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Intramuscular Myxoma of the Temporalis Muscle: Case Report and Diagnosis of an Uncommon Entity in Head and Neck

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Abstract

Objective: Intramuscular myxomas are rare tumors within the head and neck area. This article aims to collect latest findings and relevant data on head and neck and report on a case within the temporalis muscle.

Methodology: A bibliographic search in PubMed with the keywords [intramuscular] AND [myxoma] was conducted. Inclusion criteria were articles regarding the specific anatomic site of head and neck in any language. A 78-year-old patient came to our practice with this type of tumor diagnosed by CT, FNA and excisional biopsy.

Results: Excision showed a 5.2 cm soft mass within the right temporalis muscle with the histological analysis of intramuscular myxoma without extracapsular infiltration. A total of 30 publications focuses in the head and neck area.

Summary: Intramuscular myxoma is rare to be found in head and neck area with surgery described to be curative. It can be associated to other systemic syndromes with fibrous dysplasia.

Keywords: Intramuscular myxoma; Benign tumor of the head and neck; Head and neck tumor

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Introduction

Myxoma is a benign tumor first described by Virchow in 1863 [1], called like this due to its resemblance with the connective tissue found in Wharton's jelly of the umbilical cord. It was later, in 1948 where Stout [2] described this entity as a true mesenchymal neoplasm comprised of undifferentiated stellate cells in a myxoid stroma. Its etiology is not certain, although the dominant theory believes malfunctioning fibroblasts produce an excess deposit of immature collagen and glycosaminoglycans [2,3].

Myxomas can be found mainly in intestines, osseous and soft tissue like skin [4], being most frequently found within the myocardium. The largest soft tissue myxoma case series published in 1965 by Enzinger et al. [5], with 34 cases, the intramuscular myxoma was first described as a subtype and was the 17% (1/34) of the case series. Its incidence is estimated to be $1/10^6$ population per year being even more rare to be found within the head and neck area [6]. It has a slight predominance towards females and is usually diagnosed between 40- and 60-year-old patients [4].

This tumor usually involves large skeletal muscles. The main location is found within the thigh, comprising almost half of the cases. Other locations, in descending order, are gluteus, shoulders, lower and upper limb muscles [4]. On physical examination an individual, soft and elastic mass is found on the anatomic muscular site. It is painless and slow-growing without malignant characteristics [4].

Such findings recommend to perform an imaging test such as an ultrasound, CT or an MRI, which can have in specific features [7]. In order to obtain a definitive diagnosis a histologic examination through FNA or biopsy is required [4]. It is recognized through the microscopic lens because it's very poor vascularization and paucicellularity composed by fibroblasts and abundant myxoid stroma, but hypercellular and hypervascular specimens have been described previously [4].

Once the diagnosis is made a surgical excision is recommended. This paper presents a head and neck case that required surgery with optimal results and a discussion of published evidence up to

date.

Methods and Case Report

A bibliographic search in PubMed with the keywords [intramuscular] AND [myxoma] was conducted. Inclusion criteria were articles regarding the specific anatomic site of head and neck in any language; showing 223 publications in all anatomic sites, while 30 studies are focused in head and neck. A total of 34 intramuscular myxomas have been described in the head and neck area including this one. After words, a literature review was performed regarding aspects and other conditions linked to intramuscular myxoma in this particular area.

The case presented was a 78-year-old male with prior history of auricular fibrillation and hypertension and a long term slow-growing deeply adhered soft mass in the right temporal region that grew partially during the last year with no other lesions. He described having the same bulge for at least 20 years without pain, facial palsy, sensory disturbances or other associated symptoms. The most recent changes made him decide to consult.

A CT was indicated which described a homogeneous subaponeurotic cystic formation without bone erosion that had grown slightly compared to a previous MRI (Figure 1). FNA showed myxoid material few cells with atypia. Given the accessibility and benign characteristics of said tumor an excisional biopsy was decided.

