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ATYPICAL MYXOMA: CASE STUDY OF A RARE PARASPINAL TUMOR

Neurosurgery

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ABSTRACT

Myxoma is a neoplasm of mesenchymal origin composed of undifferentiated stellate cells in the myxoid stroma. Only eight cases of these benign tumors involving the paraspinal muscles were reported in literature. We report the ninth case of intramuscular myxoma in the paraspinal musculature. A 57-year-old male presented with progressively increasing painful swelling in the left paraspinal region. Imaging revealed involvement of erector spinae muscle. The tumor was resected and sent for histopathological examination which revealed atypical intramuscular myxoma. Follow-up at 1 year showed complete resolution of preoperative symptoms and no evidence of local recurrence. Although rare, intramuscular myxoma should be included in differential diagnosis of paraspinal masses.

KEYWORDS

Paraspinal, Intramuscular, Myxoma

INTRODUCTION

Myxomas (from the Greek word 'muxa' meaning mucus) are rare, benign connective tissue tumors arising from stellate mesenchymal cells, comprising entities such as fibromyxoma, cardiac myxoma and intramuscular myxoma (IM)[1]. It can arise in a variety of locations such as the heart, subcutaneous and aponeurotic tissues, bone, genitourinary tract, and skin. The most frequent site for myxoma is the heart and bony myxoma is almost exclusively limited to the jaws and palate [2-3]. Incidence rates of IM vary between 0.1 and 0.13 per 100,000 individuals per year[4]. This benign lesion favors a female predilection, occurring between the fourth and sixth decades of life [5]. We report a rare case of atypical myxoma developed within the lumbosacral paraspinal muscle of a male patient.

CASE REPORT

A 57-year-old male presented with swelling in the lower back for 2 years duration, which was gradual in onset and progressively increasing in size, associated with dull aching persistent lower back pain. There was no history of any other similar swelling in the body, no history of fever, weight loss, loss of appetite, radiating pain to the lower limbs, sensory impairment, or motor weakness in the lower limb. On physical examination there was a large palpable firm non-tender mass in the left paraspinal lumbosacral region (L2-L5), neurologic examination was within normal limits.

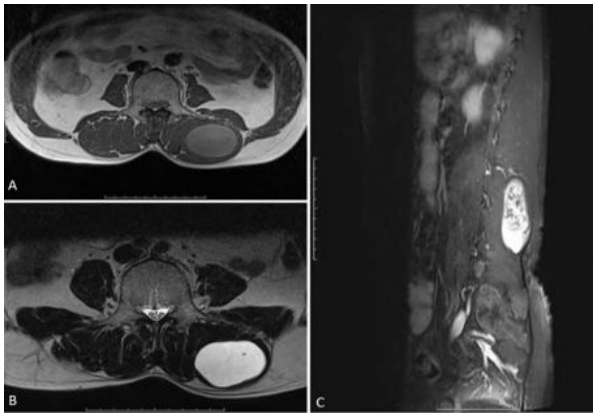

Figure 1- A- T1WI Axial section- hypointense lobulated mass in paraspinal muscle; B-T2WI Axial section- hyperintense lobulated mass in paraspinal muscle; C- Gadolinium Contrast image- Sagittal section - heterogeneously enhancing lobulated mass in paraspinal muscle

The magnetic resonance imaging (MRI) demonstrated a lobulated mass in the left paraspinal muscle (erector spinae), which was hyperintense on T2WI, hypointense on T1WI, and heterogeneously enhancing on gadolinium, measuring 4cm(Anteroposterior) X 5.2cm(Mediolateral) x 8cm(Craniocaudal) extending from L2 to L5 vertebral body **(Figure 1)**. The differential diagnosis at this point was a peripheral nerve sheath tumor. The tumor was completely resected with a left paramedian longitudinal incision.

