

## Nasal congenital fibrolipomatous hamartoma in a premature infant

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Hamartomas are tumor-like lesions composed of tissue elements normally found at the site where they are located, but which grow in a disorganized manner. They generally occur at birth or soon after, although presentations during adult life have been reported. Hamartomas of the head and neck are very rare. The exact mechanisms behind the development of hamartomas remain unknown; however, their benign nature renders complete surgical excision sufficient for their management. Here, we describe a case of a premature infant with a hamartomatous polypoid lesion containing both fibrous and adipose components, originating from the dorsum of the nose. The differential diagnosis and management of hamartomas of the head and neck are also discussed. Although several cases series on precalcaneal fibrolipomatous hamartomas have been encountered in the literature, to the best of our knowledge, this is the first report of a nasally located congenital fibrolipomatous hamartoma.

*Key words:* fibrolipomatous hamartoma, nose, premature infant.

Hamartomas are tumor-like lesions composed of tissue elements normally found at the site where they are located, but which grow in a disorganized manner. A hamartoma may contain several different tissue components with mature cellular elements. They are believed to be benign tumor-like malformations that manifest during normal tissue development<sup>1</sup>. This entity was first described by Albrecht in 1904<sup>2</sup>, and is widely believed to be a congenital condition that manifests itself during the pubertal period. Hamartomas of the head and neck are very rare<sup>3</sup>.

Congenital fibrolipomatous hamartomas (FLH) are rare hamartomas generally located in the precalcaneal area<sup>4,5</sup>. Here, we describe a case of a preterm infant with a congenital FLH, made interesting by the rare presentation of the lesion in the nasal area.

### Case Report

A preterm male infant was born in our hospital to a mother with preeclampsia after 33 weeks

of gestation. His birth weight was 1470 g, with 1<sup>st</sup> and 5<sup>th</sup> minute Apgar scores of 6 and 8, respectively. The presence of a pedunculated lesion originating from the upper edge of the right nostril, roughly 1.5 cm in size, covered in skin and occluding the right naris was immediately apparent on physical examination (Fig. 1a). Findings on an abdominal ultrasound and a cranial computerized tomography performed to identify the presence of other anomalies were normal. The nasal dorsal lesion was excised one month later (Fig. 1b) with the parents' consent.

Histopathological examination of the polypoid lesion with hematoxylin-eosin stain revealed the presence of normal-appearing epidermis and a dermal layer with hair follicles and sebaceous and sweat glands. The underlying layer contained fat tissue, vascular structures and bundles of muscle fibers, which stained negative following immunohistochemical assay with anti-human melanoma (HMB-45). Morphological findings were considered to be consistent with a benign FLH (Fig. 2). The

patient was discharged on the same day of the procedure. No complication was observed



Figure 1. The pedunculated hamartomatous polypoid lesion originating from the dorsum of the nose (a). The macroscopic appearance of the excised lesion (b).

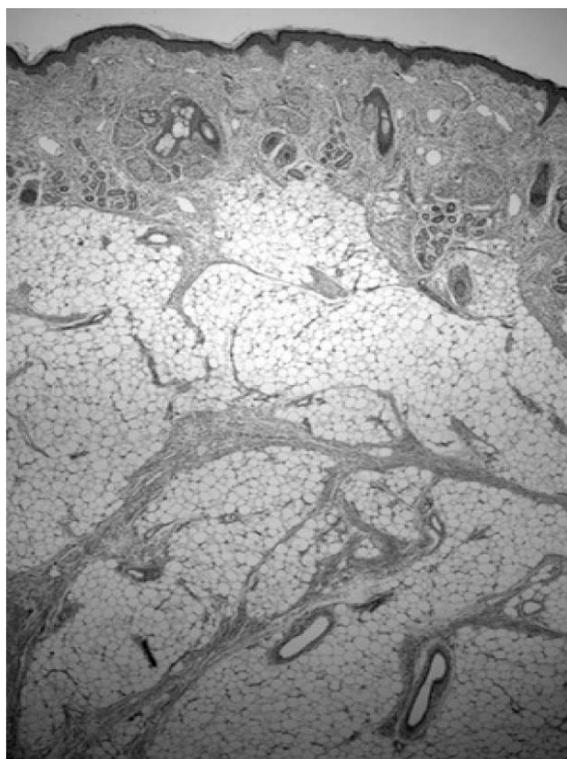


Figure 2. Microscopic appearance of the excised lesion showing lobulated mature adipose tissue protruding into the dermis and separated by collagenous fibrous sheaths (H&E x40 magnification).

in the postoperative period, with no signs of relapse after four months of follow-up.

### Discussion

Hamartomas are non-neoplastic tumor-like

malformations that may occur during normal tissue development. Unlike teratomas and dermoid cysts, hamartomas are overgrowths of normal mature tissue, which occur within the normal organ or tissue of origin. Further growth ceases after full cellular maturation is achieved<sup>6</sup>. The presence of both mature fat and fibrous components in the lesion excised from our patient was a finding consistent with a hamartoma, or more precisely a fibrolipomatous lesion of hamartomatous origin.

The presence of fibrous proliferation in infants and young children was originally described in 1956 by Reye<sup>7</sup>, and was later named as “fibrous hamartoma (FH) of infancy” in 1965 by Enzinger<sup>1</sup>. A metaanalysis involving 197 cases of FHs revealed a male predominance of 70%, with 91% of lesions manifesting within the first year of life. A FH was observed at birth in 28 patients (23%). The most commonly reported locations were the axillary region, upper arms, upper trunk, inguinal region, and external genital areas. Nasal FHs are very rare, with an estimated frequency of 0.5%<sup>8</sup>. Our patient had a FLH located in the nasal area, which shared many histopathological similarities with FH.

Most of the case reports in the literature are of congenital FLH located in the precalcaneal region<sup>5,9</sup>. The etiology of precalcaneal FLHs is unclear, although several hypotheses have been proposed. It has been suggested that an idiopathic alteration in the involution of the plantar subcutaneous tissue in the final month of pregnancy and the first months of life may lead to herniation of fat into the subcutaneous tissue<sup>10</sup>. Although several case series on precalcaneal FLHs may be encountered in the literature, to the best of our knowledge, this is the first report of a nasally located congenital FLH.

In conclusion, congenital FLH is a rare benign hamartomatous lesion with a peculiar histology of cosmetic rather than clinical significance. It is readily treatable by excision alone, and no recurrences have been reported after surgical removal of such lesions.

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