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Answer

Rhabdomyomatous mesenchymal hamartoma presenting as a polypoid lesion of the nasal skin in a child: answer

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Keywords

Hamartoma, skin, nose, genetic defects, striated muscle fibers.

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Answers

1. The histological picture is characterized by the presence of skeletal muscle fibers and adipose tissue are suggestive for hamartoma.
2. Actin muscle specific immunohistochemical stain, to confirm the presence of skeletal muscle fibers.
3. Differential diagnosis with rhabdomyoma.

Introduction

Rhabdomyomatous mesenchymal hamartoma (RMH) is a rare congenital lesion of the dermis and soft tissue [1], first described in 1986 as a striated muscle hamartoma [2]. It has been reported under various names: striated muscle hamartoma, congenital midline hamartoma, hamartoma of cutaneous adnexa and mesenchyme [3].

Etiology of this lesion is unknown; it has been hypothesized that be due to an abnormal migration of mesodermal stem cells during embryiogenesis or to right genetic defects [3, 4]. Patients with RMH occasionally have other congenital defects [5].

RMH usually presents as a polypoid or papular cutaneous 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 [6].

Here we report a case of RMH in a 2-year-old child presenting with a congenital polypoid mass on the nasal skin.

Pathological findings

Histologically the lesion was covered by a normal squamous epithelium. Mature striated muscle fibers was found within the dermis extending into the subcutaneous tissue. The fibers were admixed with fibrous connettive tissue, mature adipose tissue, adnexal structures. The striated fibers arranged perpendicular to the surface of the skin. There was no evidence of malignity [7-9].

Discussion

RMH is a rare benign lesion of the deep dermis and subcutaneous fat in the region of the head and neck [1, 10, 11]. The etiology of this condition is unknown. Cases of RMH have been reported to be associated with other genetic defect such as amniotic band syndrome, Delleman syndrome and the Goldenhar syndrome [4, 10, 12].

Distinctive characteristics of these lesions are striated muscle fibers aligned perpendicular to the surface epithelium, surrounded by connective tissue and adipose tissue. Actin muscle specific immunohistochemical stain must be performed to confirm the presence of fiber skeletal muscle (**Fig. 1**) [13].

The differential diagnosis includes rhabdomyoma, nevus lipomatosis superficialis, fibrous hamartoma of infancy, neuromuscular choristoma and cutaneous embryonal rhabdomyosarcoma [13]

Complete excision is curative [13].

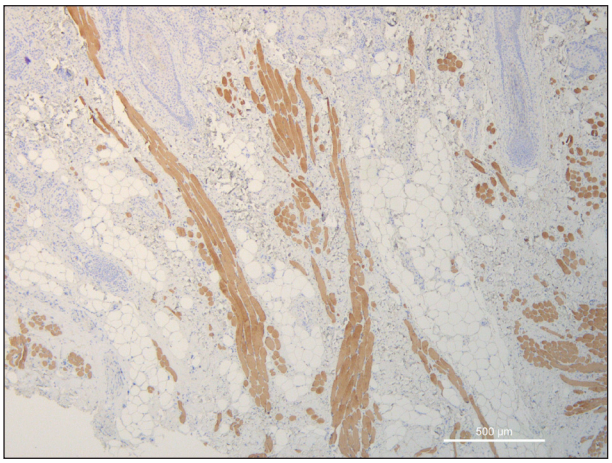

Figure 1. Actin muscle specific himmunoistochemical showed striated muscle fibers aligned perpendicular to the surface epithelium, surrounded by connective tissue and adipose tissue.

Declaration of interest

The Authors declare that there is no conflict of interest.

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