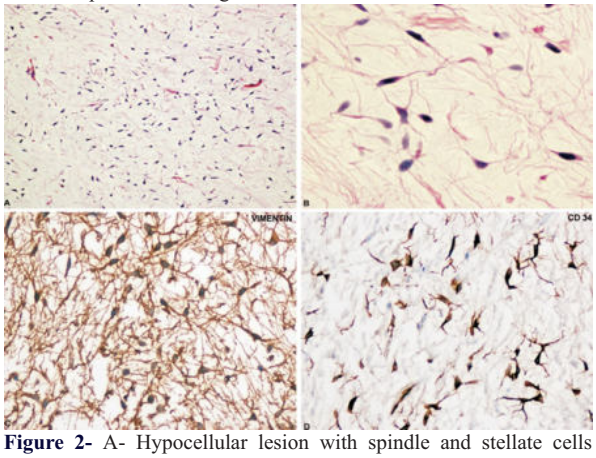

Figure 2- A- Hypocellular lesion with spindle and stellate cells embedded in loose edematous & occassional congested blood vessel; B- Spindle /stellate shaped cells with long cytoplasmic processes dispersed in a highly myxoid stroma; C- Vimentin strongly labels tumor cell with background collagen fibrils.(Immunoperoxidase x Obj20); D- CD34 highlights tumor cell cytoplasm and cytoplasmic processes. (Immunoperoxidase x Obj20)

On gross examination, the tumor was ovoid, glistening grey-white in appearance, well encapsulated, and contained gelatinous material. The patient tolerated the surgical procedure well and no postoperative

complications were noted. On a histopathological examination, it was an encapsulated hypocellular tumor with myxoid stroma and many bland-looking cells with multiple cytoplasmic processes. There were few collagen fibrils deposition in the myxoid stroma. On immunohistochemistry, the tumor cells were strongly labelled by Vimentin and CD34. The Mib1 labelling index was less than 1%. The adjacent skeletal muscle fibers and fibrofatty tissue were free of tumor **(Figure 2)**. Thus, based on histopathology and immunohistochemistry, the diagnosis was consistent with intramuscular myxoma.

DISCUSSION

Soft tissue myxomas have a rare incidence[6]. It was first described in 1863 as a benign tumor that histologically resembles mucinous substance of the umbilical cord [6]. This is a true mesenchymal neoplasm composed of undifferentiated stellate cells embedded in myxoid stroma containing weak reticular fibers [5,6]. To date, the etiopathogenesis of myxomas remains elusive. Some theories suggest trauma to be an etiological factor [9], while other theories suggest stellate cells may be altered fibroblasts that fail to produce mature collagen and there is growth of polysaccharide-producing cells to be involved in the neoplastic process[5,7-9]. It has also been proposed that there is fibrous dysplasia that usually precedes the IM [10].

Though there was no report about the prevalence by race, most of the previously reported cases were from the western population with female preponderance. This is the second case of IM being reported in the Asian population, which developed in the lumbosacral paraspinal musculature. Clinically intramuscular myxoma is usually a single, asymptomatic, painless, slowly progressing, palpable, well-defined mass[11]. This tumor may occur in isolation or in association with fibrous dysplasia or Albright syndrome. If multiple, they are usually associated with fibrous dysplasia of the bones of the same extremity, known as Mazabraud syndrome[12-13]. Radiological evaluation can be done with ultrasound, computerized tomography (CT), or MRI[14]. The ultrasound shows IM as a hypo-echoic lesion with well-defined margins and there may also be a presence of anechoic cystic foci[14]. The CT shows IM as a well-demarcated homogeneous low-density lesion within the skeletal muscle [15]. There is low signal intensity on T1-weighted images and high signal intensity on T2-weighted STIR [14-15].

The definitive diagnosis is histopathological evaluation which shows a hypocellular and hypovascular tumor with undifferentiated spindle-shaped cells, separated by reticulin fibers in an abundant myxoid matrix of hyaluronic acid. It shows benign characteristics with the absence of mitosis, nuclear atypia, and necrosis [11, 16]. The majority of lesions infiltrate into the adjacent muscles, as the delicate pseudocapsules are incomplete on close inspection[17]. On immunohistochemical staining, IM cells stain positively for vimentin, and variably for CD34 and actin; immunostain for S-100 protein is typically negative[17]. The absence of vascularity decreases the likelihood of sarcoma, and the absence of S-100 protein excludes myxoid neurofibromas and low-grade malignant peripheral nerve sheath tumors[18]. The optimal treatment for IM is complete surgical excision. Although IM is a benign tumor, it tends to recur, with an accompanying risk of malignant transformation if incompletely excised[19-20].

CONCLUSION

In the present study, we report a rare case of IM, presenting as a swelling in the paraspinal musculature. A thorough surgical resection showed complete resolution of preoperative symptoms and there was no evidence of local recurrence at the end of 1-year follow-up. Clinicians should be aware of this rare entity and consider it in the differential diagnosis of paraspinal masses to successfully manage these patients. Surgical excision has shown to be curative in the vast majority of cases, with minimal recurrence rates.

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