Intramuscular myxoma of the cervical paraspinal muscle

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Abstract Myxoma is a neoplasm of mesenchymal origin composed of undifferentiated stellate cells in a myxoid stroma. This tumor can develop in a variety of locations. Myxomas that arise from skeletal muscles are called intramuscular myxomas. They usually occur in large skeletal muscles. Only ten cases of these benign tumors involving the neck muscles were reported in literature. Of them, only three were located at the paraspinal muscles. A 64-year-old woman presented with occipital and neck pain over 5 years noted an expansive painful lesion located at posterior cervical region with progressive volume increase in the last 12 months. Image exams revealed a large mass located in the left posterior region of the neck in contact with the C2, C3 and C4 laminae with no invasion of the vertebra. Tumor total removal was performed through

normal muscle margins and the vertebral periosteum was scraped. The tumor was encapsulated, lobulated with a gray-white appearance. The histological examination yielded the diagnosis of intramuscular myxoma. Follow-up at 1 year showed complete resolution of preoperative symptoms and no evidence of local recurrence. In conclusion, although rare, intramuscular myxoma should be included in differential diagnosis of cervical paraspinal tumors. We reported the fourth case of intramuscular myxoma in the paraspinal musculature of the neck. Despite its benign characteristics, local recurrence was reported after subtotal resection. Tumor total removal should be the goal of surgery.

Keywords Intramuscular myxoma · Benign tumor · Neck · Paraspinal muscle

Introduction

Myxoma is a true mesenchymal benign tumor of fibroblastic origin that produces an excess of mucopolysaccharide and is incapable of producing mature collagen [17]. The tumor, histologically, resembles umbilical cord [12]. These tumors can occur in a variety of locations including the heart, subcutaneous and aponeurotic tissues, bones, genitourinary tract, skin, retroperitoneum, intestine, pharynx, joints and skeletal muscles [15]. Intramuscular myxoma (IM) is a rare tumor that more commonly occurs in large skeletal muscles [2]. The incidence is estimated at one case of IM per million population per year [10]. Myxomas that arise in neck muscles are extremely rare with only ten cases reported in literature [3–7, 9, 12, 14, 16, 17]. We report a case of an IM presented as a posterolateral neck mass in an adult and briefly review the literature concerning this subject.

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Fig. 1 Clinical appearance of the posterior cervical mass reveals a 6 × 6-cm fixed mass in the left posterior aspect of the neck

Case

A 64-year-old woman presented with increasing occipital and neck pain over 5 years. She noted an expansive painful lesion located at posterior cervical region with progressive

volume increase in the last 12 months (Fig. 1). There was no history of trauma, fever, weight loss and no other remarkable medical or familial history. Clinical examination revealed a 12-cm diameter, firm tender mass in the posterior cervical region. The neurological examination was normal.

Computed tomography (CT) of neck showed a hypodense lesion of 15 cm located in the left posterior region of the neck in contact with the C2, C3 and C4 laminae (Fig. 2a). No enhancement in the lesion with intravenous contrast medium was observed on CT scan. The magnetic resonance imaging (MRI) evidenced a mass with irregular borders hypointense in T1 (Fig. 2b) and hyperintense in T2 (Fig. 2c), with no invasion of the vertebrae.

Surgical resection was accomplished through a posterior vertical incision over the tumor site. The tumor was easily separated from the paravertebral musculature and adherent to the C2, C3 and C4 left laminae and articular joint. The tumor was encapsulated, lobulated with a gray-white appearance (Fig. 3). Tumor total removal was performed through normal muscle margins and the vertebral periosteum was scraped (Fig. 3b). The surgical time was 2 h with bleeding of 45 mL. Postoperatively, the patient made an excellent recovery of the pain symptoms. Follow-up at 1 year showed complete resolution of preoperative symptoms and no evidence of local recurrence.

Fig. 2 Cervical CT scan shows a septated, well-defined and hypodense mass in the left posterior neck. The mass does not enhance with intravenous contrast medium (a). Sagittal and axial T1-weighted MRI shows an intermediate to low signal (b) and the T2-weighted shows an increased signal (c) in the septated cystic-looking mass

Fig. 3 Intraoperative aspect of the tumor, which was encapsulated with good cleavage plan (**a**). Tumor total removal was performed through normal muscle margins and the vertebral periosteum was scraped (**b**). Macroscopically cut surface of specimen is ovoid, graywish and gelatinous (**c**)

Fig. 4 Histopathologic views of the intramuscular myxoma. **a** Photomicrography shows the capsulae of the tumor (*purple set*) (hematoxylin–eosin; $\times 4$). **b** Low magnification appearance of the tumor characterized by a paucity of cells, abundance of mucoid material, and almost complete absence of vascular structures (hematoxylin–eosin; $\times 10$). **c** Photomicrography at higher magnification reveals a myxoid stroma sparsely populated by small spindle shape cells and stellate cells without nuclear atypia and fibers (hematoxylin–eosin; $\times 40$). **d** Low magnification show the tumor (*purple set*) surrounded by normal skeletal muscle (*green set*) (hematoxylin–eosin; $\times 4$)

The histological examination yielded the diagnosis of IM, showing small spindle cells, stellate cells and fibers in abundant myxoid stroma. Mitotic activity, necrosis and nuclear atypias were absent (Fig. 4).

