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Post-traumatic lip lesion mimicking rhabdomyomatous mesenchymal hamartoma in a pediatric patient

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Abstract

We report a 7-year-old boy who presented with a nodule on the upper lip. A previous clinical history of mechanical trauma in the lesional area had been noted. After surgical excision, microscopy revealed fibrocollagenous fascicles associated with neurovascular bundles and skeletal striated muscle fibers in diffuse subepithelial distribution, suggesting rhabdomyomatous mesenchymal hamartoma. However, strict clinicopathological correlation favored a healing process with trapped striated skeletal muscle tissue. After three years of follow-up, an improvement in the aesthetic appearance of the upper lip was observed. To the best of our knowledge, a case of pseudo-rhabdomyomatous mesenchymal hamartoma has not been reported to date.

Keywords: pediatric dentistry, lip, hamartoma, oral diagnosis, immunohistochemistry

Introduction

Rhabdomyomatous mesenchymal hamartoma (RMH), also known as hamartoma of the congenital midline or hamartoma of striated muscle, is a rare congenital lesion microscopically characterized by skeletal striated muscle in the superficial dermis admixed with adipose tissue, nerves bundles, and adnexal elements. It was first described as "striated muscle hamartoma" by Hendrick et al. (1986). Three years later, these skin lesions were renamed RMH [1-4].

Rhabdomyomatous mesenchymal hamartoma occurs mainly in newborns and children, with no predilection for gender. It is commonly described affecting soft tissue of the head and neck region, especially of the chin, nose, forehead, and oral cavity. To date, approximately 5 cases of intraoral RMH (3 of tongue, one of upper lip, and one of lower lip) have been reported [1,5].

The etiology of RMH remains unknown. However, it has been suggested that changes associated with embryogenesis or genetic factors may promote its formation. The clinical presentation of RMH is varied, being commonly polypoid papules or tags on the skin, usually asymptomatic. Owing to its various clinical presentations, histopathological analysis is important for diagnostic confirmation of RMH [1-5].

Case Synopsis

A 7-year-old boy was referred to our clinic complaining of swelling on the right side of the upper lip, with a time of evolution of one year. The history revealed that the patient suffered an accident with facial trauma two years prior, involving the intrusion of the deciduous maxillary central incisor and laceration of the upper lip. The patient's mother stated that the child did not have any type of visible changes on the lips before the accident. The medical history was noncontributory. During the clinical

