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RHABDOMIOMATOUS MESENCHIMAL HAMARTOMA OF NOSE. CASE REPORT

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ABSTRACT Rhabdomyomatous mesenchymal hamartoma (RMH) is an extremely rare congenital lesion of the dermis and soft tissues consisting of a disordered and varied collection of mature adipose tissue, skeletal muscle, adnexal elements and nerve bundles. A case of subcutaneous RMH of nasal dorsum in a 21-year old woman is reported. The surgical removing of lesion with simultaneously rhinoplasty was performed.

KEYWORDS : rhabdomyomatous mesenchymal hamartoma, connective tissue disease, rhinoplasty, immunohistochemistry

Introduction

Rhabdomyomatous mesenchymal hamartoma (RMH) is a rare dermal or subcutaneous lesion comprising disordered mature adipose tissue, skeletal muscle, adnexal elements, nerve bundles and collagen (Rosenberg *et al.,* 2012).

It was, first described as Striated Muscle Hamartoma by *Hendrick et al* in 1986, and then has been reported under various names: striated muscle hamartoma, congenital midline hamartoma, hamartoma of cutaneous adnexa and mesenchyme (Rosenberg *et al.,* 2012).

RMH usually presents as a polypoid or papular cutaneous lesion, sometimes even plaque-like lesion, that ranges in size from a few millimeters to 1-2 cm and occurs in areas where there is a superficial striated muscle, as the nose, chin, periorbital and anterior neck areas and have distinctive histological characteristics (Chang and Chen, 2005; *Solis-Coria et al.*,2007; *Saliba et al.,* 2012). They may be solitary, independent lesions or multiple and associated with other congenital abnormalities (*Sahn et al.,* 1990; *Takeyama et al.*, 2005). The case of 21 year old women with RMH of nasal dorsum is presented.

Case report

A 21 year –old female apply to YSMU "Heratsi"#1 hospital for purpose of rhinoplasty with complaints of nose esthetic defect (fig. 1). She had Caucasian nose and noticed, that the dorsal area of the nose had grown gradually without any other symptoms approximately 6 years. She had a history of polycystosis 3-4 year earlier and undergoes some hormonal treatment by gynecologists.

Upon physical examination, we found a subcutaneous mass, 20 × 10 mm in size at its widest point at the dorsal nasal area. The mass was painless, elastic-soft without tenderness. The skin was a little hyperemic. Based on the clinical manifestation, the primary diagnostic impression was a dermoid cyst.

The rhinoplasty with simultaneous tumor-like lesion enucleation was planned.

Under general anesthesia a lesion was surgically removed via open rhinoplasty incision (fig. 2a, b). Incision revealed a tumor, which involving the dermis, subcutaneous tissue and muscles. It was reddish-white and meaty in texture and there was no evidence of cysts (fig.2 c). Open rhinoplasty was perform after tumor removing (fig. 2 d,f).

Tissue samples were fixed in 10% buffered neutral formalin. On histological examination with routine haematoxylin-eosin, it showed a disordered collection of bundles of mature striated muscle fibers arranged in a haphazard manner and interspersed with adipose tissue, fibrocytes and collagen, and had a eodematous matrix, mitotic figures were typically rare or absent altogether (fig. 3). On immunohistochemical examination mature striated muscle was positive for desmin, adipose tissue and nerves were positive for S-100 protein, and fibrocytes and collagen were positive for vimentin (fig. 4 a,b,c). The clinical, macroscopic, histologic and immunochemical characteristics allowed diagnosis of rabdomyomatous mesenchymal hamartoma.

After 3 weeks follow-up examination the patient was satisfied with her facial esthetic results and had no any complains (fig.5).

Discussion

RMH is a rare benign dermal or subcutaneous lesion comprising disordered mature adipose tissue, skeletal muscle, adnexal elements, nerve bundles and collagen (*Rosenberg et al.,* 2002; *Ortak et al*., 2005). RMH occurs most commonly in areas where there is superficial striated muscle , mainly on the middle of the head and neck including forhead, nose,upper lip, chin, followed by the periorbital and anterior neck areas (Elgart and *Patterson,* 1990; Wang *et al.,*2014). They typically appear as a single polypoid, pedunculated or popular cutaneous lesion or subcutaneous swellings as in our case, but they can also occur as multiple lesions or as a plaque-type variant (Magro et al., 2005; Chang and Chen, 2005; Fontecilla *et al*., 2016). As *Saliba et al.* (2012) noted, there have been 48 cases of RMH reported in the literature and, nonetheless, it remains a rare condition poorly known to clinicians with an unclear etiology (*McKinnon et al*., 2015). However, RMH is believed to result from an abnormality in the migration of mesodermal tissue during embryogenesis or from genetic defects, as it is associated with other congenital defects like the amniotic band syndrome, Delleman syndrome, and Goldenhar syndrome (*Sahn et al.,* 1990; *Solis-Coria et al.,* 2007; *McKinnon et al*., 2015).