A temporal incision posterior to the preauricular crease under general anesthesia was performed, dissecting skin and subcutaneous tissue superficial to the temporalis fascia, with careful dissection in order to avoid the frontalis branch of the facial nerve. After an incision through the belly of the temporalis muscle the tumor started to protrude, being easily dissectable and sliding out of the incision after a bit of pressure was applied (Figure 2).

Once excised a careful check for hemostasia was performed and a vacuum drain was placed. The muscle was reconstructed with Vicryl and the skin with staples.

Analysis described a smooth-surfaced, soft, encapsulated nodular formation of 5.3 by 5.3 cm with mucoid content. A small resection muscular margin showed no tumor infiltration. A histologic diagnosis of Intramuscular myxoma was made (Figure 3).

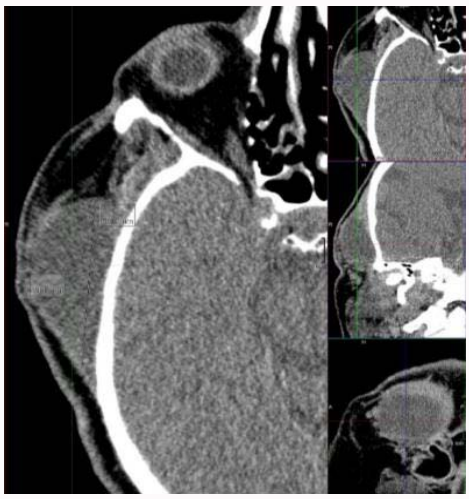

Figure 1: CT scan.

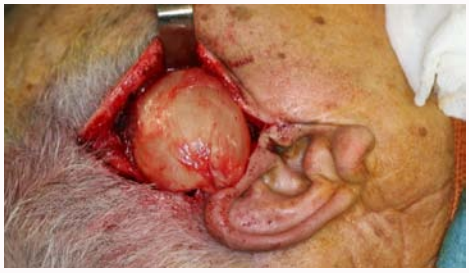

Figure 2: Dissecting skin and subcutaneous tissue superficial to the temporalis fascia.

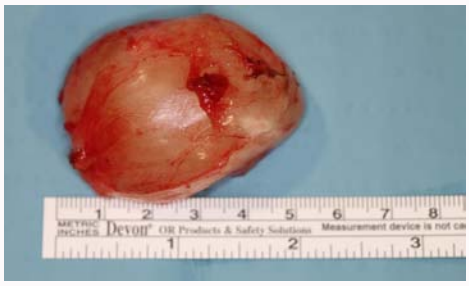

Figure 3: Smooth-surfaced, soft, encapsulated nodular formation of 5.3 cm × 5.3 cm with mucoid content.

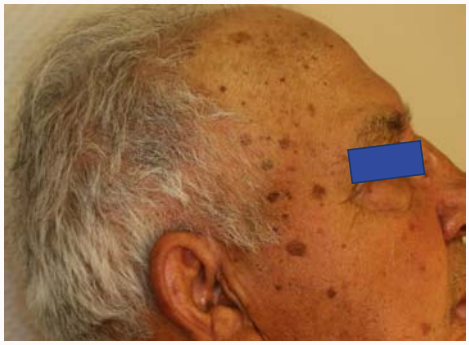

Figure 4: Posterior follow-up showed no complications in short and long term.

Posterior follow-up showed no complications in short and long term. Two years later, the patient refers satisfaction with the cosmetic outcome of the scar and shows no sequels or signs of relapse (Figure 4).

Results

The main locations found in head and neck have been the masseter and temporalis muscle (4 cases each) and cheek (3 cases). The next locations are paraspinal muscles, cervical muscles, trapezius, levator scapulae and SCM. The rest of locations has 1 case described in literature (Figure 5).

