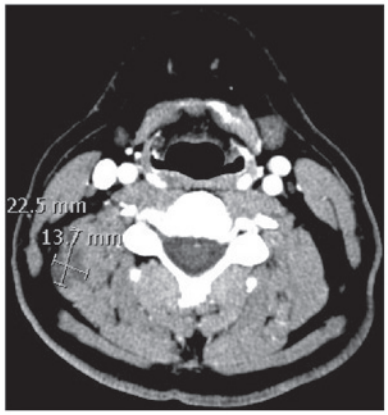

Figure 1. Cross-section contrast computed tomography scan showing the tumor in the right medial scalene and levator scapulae muscles.

Case report

Presentation. A 45-year-old, otherwise healthy, male presented to the Medical University of South Carolina (Charleston, USA) in March 2013 with a painless mass in the posterior of the neck that had been noticed by the patient 2 months earlier. The patient exhibited no sensory impairment, numbness or weakness of the right extremities. Physical examination revealed a deep, fixed, non-tender mass, with ill-defined borders.

Diagnosis. Ultrasound imaging showed an ill-defined, hypoechoic irregularity of a deep muscle of the right posterior neck, which corresponded to a hypodense lesion within the levator scapulae muscle on a contrast CT scan (Fig. 1). There was no significant internal flow on Doppler imaging and no lymphadenopathy. MRI revealed a 2.7x2.5x1.4-cm mass within the right superficial paraspinal musculature, likely involving the levator scapulae and scalene muscles. Pathological examination of a core needle biopsy showed spindle cells with a bland appearance, in a hypovascular, myxoid stroma, confirming the diagnosis of IM (Fig. 2).

Surgical excision. Dissection was performed down to the sternocleidomastoid muscle, which was mobilized along the posterior border. The spinal accessory nerve was identified and preserved. Electromyography was used to monitor the brachial plexus and the spinal accessory nerve. Erb's point was identified and the greater auricular nerve was preserved. Level 5 dissection of the lymph nodes was undertaken, preserving cranial nerve XI, to provide access to the tumor. The mass was palpated deep to these lymph nodes, and was located within the levator scapulae muscle, extending medially to involve the posterior and middle scalene muscles. Dissection was performed around the tumor, taking an ~1-cm cuff of muscle circumferentially around the tumor. The lesion was fully resected (Fig. 3) with tumor-free borders. Following 30 months of follow-up, no further treatment was needed and the tumor did not recur.

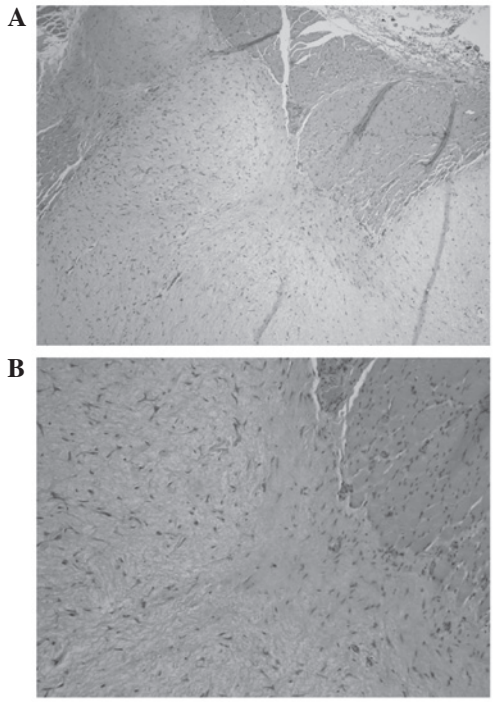

Figure 2. Hematoxylin and eosin staining of the intramuscular myxoma. (A) Paucicellular, hypovascular lesion with myxoid appearance (x10 magnification). (B) Small, spindle- or stellate-shaped tumor cells (x100 magnification).

