Case report :

Rhabdomyomatous mesenchymal hamartoma: Report of the first case in the Middle East

Abdul Majeed AlHadlaq

Imam Mohammad Ibn Saud Islamic University

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Corresponding Author:

Abdul Majeed AlHadlaq
Imam Mohammad Ibn Saud Islamic University
aalhadlaq@imamu.edu.sa
Phone: +966557777490

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Abstract

Rhabdomyomatous mesenchymal hamartoma (RMH) is a rare congenital tumor of the dermal and subcutaneous tissues originating in areas where there is a superficial striated muscle. To the best of our knowledge, only 77 cases were found in the literature, and in most of these cases, it presents in the midline head and neck, with a few cases with unusual presentations being reported. Although RMH is usually a benign and incidental finding, few reported cases were associated with syndromic or congenital anomalies. To our knowledge, this is the first case reported in the Middle East, indicating that the incidence of RMH and race are not associated.

Introduction

In 1986, Hendrick first described Rhabdomyomatous mesenchymal hamartoma (RMH) as a striated muscle hamartoma [1], a name first given by Mills in 1989 [2].

RMH is a rare congenital tumor of the dermal and subcutaneous tissues originating in areas where there is superficial striated muscle [1]. The literature review was carried out using the PubMed search engine with the following keywords: Rhabdomyomatous mesenchymal hamartoma, achrodon, skin tag, and striated muscle hamartoma. Only articles written in English were included in the search results. The pathological diagnosis for the inclusion criteria was "Rhabdomyomatous mesenchymal hamartoma." To the best of our knowledge, only 77 cases are found in the literature. In most of these cases, it presents in the midline head and neck, with a few cases with unusual presentations being reported. Ten cases had unusual presentations: 3 cases in the tongue; 4 cases in the perianal region; 3 cases in the tonsil; 1 case in the vagina; 1 case in the great toe [3-6].

Although RMH is usually a benign and incidental finding, few reported cases were

associated with syndromic or congenital anomalies. The differential diagnosis for a similar lesion is acrochordon, hair follicle nevus, and pedunculated seborrheic keratosis. To our knowledge, this is the first case reported in the Middle East, implying that the incidence of RMH and race are not associated.

Case

A 7-month-old girl presented to the clinic with her parents for a pedunculated lesion on the chin (Fig. 1-A). Her birth was a normal spontaneous vaginal delivery. The patient was born with the lesion, and her parents did not notice any change in size, color, or shape.

There was no family history on both sides of the same anomaly or similar lesions. It was a pedunculated, smooth, and soft lesion. There was no change in color or temperature of the lesion or the surrounding area. Moreover, the lesion had no opening or discharge. It measured around 0.7 cm in length and 0.5 cm in diameter. The patient was fully examined for other anomalies, but nothing was found.

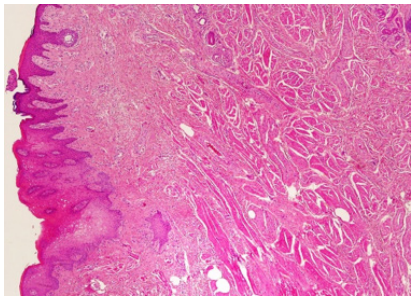
The family was concerned regarding the pathology and the cosmetic appearance of the lesion and wanted it to be removed. An excisional biopsy was planned under local anesthesia. The incision was elliptical. Then, the lesion was excised, and the incision was sutured with deep dermal absorbable suture 4.0 and three simple interrupted nonabsorbable sutures 6.0 (Fig. 1-B). The patient healed with no postoperative complications.





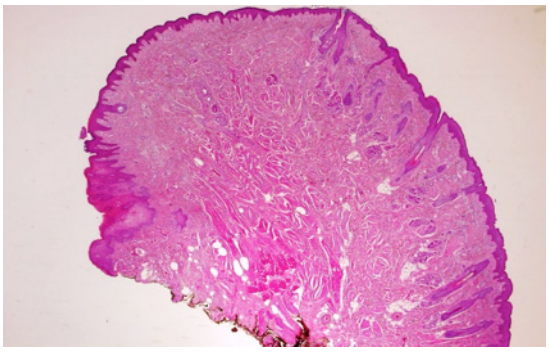
Figure 1-A: The lesion pre-operatively.
Figure 1-B: The lesion post-operatively.

The pathology notes were “polypoid epidermis with underlying mixed connective tissue including skeletal muscle, peripheral nerves, blood vessels and adipose tissue with no signs of malignancy.” The pathological diagnosis was Rhabdomyomatous mesenchymal hamartoma (Fig. 2-4).



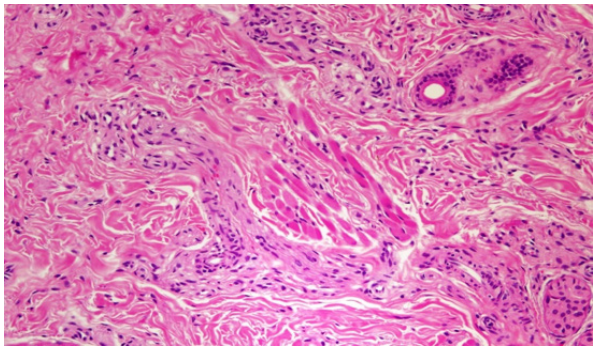
(FIG 2)

Figure 2: low power view shows polypoid lesion covered by epidermis and partially by non-keratinizing stratified squamous mucosa (H/E stain, original magnification x 20).



(FIG 3)

Figure 3: photomicrograph of medium power slide reveals underlying dermis with skeletal muscle and peripheral nerve. (H/E stain, original magnification x 100).



(FIG 4)

Figure 4: high power focusing on the skeletal muscle and peripheral nerve tissue. (H/E stain, original magnification x 400).

Discussion

RMH is a rare benign congenital skin and soft tissue lesion. Mills classically described it as hamartoma of mature striated muscle fibers occupying the greater part of the mass but admixed with some mature adipose elements [2].

The classical presentation of the cases is midline head and neck region and areas of superficial smooth muscle; RMH appears as a solitary lesion, papule, or pedunculated. Scattered cases had unusual presentations, such as in the tongue and perianal region, vagina, and great toe [7].

RMH is a benign lesion and has no malignant transformation. First, Williams has reported one case with regression of the lesion 8, while Mazza et al. have recommended waiting 1-2 years in another case to prove the regression by biopsy [3].

Hendrick has documented a second case with amniotic band syndrome (ABS), which prompted him to consider possible relation that these lesions could have originated during the embryonic period by amniotic adhesion due to pulling of the skin [1].

Regarding the presentation of the classical cases, involvement of the facial expression muscles and platysma, which arise from the primitive mesoderm of the second branchial arch, could be responsible for the haphazard migration of these muscles' fibers to the dermis [9]. Other cases of RMH have been reported in association with syndromes or congenital anomalies other than ABS, Delleman (oculocerebrocutaneous) syndrome, and Goldenhar spectrum, cleft lip, cleft gum, and nasofrontal meningocele with a dermoid cyst [10].

In summary, RMH is the histopathological diagnosis. It is a rare, benign lesion and has no malignant transformation. The classical presentation is the midline head and neck region and areas of superficial smooth muscle, appearing as a solitary lesion, papule, or pedunculated. The treatment of choice is surgical removal. Most of the cases are treated for cosmetic reasons. This presented case is the first reported case in the Middle East.

- Verbal consent for photography and publishing was taken from the parents.
- Ethical approval was granted by IRB at Imam Mohammad Ibn Saud Islamic University approval for project number "143-2021."

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