This case represents the fourth temporalis muscle intramuscular myxoma described to date in the medical bibliography. The first one by Serrat et al. [8], the second one by Robin et al. [9] and the third one by Higashida et al. [10].

Discussion

This tumor was found to be more common among females (56.7 *vs.* 43.3%) with a mean age around 50 years old [4] being

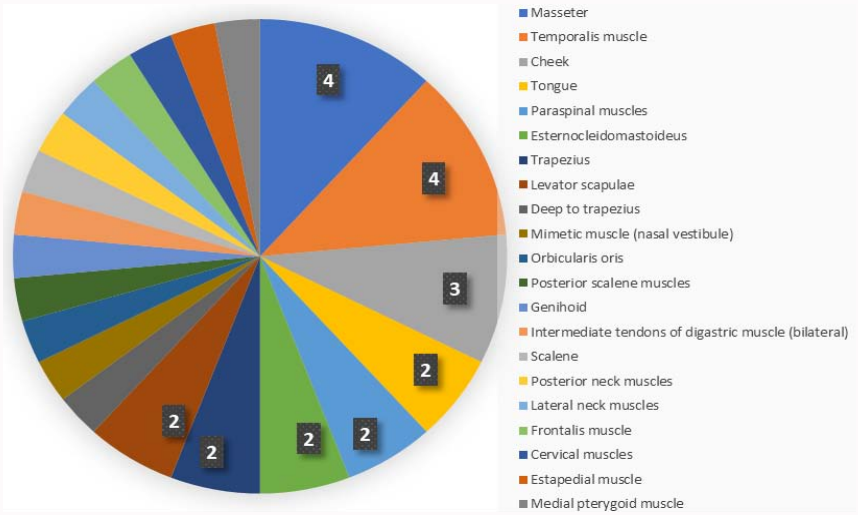

Figure 5: Results.

rare in population younger than 20. The patient described is found close to the age group described in previous articles although the exact beginning remains unknown. It has also been described in the pediatric population, in at least three cases, within the posterior neck region [11] and one unique case of bilateral intramuscular myxomas affecting the anterior belly of digastric muscles [3].

The usual evolution is a slow-growing and painless tumor. Its size has been described between 2 and 8 cm [12,13] in its largest diameter in the head and neck. One case, found within the hyoglossus muscle measuring ~8 cm × 8 cm, made speech and swallowing difficult [13]. Another case reported presented with an intense gag reflex on palpation, was to be found within the left sternocleidomastoid muscle. Local compression of the vagus and glossopharyngeal nerves was thought to be the cause of her nausea. It was solved after the excision [14].

Previous articles describing temporalis muscle affectation refer the leading symptoms to be a palpable enlarging mass without other symptoms [8-10]. In our case the patient didn't show any pain or limitation other than a visible bulge.

On more rare occasions it has been associated with tissue infiltration and bone lysis, the latter being described up to seven times in the literature [15], and even subcutaneous infiltration, although this manifestation is explained by its formation within any of the facial mimic muscles [16].

In case of a soft tissue mass like the intramuscular myxoma CT, MRI or ultrasonography are advised, although the definitive diagnosis is by biopsy [17].

On CT and MRI intramuscular myxomas are most commonly seen as ovoid lesions with well-defined margins. Due to the high-water content of myxoid matrix, they show a cystic appearance on unenhanced imaging examinations, this is low attenuation on CT and high signal intensity on T2 weighted MR images. Nevertheless, they demonstrate variable internal enhancement after the administration of contrast material, consistently with solid masses. Focal areas of heterogeneous internal enhancement can be found, favoring confusion with other malignant tumors [11]. The content within its smooth walls was homogeneous close to 20 Hounsfield Units, which is lighter than muscle and more dense than cerebrospinal fluid, confirming its cystic nature.

Distinctively, these tumors have a perilesional fat rind and adjacent muscular edema [18]. On PET/CT intramuscular myxomas show mildly F-18 FDG uptake [19].

