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Intramuscular myxoma of the cervical paraspinal musculature: case report and review of the literature ☆

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ABSTRACT

Case: A 57 year old female patient presented with a painful right-sided neck mass that on MRI was shown to be adherent to the posterior aspect of the sternocleidomastoid muscle. The mass was surgically resected en bloc without complications. Histopathologic analysis revealed the mass to be a myxoma.

Review: To date, there have been several case reports of myxomas, although very few involving the head or neck. The majority of the literature available concurs that myxomas are benign neoplasms that exhibit characteristic qualities on MRI imaging. The definitive treatment by consensus is surgical excision.

Conclusions: Although the incidence of head and neck myxomas is low, it is important to include in the differential of a neck mass with certain radiographic findings.

1. Introduction

The term "myxoma" was originally utilized by Virchow in 1871 in his descriptions of tumors histologically resembling tissue of the umbilical cord [1]. Stout further clarified the term by establishing criteria for the classification of tumors as myxomas: a mesenchymal non-metastatic neoplasm composed solely of undifferentiated stellate cells, surrounded by a loose mucoid stroma [2]. Myxomas are benign, rare, locally invasive tumors of connective tissue that may arise in both somatic soft tissues and bone. In the head and neck, myxomas are most commonly found in the maxilla, mandible and soft tissues of the face. The subtype of intramuscular myxoma, that is, myxomas arising from skeletal muscle, was first described by Enzinger in 1965 [3]. There have been fewer than 200 reported cases of myxoma of the head or neck since the establishment of diagnostic criteria in 1948. Fifteen of these have been noted to be intramuscular, with four arising from cervical paraspinal musculature. A rare case of an intramuscular myxoma of the cervical paraspinal musculature and a comprehensive review of the literature to date are presented.

2. Case

A 57 year old female patient presented with a painful lump on the right side of her neck that had developed over 3 months. She stated that she had a tingling sensation that radiated up to the top of her head when this lump was depressed. The patient had undergone right sided microvascular decompression for trigeminal neuralgia four years prior. There was no history of trauma, fever, dysphagia, hoarseness, weight loss, and no other remarkable medical or familial history. Clinical examination revealed a firm, fixed mass posterior to the sternocleidomastoid muscle with no other significant lymphadenopathy. She exhibited right V3 distribution numbness

☆ Presentations: None.

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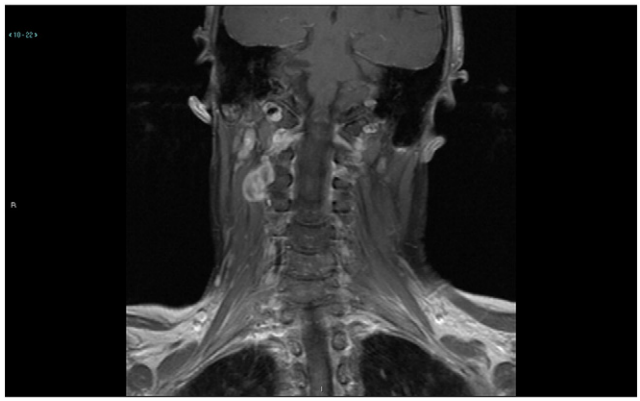

Fig. 1 – T1 coronal (post-gadolinium).

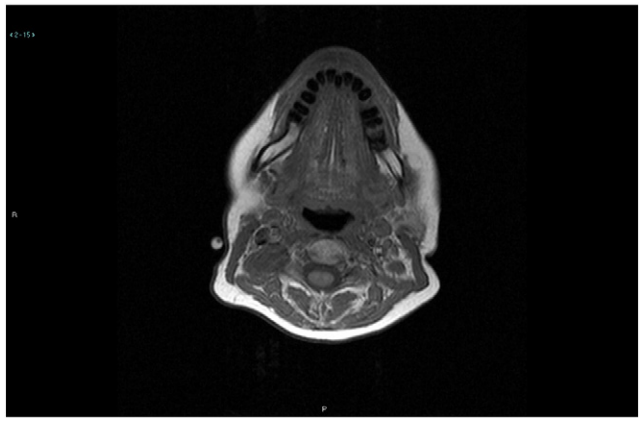

Fig. 2 – T1 axial (pre-gadolinium).

that remained unchanged since her prior surgery. All other cranial nerve functions were intact and symmetric.

