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Neuromuscular Hamartoma in A 22 Year Male: A Case Report

KEYWORDS: Neuromuscular hamartoma, benign Triton tumour, peripheral nerve tumour

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ABSTRACT *Neuromuscular hamartoma (choristoma) or benign "triton" tumor is a very rare peripheral-nerve tumor composed of mature nerve fibers and well-differentiated striated muscle fibers. Only seven cases have been reported in the literature. A new case of this rare entity is described in a 22-year-old male patient with a recurrent left manubrium mass . Unlike the previous cases, this patient had continued to have proliferation of her tumor since childhood, requiring total resection in adulthood. It is concluded that total resection of these tumors should be attempted at initial diagnosis. The histogenesis of these tumors remains unclear.*

INTRODUCTION

The neuromuscular hamartoma (also referred to as the neuromuscular choristoma or benign triton tumour) is a rare developmental lesion composed of mature elements of striated muscle and nerve. [1,2].Of the fewer than 50 cases from the literature,the majority occurred in young children as solitary masses involving large nerve trunks,particularly the brachial and sciatic.40 Tumors arising from the cranial nerves also occur but usually present during adult life.41.at times, fibrous component surrounding the lesions is so dense and cellular that it suggests the diagnosis of fibromatosis replacing nerve and muscle. We present a rare case of a neuromuscular hamartoma on chest wall near manubrium in a twenty two year old male patient with no neurological deficit.

CASE REPORT-

A 22-year-old male presented to the surgical outpatient clinic with history of painless diffuse swelling over left lower chest area near manubrium since birth. There was no complaint of pain, restriction of movement, paraesthesia or any other symptom associated with the swelling. The patient had no significant past/surgical/ medical/ family/ personal history.

Local examination revealed a diffuse, non- tender, soft to firm swelling with ill- defined borders ,immobile mass measuring 10x8x4 cm on the left sided lower chest area ,unattached to overlying skin was noted. Chest x ray at this stage showed a peripheral soft tissue lesion anterior to manubrium sterni. The CT-scan revealed small loculated soft tissue lesion anterior to manubrium sterni measuring approx. 27x56x54mm in longest dimension with periosteal reaction and minimal erosion of the cortex of manubrium. Anterior abnormal projection of the manubrium sterni was also seen. Other routine investigations of the patient were unremarkable.

The tumour was excised and sent for histopathological examination. On gross examination, the specimen consists of two fibromuscular greyish brown soft to firm tissue piece , the largest of which measured 6 x 3 x 2 cm. On cut section, it was grey-white to grey brown and subdivided by collagenous connective tissue into smaller nodules.

Microscopy showed nodules and fascicles composed of well differentiated skeletal muscle fibres of varying sizes separated by fibrous tissue bands admixed with nerve bundles, some of which were enclosed within the perimysium of the skeletal muscles . In some areas, the nerve fibers and muscle fibers were individually scattered in a collagenous stroma. No cytologic atypia or mitoses were present. . These microscopic findings were diagnostic of neuromuscular hamartoma. On immunohistochemical staining, the skeletal muscle fibres and nerve tissue were strongly positive for myoglobin and S-100 respectively.

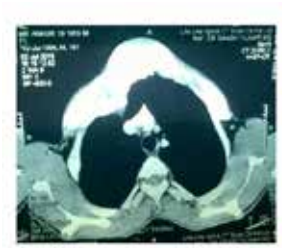

FIG.1 Helical CT thorax

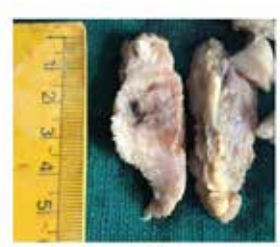

FIG.2 Gross specimen

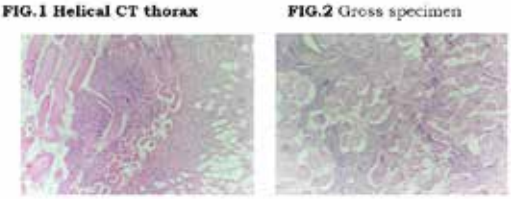

FIG.3,4 Fig.4: Photomicrograph showing mature skeletal muscle fibres admixed with nervous and adipose tissue elements

FIG.3,4 Fig.4: Photomicrograph showing mature skeletal

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muscle fibres admixed with nervous and adipose tissue elements

DISCUSSION-

Benign triton tumours, also referred to as neuromuscular hamartomas or neuromuscular choristomas are extremely rare neoplasms with a variable combination of peripheral nerve and skeletal muscle differentiation in an organized fashion in a manner somewhat akin to neuroskeletal development.They are believed to be hamartomatous lesions of neuroectodermal- mesenchymal origin which could have resulted due to incorporation of mesenchymal tissue into nerve sheaths during embryogenesis or aberrant differentiation of neuroectodermal elements into mesenchymal components or hamartomatous growth of muscle spindles [5-7] . It usually occurs in infants and young children. [1,2]

The histogenesis of neuromuscular choristoma remains a controversial issue.1-17 Louhimo and Rapola2 suggested that the skeletal muscle might originate from the limb mesenchymal cells trapped within the nerve sheath during embryogenesis. Another possibility is that the neuroectoderm, the so-called ectomesenchyme, can give rise to mature skeletal muscle in much the same way as the neuroectodermal eye cup gives rise to the iris muscle.2,4,5,15,16 Masson has offered that endoneurial cells of neuromas, under the organizing influence of the motor nerve fibers, may be able to differentiate into muscular tissue and this hypothesis was used to explain the histogenesis of malignant triton tumor.(8,9)

The first reported case of neuromuscular hamartoma, published in 1895, involved the sciatic nerve. [10] In earlier case reports, neuromuscular hamartomas were generally associated with a large peripheral nerve, such as the brachial plexus (25%) [11] or sciatic nerve (15%). [8,9] Cranial nerve involvement, including that of the trigeminal, [10] facial, occulomotor, cochlear, occipital and optic nerves, was subsequently reported in 18 cases (45%). [4] Cutaneous lesions have been reported in four cases

A variety of lesions including neuromesenchymal hamartoma, ectomesenchymal hamartoma and rhabdomyomatous hamartoma can be included in the differential diagnosis of this tumour.(12)

This case is also unique in that the patient had no associated neurological symptoms at presentation. Often, total excision of neuromuscular hamartoma is not possible because of its close association with a major nerve. Therefore, only partial excision or biopsy can be performed in many cases. [2] Even an incomplete excision has resulted in symptomatic relief and tumour regression. Treatment should be conservative and aimed at preserving nerve integrity

CONCLUSION

Diagnosis of a benign triton tumour depends solely on the histopathological examination since the clinical diagnosis is difficult. Sometimes, the fibrous component may predominate, thereby mimicking fibromatosis. After a correct diagnosis, the treatment should be conservative and primarily aimed at maintaining the integrity of the nerve. A complete excision is totally curative.

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