

## RHABDOMYOMATOUS MESENCHYMAL HAMARTOMA OF THE ARM : A RARE CASE REPORT

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### Abstract

Hamartomas are benign lesions that contain disorganized growth of mature tissues[5,6]. Choristomas are similar, except they are comprised of tissues that would not typically be found at the anatomic site at which the lesion is arising. A variety of hamartomas and choristomas can occur in skin and soft tissue and be challenging to diagnose. Rhabdomyomatous mesenchymal hamartomas (RMH) or striated muscle hamartomas (SMH) are an unusual, benign, congenital or acquired neoplasms affecting mainly children, most of the times in the head and neck region. They are anormal dermal and subcutis aggregates of disorderly striped muscle rays teamed with regular mesenchymal elements. Management is typically surgical excision, with rare descriptions of recurrence.

**Keywords:** mesenchymal, rhabdomyomatous, striated muscle, surgery

### INTRODUCTION

The word hamartoma comes from Greek ‘hamartanein’ which translates to err/fault, and ‘oma’ which means a tumour like growth. They are considered benign lesions, attributable to an error of development, which are often present at birth, although they can also be obtained at a later age, and consist of aberrant development of mature structure elements, but generally characterised by a disorganised architecture. (1) Hamartomas are usually asymptomatic, incidentally detected masses located at any site. They can also cause cosmetic concerns based on their location, and may need to be surgically excised. The original description was captured by Hendrick et al. as striated muscle hamartoma or rhabdomyomatous mesenchymal hamartoma. in 1986 as a rare congenital tumour

mainly found on the face and neck of babies. (2-4) The etiology of RMH is unknown, but it is histologically defined by striated muscle. (5) RMH presents as one or more lesions, often polypoid, typically on the midline, and is defined by mesenchymal elements (fat, connective tissue, blood vessel, nervous tissue) and striated muscles randomly disposed in the dermis and subcutaneous tissues. (2,4,6,7)

### Case Report

A 20 year old female patient came to us with a swelling back of the left arm since 7 years. ,which was insidious in onset and gradually increase to the current size. It was visceral aching pain and non-radiating. There was no history of trauma, ulceration, discharge or bleeding. No visible changes in the size of the swelling .Nocomorbidities history. On physical examination, there was a firm and tender swelling over the left triceps region measuring approximately 6 x 3 cm. (Fig. 1) On palpation it was tender, mobile in transverse direction, upon contracting for triceps the swelling became less prominent and regional lymphadenopathy was not present. MRI demonstrated a well-defined focal intramuscular lesion centred in the triceps muscle that was T2 and fat-saturated hyperintense with mild perifocal intramuscular edema and suspicious internal flow voids and calcifications (suggestive of vascular or calcified soft tissue lesion). (Fig. 2)

We were prepared for exploration and surgical excision of the lesion. Aseptic precautions (part painted and draped). An incision was given over the most bulging part of swelling and deepened in layers. (Fig. 3) The tricep muscle was divided, the edema recognized and excised in toto. (Fig. 4) superior aspect, however vascular pedicle was identified, clamped and ligated, specimen was sent for histopathology. (Fig. 5) The severed tricep muscle was apposed with 3-0 polyglactin sutures and the skin with 2-0 nylon sutures over 12 Fr suction drain. (Fig. 6) POP slab with sterile dressing applied. Histopathological examination showed skeletal muscle bundles and a lesion consisted chiefly of cavernous spaces filled with blood, ectatic lymph spaces, adipose tissue, skeletal muscle fibres, fibrocollagenous tissue with focal lymphoid aggregates and vitalised bony tissue in keeping with benign striated muscle hamartomatous lesion. (Fig. 7) On postoperative day 10, the sutures were removed