The diagnostic process needs to rule out malignancies such as liposarcoma as the main differential diagnosis (with myxoid degeneration), angiomyxoma, extra skeletal chondrosarcoma, lymphoma, fibrosarcoma, rhabdomyosarcoma, malignant histiocytoma, myxoid neurofibroma [2], low-grade fibromyxoid sarcoma or metastatic disease [8]. It is an important first step in order to discard more preferential management.

Other etiologies to be found are related to infection such as subcutaneous abscess or tuberculoma, or benign tumors. The differential diagnosis includes lipoma, dermoid cyst, lymphadenopathy, branchial cyst, lateral cervical cyst, rhabdomyoma, leiomyoma, nodular fasciitis and oral focal mucinosis [10], nerve sheath tumors or hemangioma [8,14].

The vast majority of these tumors do not show recurrence after surgical excision except a few cases of recurrence that have been reported, mainly after incomplete resection and enucleation. Although the macroscopic aspect is such as of encapsulated, it's been described previously [8] as an incomplete fibrous capsule with the ability of infiltrating surrounding muscle tissue. Sporadic recurrence is very rare and no metastases have been described [4,20].

After a positive imaging test, a fine-needle aspiration cytology is usually advised in order to confirm diagnosis. It is characterized by a rich myxoid stroma filled with mucopolysaccharides and fibrous structural proteins with few fibroblasts [21]. They are furtherly characterized by the presence of spindle cells without atypia or mitosis [22]. Paucicellularity and minimal vascularity are most commonly encountered in accordance to its nature as a benign tumor. In some cases, hypercellularity and a higher rate of vascularization have been found, becoming necessary to rule out sarcoma lineage [2]. The main differences in its behavior are that the latter have a rapid growth tendency, ill-defined limits, capacity of infiltration to surrounding tissues, rich blood supply and heterogeneous components within the tumor. The variety of areas within the tumor account for specific tissues, such as fatty tissue, chondroid matrix and focal areas of necrosis

[11]. An important difference to account for is myxoid degeneration changes within some other types of tumors such as liposarcoma, rhabdomyosarcoma or chondrosarcoma [10]. Histopathological analysis (FNAA and biopsy) showed myxoid content, low cellularity, a focal cyst and capsule continuity without signs of infiltration to the surrounding tissues which adheres to its described general behavior. The findings and clinical follow-up point to the fact that surgery was curative. This case describes an archetypical appearance and treatment of this pathology but it is wise to consider other syndromes in particular scenarios.

If multiple IM were to be found, discarding systemic syndromes is advised. Mazabraud Syndrome (MS) is a rare syndrome that is characterized by a higher recurrence and multiple IM with polyostotic fibrous dysplasia [23] with a higher incidence in females (× 2.3) [24]. Fibrous dysplasia is more prevalent and usually precedes the onset of IM, which is a single tumor in half the cases [23]. Polyostotic dysplasia is the most common type over monostotic affection [23,25].

In the largest cohort found in the bibliography, out of 1,446 patients with intramuscular myxoma the prevalence of MS was found to be 2.2%. The disparity of onset timing postpones the diagnosis of MS to an average of 10.1 years. In this specific population, almost a third of the cases have had recurrence within the first 8.5 years after surgery despite free margins on average. The results of said multicenter study have found association between higher cellularity of myxomas and recurrence [25]. Even though this is not the situation in the case reported, we feel like it is advisable to consider this diagnosis in the forementioned situations.

Summary

- Intramuscular myxoma is a rare, slow-growing and painless tumor to be found within head and neck area. The main referred symptoms are bulge and compression by mass effect.
- CT, MRI, ultrasound and FNA or biopsy is the basis for diagnosis.
- Surgery has been described to be curative in the vast majority of cases, with a very low recurrence rate. Myxomas with increased cellularity may have an indication for longer follow-up.
- Mazabraud syndrome should be considered in cases associated with fibrous dysplasia.

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