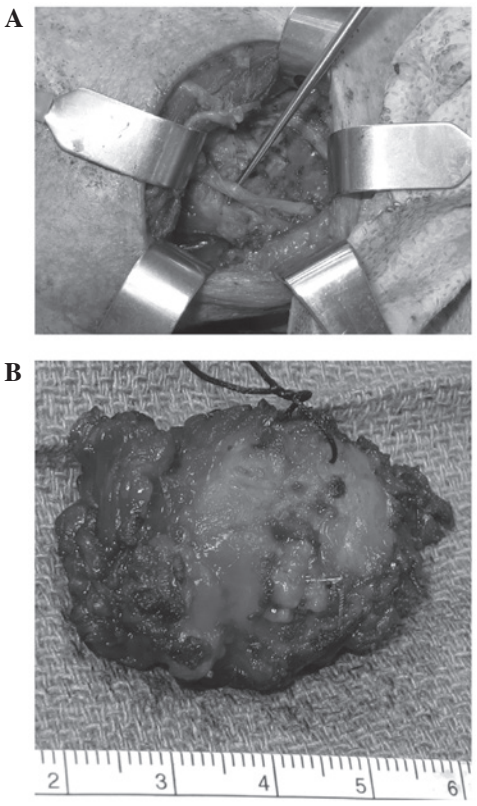

Figure 3. Gross tumor pathology. (A) Tumor prior to excision, situated deep to the spinal accessory nerve. (B) Excised tumor measuring 4 cm in length.

Literature review

Methods

Pubmed search. A comprehensive literature review of the literature was performed by searching the Pubmed-National Center for Biotechnology Information database, using the keyword search 'intramuscular AND myxoma'. The search yielded 158 studies published prior to December 2014, 15 of which were excluded, as

Table I. Summary of the head and neck intramuscular myxomas reported in the literature.

Reference no.	Gender	Age, years	Ethnicity	Anatomical location	Treatment	FU, years	Recurrence status	Size on PE, cm	Size on CT /MRI, cm	Size after excision, cm
(12)	F	57	NA	Paraspinal muscles	Surgical excision	NA	NA	NA	2	NA
(13)	F	63	NA	Parasipnal muscles	Surgical excision	1.5	No recurrence	NA	27x16x38	NA
(14)	M	74	NA	Hyoglossus	Surgical excision	3	No recurrence	8	8	NA
(15)	M	51	NA	Temporalis	Surgical excision	0.5	No recurrence	>5	NA	6.5x4x3
(26)	F	70	NA	SCM	Surgical excision	5	No recurrence	2	2x1.2x1.6	NA
(27)	F	45	Asian	Trapezius	Surgical excision	1	No recurrence	2.5x3	4.1x2.8x4.9	3.5x2.5x2
(28)	M	52	NA	Nasal vestibule (mimetic muscle)	Surgical excision	1	No recurrence	NA	2x1.3	2.5
(19)	F	64	NA	Paraspinal muscles	Surgical excision	1	No recurrence	12	15	NA
(20)	M	74	NA	Masseter	Surgical excision	2	No recurrence	2x3	NA	NA
(21)	F	2	NA	Trapezius and paraspinal muscles	Surgical excision	2	No recurrence	4x5	4.1x2.8x4.9	NA
(22)	M	22	NA	Scalene and SCM	Surgical excision	4	No recurrence	6x4	7x3	7x4x3
(23)	F	43	B	Temporalis	Surgical excision	1.5	No recurrence	3.5x2.5	NA	3.5x2x2.6
(24)	F	5	NA	Deep to trapezius	Surgical excision	1	No recurrence	4	4	NA
(25)	F	60	NA	Posterior scapular muscles	No intervention (monitor only)	1	No change in size by MRI	Four deep nodules	NA	NA
(25)	M	56	NA	Right cheek	Surgical excision	4	No recurrence	NA	3	NA
(26)	M	62	NA	Temporalis	Surgical excision	1	No recurrence	5x4	NA	NA
(27)	M	46	NA	Orbicularis oris	Surgical excision	2	Recurrence in 5 months, then no recurrence	3	NA	NA
(28)	F	60	NA	Tongue	Surgical excision	NA	NA	2	NA	NA
(29)	F	69	B	Levator scapula	Surgical excision	1	No recurrence	4x3	4	NA
(30)	F	43	NA	Masseter	Surgical excision	5	No recurrence	NA	NA	2x1
(31)	F	16	NA	Intermediary tendons of the digastric muscles, bilaterally	Surgical excision	1	No recurrence	6x1.5	NA	1.5 each
(32)	M	51	NA	Posterior neck (recurrent after previous excision)	Surgical excision	16	No recurrence	NA	NA	NA
(33)	F	79	NA	Masseter	Surgical excision	NA	NA	1	NA	NA
(34)	F	62	W	Posterior neck	Surgical excision	10	No recurrence	NA	NA	3
(35)	F	46	NA	Lateral neck	Surgical excision	2	No recurrence	NA	NA	NA

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Table I. Continued.