Discussion

In 1863, Virchow [19] introduced the term myxoma to describe a benign tumor that histologically resemble

mucinous substance of umbilical cord tissue. Histological criteria for the diagnosis of myxoma was then proposed by Stout [18] deeming it as a true neoplasm of mesenchymal origin composed of undifferentiated stellate cells in a myxoid stroma with delicate reticulin fibers. These tumors may occur in various locations. The myxomas that arise from skeletal muscle are called IM. This tumor was described as a distinct subtype of myxomas in 1965 by Enzinger [6], and comprised only 17% of all soft tissue myxoma in his report. The majority of IM cases are in the fourth to sixth decade of life and there is a slight female predominance [6, 10].

Almost half of IM are situated in the thigh [2]. Other less-common locations in descending order of frequency are buttock, shoulder, lower leg, arm and trunk [2]. IM rarely occurs in neck. To our knowledge, there have been reported only ten cases of IM in the neck in literature [3–7, 9, 12, 14, 16, 17] (Table 1). Three of these cases were reported in the cervical paraspinal muscles [5, 7, 17]. The clinical manifestations of IM of the neck are non-specific, and it is difficult to diagnose the tumor before microscopical examination. It is a slow-growing tumor and usually presents as a painless mass that may produce symptoms due to the compression of surrounding structures [5, 7, 17]. Only approximately 20% are painful [6]. In the case described, the evolution of the pain was 5 years and the patient noted an enlarging mass over 1 year. In the cases of IM in the cervical paraspinal muscles reported by Shugar et al. [17.] and Crankson et al. [5], the clinical manifestation was a gradually enlarging non-painful mass in the posterior cervical region.

Because of the infrequent occurrence of IM, it may not even be suspected after radiological investigations. CT typically reveals a cystic-looking mass, as was this case, with an attenuation value greater than water, but less than surrounding normal muscle and no enhancement after intravenous administration of contrast medium [5, 8, 9]. On MRI, IM have sharp borders with homogeneous low-signal

intensity, lower than skeletal muscle, on T1-weighted images and homogenous high intensity, brighter than fat, on T2-weighted images [1]. These radiological features may be presented in other fluid-filled lesions such as cystic hygroma, cystic teratoma, lipoma, hematoma, abscesses, lymph nodes, neuroma and myxoid sarcoma.

The gross appearance is characteristic and changes little from case to case. Most tumors are ovoid or globular and have a glistening gray-white or white appearance, depending on the relative amounts of collagen and myxoid material. They consist of mass of stringy gelatinous material with occasional small fluid-filled, cyst-like spaces, and they are covered by bundles of skeletal muscle of fascial tissue [20]. Although on gross examination most of the tumors appear to be well circumscribed or encapsulated, on close inspection, the delicate fibrous capsule is incomplete, and most lesions infiltrate adjacent muscle [14].

Histologically, IM are hypocellular, hypovascular, intensely mucoid and basophilic using the hematoxylin–eosin stain and predominantly solid [2]. The four main components are interstitial mucin, sparse spindle-shaped stromal cells, fine fibrillary reticulin fibers and varying numbers of strands or trabeculae of fibrous tissue. A few residual, shrunken, degenerating voluntary muscle fibers with crowded nuclei surrounded by myxoma tissue usually can be seen at the periphery [2]. Inflammatory cells are not a feature, but occasional macrophages and rare mast cells may be seen. IM are mainly solid, although a few irregular small cystic spaces or holes may be found. The stromal cells are scattered throughout the mucinous material, they usually are small, with darkly stained nuclei, and may be spindle-shaped, stellate or even oval to round. Mitoses and necrosis are not seen. The stromal cells cytoplasm is often continuous with fine fibrillary reticulin, resembling fibers of frayed string, that course irregularly through the lesion [2, 20]. Some IM show focal areas of hypercellularity and hypervascularity, which may cause further confusion with sarcoma [2, 11, 20]. Hypercellular zones can appear in 76%

Table 1 Location of cervical intramuscular myxomas reported in literature

Number	References	Age, sex	Location
1	Enzinger [6]	55, Male	Sternocleidomastoid muscle
2	Rosin [16]	44, Male	Geniohyoid muscle
3	Canallis et al. [4]	46, Female	Lateral part of neck
4	Feldman [7]	62, Female	Cervical paraspinal musculature
5	Nishijima et al. [12]	16, Female	Anterior part of the neck
6	Shugar et al. [17]	68, Female	Cervical paraspinal musculature
7	Andrews et al. [3]	61, Female	Neck (no more information)
8	Crankson et al. [5]	5, Female	Cervical paraspinal musculature
9	Ozawa et al. [14]	22, Male	Scalene
10	Ishoo [9]	2, Female	Trapezius
11	Case reported	64, Female	Cervical paraspinal musculature

of IM and it can occur 10–80% of the tumor [11]. However, even in these hypercellular zones, the absence of mitoses, nuclear atypia and necrosis indicate IM [2, 11, 20].

Surgical excision is the treatment for IM. There is no report of metastasis or malignant change, but some cases have been reported to recur when the surgical procedure has been enucleation or incomplete resection [11, 13]. If possible, it is better to extract this tumor with a part of surrounding muscle, where the boundary is not clear, since IM has an infiltrative tendency to muscles as a part of its character. In this case, tumor total removal was performed through normal muscle margins and no recurrence was observed at 1 year of follow-up.

In conclusion, IM is infrequent in the neck. We reported the fourth case of this tumor in the cervical paraspinal musculature. Its difficult to make a diagnosis before operation because of the lack of characteristic clinical history and radiographic findings. There is no report of malignant change of myxoma, but occasionally recurrence resulting from incomplete excision has been reported. Although rare, IM should be included in differential diagnosis of cervical paraspinal tumors.

Conflict of interest statement None of the authors has any potential conflict of interest.

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