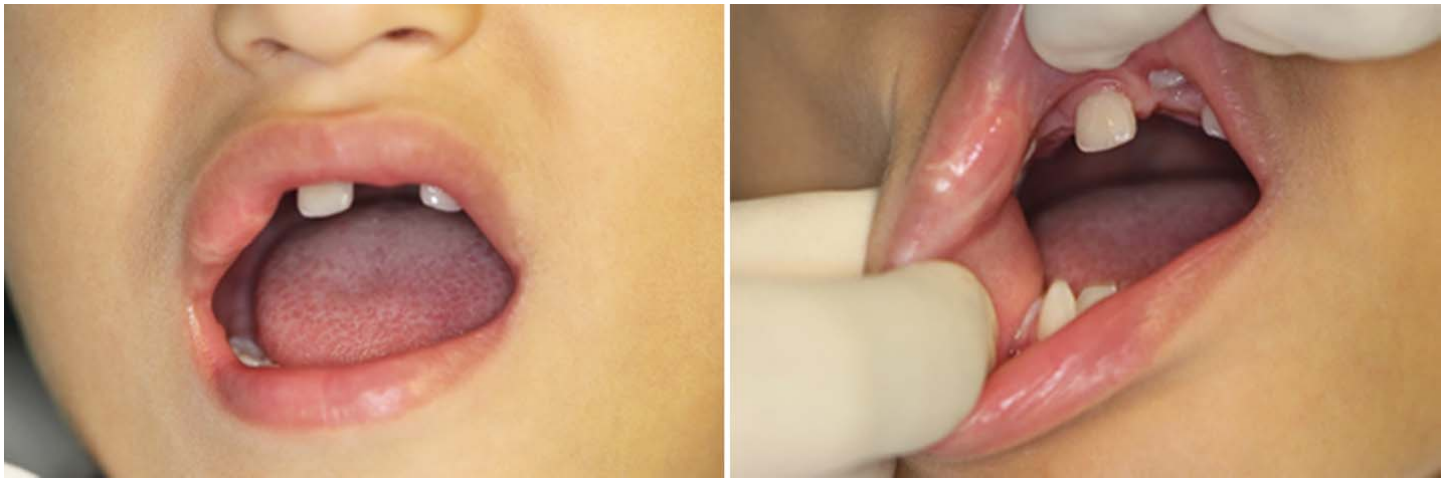

Figure 1. *Clinical examination showing a nodule on right side of the upper lip.*

examination, a nodule with firm consistency on the right side of the upper lip was exhibited (**Figure 1**). The main clinical hypothesis was that it was related to the healing process. However, other reactive or benign (mesenchymal or epithelial origin) proliferations could not be totally excluded. Thus, the nodule was surgically removed under local anesthesia.

Histopathologically, the growth showed a large area of fibrous connective tissue associated with neurovascular bundles and skeletal striated muscle tissue in a diffuse subepithelial distribution. At the periphery, a stratified squamous epithelial lining was observed (**Figure 2A**, **B)** The Masson trichrome stain (**Figure 2C)** and desmin immunostaining (**Figure 2D)** highlighted bundles of skeletal striated muscle trapped by fibrous stroma. The diagnosis of a healing process mimicking RMH was made.

After 3-years of follow-up the patient is well, without persistent clinical alterations in the lesional area.

Case Discussion

Rhabdomyomatous mesenchymal hamartoma is a rare congenital lesion characterized by dermal and subcutaneous tissue abnormalities. To date, approximately 46 RMH cases have been reported in the literature. The data indicate that 26 were male and 20 were female. Most RMH cases were seen affecting newborns (10 cases) and children (29 cases, mean age, 5 years). However, it can also be observed in adult patients (7 cases, mean age, 52 years), [1,4,5]. Rhabdomyomatous mesenchymal hamartoma shows preferential involvement of the chin, nose, periorbital region, tongue, lip, and sternoclavicular area. However, other sites including thorax, anus, and vagina have also been reported. On clinical examination, RMH can be visualized as a polypoid lesion, papule, plaque, or "skin tag." The differential diagnosis includes fibrous hamartoma of infancy, benign Triton tumor, nevus lipomatosus superficialis, rhabdomyoma, and cutaneous embryonal rhabdomyosarcoma. Although the etiology of RMH is unknown, it has been suggested that changes in embryogenesis or genetic factors may promote its formation. In this pathway, congenital anomalies, such as amniotic band syndrome, Dalleman syndrome, and Goldenhar syndrome, have been reported to be associated with RMH [1,2,4,5].

The treatment of choice for RMH is surgery, regardless of its location. In general, there is no occurrence of postoperative complications or clinical alterations. However, clinical regression in post-biopsy or follow-up without surgery have also been observed in some RMH cases, suggesting that strict clinical follow-up should be performed [1,3,5].

Conclusion

In the current case, the clinical history indicated that the patient suffered an accident with facial trauma. After two years, a nodule on the right side of the upper lip was detected, suggesting also the