Reference no.	Gender	Age, years	Ethnicity	Anatomical location	Treatment	FU, years	Recurrence status	Size on PE, cm	Size on CT /MRI, cm	Size after excision, cm
(36)	F	42	NA	Forehead	Surgical excision	12	No recurrence	NA	NA	NA
(37)	M	44	NA	Geniohyoid	Surgical excision	1.5	No recurrence	2	NA	NA
(38)	M	15	W	Cheek	Surgical excision	5	No recurrence	NA	NA	2x1.5
(39)	M	74	NA	Cheek muscles	Surgical excision	NA	NA	NA	NA	NA
Present study	M	45	W	Levator scapulae and scalene	Surgical excision	1	No recurrence	NA	2.7x2.5x1.4	4

F, female; M, male; W, white; B, black; NA, not available; FU, follow-up; PE, physical examination; CT, computed tomography; MRI, magnetic resonance imaging; SCM, sternocleidomastoid.

Figure 4. Outline of the case selection method used in the systematic review. The keyword search 'intramuscular AND myxoma' was performed in Pubmed database and English studies reporting cases in the head and neck region were included in Table I.

they were in a language other than English, leaving 143 studies. Of these, 28 included cases in the head and neck region, and were included in the present literature review. The inclusion criteria encompassed all studies with IM cases of the head and neck region that were published prior to December 2014. The exclusion criteria were as follows: i) Reports published in a non-English language; and ii) cases that had been already published in another study (i.e., duplicated cases). This yielded 28 studies (12-39), with 29 cases, in addition to the currently presented case. Fig. 4 outlines the case selection method.

Statistical analysis. Using Excel software (Microsoft Corporation, Redmond, WA, USA), two-tailed Student's t-test for independent samples was performed to compare the ages of the two genders. Values are reported as mean ± standard deviation (SD). $P<0.05$ was used to indicate a statistically significant difference.

Results. A total of 28 studies were included in this review, constituting 29 cases of IM in the head and neck region, in addition to the currently presented case (n=30; Table I).

The cases consisted of 43.3% males (n=13) and 56.7% females (n=17), with an age range of 2-79 years and a mean age (mean ± SD) of 49.7±20.4 years (males, 51.2±18.2 years; females, 48.6±22.4 years; P=0.73). The most common head and neck site was the paraspinal muscles, followed by the trapezius, masseter, cheek and temporal muscles (n=3 each).

The size of the mass on physical examination was available for 17 cases, with a length range of 2-12 cm and a mean length of 4.4±2.7 cm. Of all the cases, 96.7% (29 of 30) underwent surgery as the treatment of choice, with a recurrence rate of 3.3% (n=1). One case was monitored only and no change in size was observed upon MRI at 1 year post-diagnosis. The mean follow-up time for all patients was 3.3±3.8 years.

Discussion

IM is a benign tumor that commonly affects the skeletal muscles of the thigh (4). IM of the neck paraspinal muscles

is extremely rare. Although non-invasive and non-metastatic, local impingement of adjacent muscles, nerves or arteries could result in significant functional impairments.

IM could present as part of Mazbraud's syndrome, a rare disease displaying one or more IMs with fibrous dysplasia in one or more bones (40). Therefore, patients presenting with IMs should be examined for bone lesions. The number of reported cases of this syndrome in 2004 was 55 (41).

IM is the most common form of myxoma after myocardial myxoma. In addition to IM, soft-tissue myxomas include juxta-articular myxoma, superficial angiomyxoma, aggressive angiomyxoma and nerve sheath myxoma (42). The incidence of IM is ~1 per million individuals (4,43). In descending order, IMs most commonly arise in the thighs, shoulders, buttocks and upper arms. Other organs reported in the literature include the hands, face, tongue and abdominal muscles.

We recommend imaging of deep neck masses, and when surgical resection is performed, consideration of the proximity to the phrenic nerve and brachial plexus is important.

In summary, the present study reports a case of IM of the paraspinal muscles in a 45-year-old man. Following radiographic imaging with ultrasound, CT scan and MRI, a core needle biopsy confirmed the diagnosis. Surgical excision was performed and follow-up for 30 months demonstrated no recurrence. IMs should be considered in the differential diagnosis of deep neck masses. Surgical excision has shown to be curative in the vast majority of cases, with minimal recurrence rates.

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