In most reported cases the lesion was described in newborns and children, and only several (eleven cases) were reported in adults (*Díaz-Pérez et al.,* 2008; *Saliba et al.,* 2012).

Clinically, these lesions are often mistaken for skin tag, fibroepithelial polyp, lipomatous nevus, dermoid cyst or accessory tragus (*Elgart and Patterson*,1990; *Rosenberg et al.,2012*). Our patient has not any complaints regarding of existing lesion, as she never know about it. Her main complain was her typical Caucasian nose (very common in Armenians), so she apply for esthetic rhinoplasty.

Treatment is local surgical excision, with the generally accepted notion that RMH has no tendency to recur after removal (*Saliba et al.,* 2012). The presented case is interesting since we didn't find any case in literature where lesion surgical removal and rhinoplasty were done simultaneously.

Microscopically these lesions are constituted by striated muscle fibers randomly distributed, intermingled with mature adipose tissue, collagen bands, blood vessels, and elastic fibers.

IHC in RMH shows positivity for desmin, S-100 protein and vimentin.

CONCLUSION

RMH is a rare benign congenital lesion, which typically presenting in the midline of head and neck region. In case of nasal dorsum localization one-stage removal of lesion and rhinoplasty is possible.

CONFLICT OF INTEREST

There is no conflict of interest that has to be indicated by the authors.

Figure 1. Patient before operation

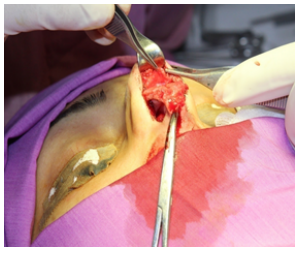

Figure 2a

Figure 2b

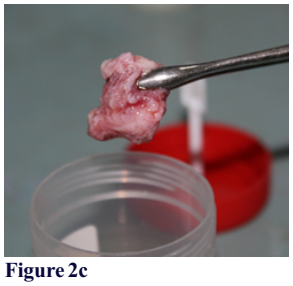

Figure 2c

Figure 2d

Figure 2 f

Figure 2: Operation steps: (a, b) excision of lesion by open rhinoplasty incision access, (c) macroscopic view of excited mass, (d, f) rhinoplasty performing

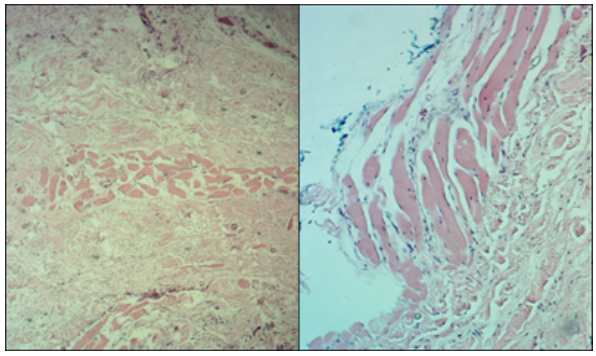

Figure 3. Histological examinations by haematoxylin-eosin

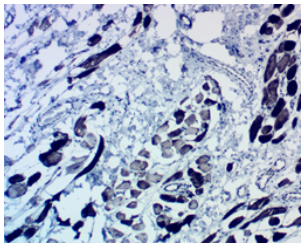

Figure 4 a

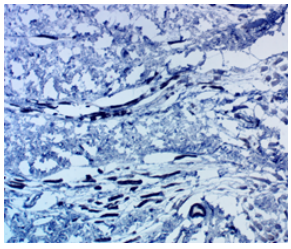

Figure 4 b

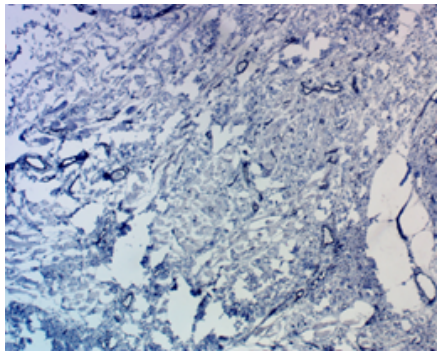

Figure 4 c

Figure 4: Immunohisochemical examination: (a) Desmin, (b) S-100 Protein, (c) Vimentin

Figure 5. Patient after 3 weeks follow-up

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