MRI of the neck revealed a 2 cm mass at the level of C3-4, just lateral to the right C3 lateral mass The lesion was T2 hyperintense and T1 hypointense with heterogenous enhancement (Figs. 1–3).

Surgical resection was accomplished through a vertical incision in the high posterior triangle of the neck. The mass was found to be firm, encapsulated and gray-white in appearance. It was easily dissected off the surrounding soft tissues and completely excised (Fig. 4). The patient was discharged the same day and reported complete resolution of her symptoms at follow-up.

Final histologic examination demonstrated stellate and small spindle cells and fibers surrounded by myxoid stroma, consistent with a diagnosis of intramuscular myxoma.

3. Discussion

Myxomas are benign mesenchymal tumors of fibroblastic origin which produce excess mucopolysaccharide, are incapable of producing mature collagen, and histologically resemble the umbilical cord [4,5]. To date, a definitive etiology for intramuscular myxomas has not yet been described; however, it is commonly thought that malfunctioning fibroblasts are responsible for both the presence of immature collagen fibers and the abundance of glycosaminoglycans [6]. They may occur in many locations in the body, including the bones, heart, skin, genitourinary tract, retroperitoneal tissues, intestinal tract, pharynx, joints, and skeletal muscles [2]. Intramuscular myxomas usually arise in large skeletal muscles, and have an estimated incidence of 1 in 1,000,000 [7]. Roughly half of intramuscular myxomas are found in the large muscles of the thigh, with less common locations including the buttock, shoulder, lower leg, arm, and trunk [8]. Intramuscular myxomas of the head and neck are rare, with only fifteen cases reported in the literature to date (Table 1). Of these, only four of these cases were localized to the cervical paraspinal musculature [4,9–11].

Myxomas are indolent-growing masses, and as a result the primary chief complaint that patients have is a painless mass; it should be noted, however, that up to 20% present with pain or tenderness in the affected area [3]. With regard specifically to cervical paraspinal muscular myxomas, two of the four reported cases describe an enlarging, painful mass [10,11]. A recent case reported by Kalsi et al. described a case of an intramuscular myxoma in the left sternocleidomastoid muscle that presented with an intense gag reflex on palpation; this was thought to be due to local compression of the vagus and glossopharyngeal nerves [12].

Intramuscular myxomas are cystic-appearing on CT scans, and do not typically enhance with contrast [13]. MRI imaging reveals a mass with sharp borders that demonstrate generally homogenous low signal intensity on T1-weighted images (i.e. lower than muscle) and high signal intensity on T2-weighted images (i.e. higher than fat). It is difficult to correctly diagnose an intramuscular myxoma with radiographic imaging, as the above noted features can be seen in other lesions such as cystic teratomas, lipomas, lymphatic malformations, abscesses, enlarged lymph nodes, myxoid sarcomas and neuromas [11,14].

On gross examination, most intramuscular myxomas are gray-white, shiny, and are circular or ovoid in shape. On close examination of the borders, most intramuscular myxomas are found to infiltrate surrounding muscle [15]. Gross examination of the dissected mass usually reveals a composition of stringy gelatinous material with intermittent fluid-filled cystic pockets [11].

On histologic examination, intramuscular myxomas are found to be hypovascular, hypocellular, predominantly solid with occasional small cystic spaces, and intensely mucoid and basophilic on hematoxylin-eosin staining. Intramuscular myxoma demonstrates an abundance of interstitial mucin, fine fibrillary reticulin fibers, sparse spindle-shaped stromal cells with darkly-stained nuclei, and occasional strands of fibrinous tissue. At the periphery of the lesion, degenerating residual muscle fibers with crowded nuclei are sometimes encountered [8]. Areas of necrosis and mitosis are not typically encountered, nor are differentiated elements such as lipoblasts, chondroblasts, or rhabdomyoblasts [16]. Up to 76% of intramuscular myxomas may exhibit areas of hypercellularity and hypervascularity, which may cause confusion with sarcoma (however, the absence of necrosis, mitosis and nuclear atypia favor the diagnosis of intramuscular myxoma) [17].