Fig. 1 – Clinical photograph of the lesion

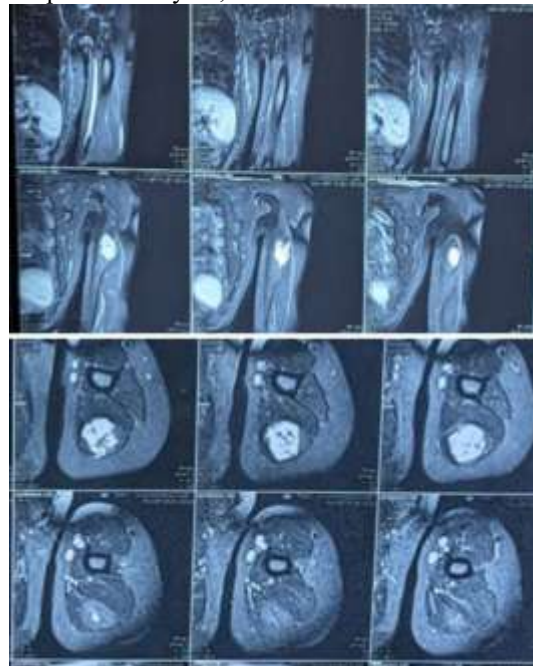


Fig. 2 – MRI findings of the lesion



Fig. 3 – Lesion being excised



Fig. 4 – Photograph following excision



Fig. 5 – Specimen after excision



Fig. 6 – Photograph after skin closure

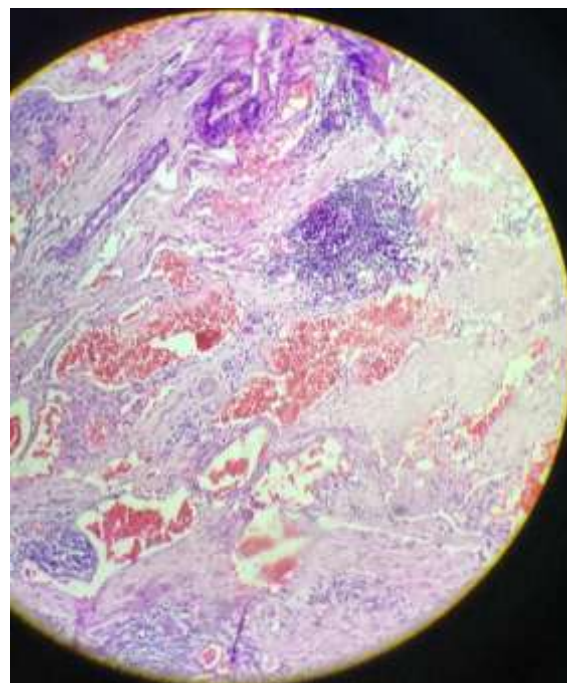


Fig. 7 – Microscopic findings of the lesion

## DISCUSSION



Rhabdomyomatous mesenchymal hamartomas are uncommon, benign cutaneous lesions that may appear in utero or later in life and consist of an aberrant of striated muscle, adipose tissue, and collagen fibres. Clinical features: They most commonly arise as a single (sometimes multiple) identically shaped polypoid or nodular mass, with major or no change in the colour of the overlying skin; usually seen on the head and neck region. (9,10) It is a deep dermal and subcutaneous tissue lesion composed of mature skeletal muscle fibres intermixed with skin adnexa such as sebaceous glands, hair follicles and sweat glands. Adipose tissue, nerves, blood vessels and collagen may also be affected in these lesions. It has shown a slight male predominance and most often occurring in developmentally prepubescent children. Histopathological findings show randomly arranged bundles of mature striated muscle interspersed with mesenchymal, vascular, and neural elements throughout dermis and hypodermis with immunohistochemical staining positive for actin and desmin, myoglobin and Masson's trichrome. (9,11) (11) The etiology of SMH is unknown, but it is believed that it may result from an abnormality in the migration of embryonic mesodermal tissues. (12) It may be associated with other congenital anomalies, including cleft lip, microtia, thyroglossal duct cyst, occult spinal dysraphism, Goldenhar and Delleman's syndromes. (9,13) Hence, overall examination of all other systems needs to be performed to rule out other findings. (10–12) Other cases have been described on the tongue, perianal region, vagina, and great toe. (15) Rhabdomyomatous mesenchymal hamartoma is thought to be a benign tumour, as there were no reported cases of malignant transformation. (16) Thus, the treatment of rhabdomyomatous mesenchymal hamartoma is excision and no recurrences have been reported following excision. (15) Two of them were spontaneously regressed at presentation even in the absence of excision. (9,13)

## CONCLUSION

This case is reported because RMH is rare and presents with unusual symptoms. A meticulous surgical excision was performed without compromising the structural integrity of the philtrum of the upper lip. Despite the benign and rare nature of RMHs, their relationship to other congenital anomalies and embryological faults should always be considered in patients with this diagnosis.

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