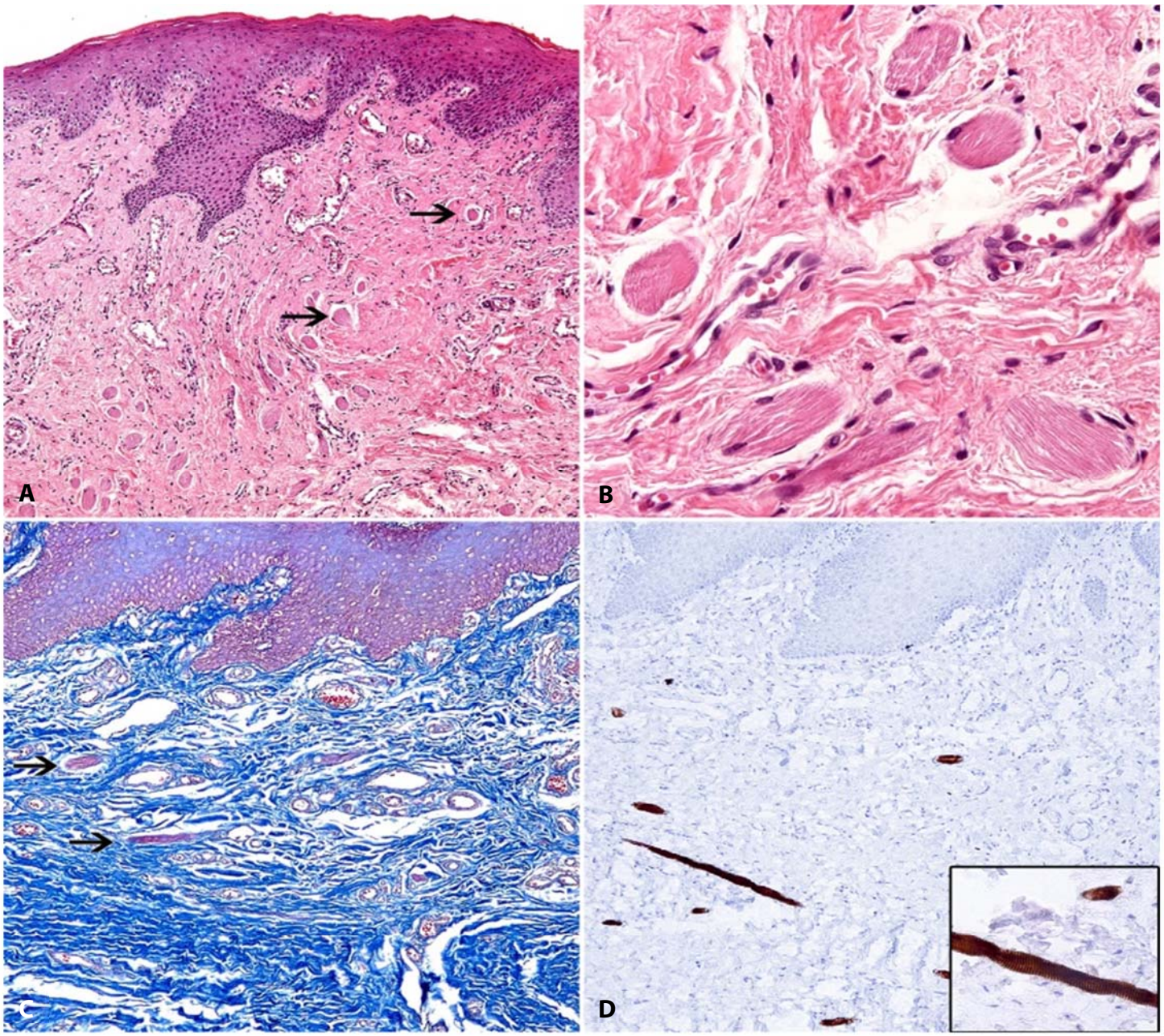

Figure 2. ***A)*** *Histopathological analysis exhibited the presence of small bundles of skeletal striated muscle (arrows) surrounded by a fibrous stroma. At the periphery, stratified squamous epithelium lining. H&E, 10×.* ***B)*** *Close-up view showing skeletal striated muscle bundles. H&E, 40×.* ***C)*** *Masson trichrome stain highlighting skeletal muscle fibers (arrows) associated with bundles of stromal collagen, 40×.* ***D)*** *Desmin immunopositivity for skeletal striated muscle,10×; inset, 40×.*

possibility of a neoplasm. After histopathological analysis, a diagnosis of RMH was suspected, especially considering the age of the patient. However, strict clinicopathological correlation favored a healing process showing RMH-like features. In fact, to date, two RMH cases affecting the upper and lower lip have been reported [1,5], which should be distinguished from its mimics, such as reported in the current case.

Potential conflicts of interest

The authors declare no conflicts of interests.

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