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CONGENITAL NASAL FIBROLIPOMATOUS HAMARTOMA – A CASE REPORT

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ABSTRACT
Hamartomas are tumor like growths composed of tissue components growing in an irregular manner normally found at the site where they are located. The exact mechanisms being not known, nasal hamartomas generally are reported in premature infants and are mostly composed of chondromesenchymal components. We describe a rare case of a full term infant presenting with a hamartomatous lesion in the left nasal cavity composed of adipose tissue and fibrous elements.

INTRODUCTION

Nasal mass in infancy is a relatively rare entity. It may be a developmental anomaly or a neoplasm. The former includes encephalocele, nasolacrimal duct cyst, hamartomas & gliomas while the latter are mostly dermoid cysts or teratomas [1]. Most of the paediatric neoplasms of head and neck region are benign in nature, the malignant constituting only 5% of them [2]. Unlike neoplasms, hamartomas are tumor like benign growths composed of tissue elements normally found at site where they are located but which grow in an irregular manner. First described by Albrecht in 1904, hamartomas are formed during normal tissue development [3]. Literature reveals nasal hamartomas to be mostly composed of chondromesenchymal components. Till date only 30 cases of nasal chondromesenchymal hamartomas have been reported following its first description by McDermott et al in 1998 [4]. We describe a rare case of a term infant having a fibrolipomatous hamartoma in the nasal area. The uniqueness lies not only in its rarity and age of presentation but also in the diagnostic dilemma encountered which can have serious implications in its management.

CASE REPORT

A 7 day old full term male infant presented with smooth mass occluding the left nostril and overhanging out of it [Fig. I]. History was inconspicuous with the child having normal birth weight, being delivered by normal vaginal route and cried immediately after birth. Nasal endoscopy revealed a broad based mass in the left vestibule which was attached medially to the membranous septum. Posterior part of nasal cavity revealed normal anatomy. CT brain and ultrasound of the abdomen was performed to look for other anomalies which showed normal study. The mass was removed by simple surgical excision the next day after taking parents' consent.

Histopathological study revealed hair follicles, sebaceous and sweat glands embedded in the dermal layer with normal looking epidermis [Fig. II]. Underlying layer consisted of anti-human melanoma (HMB-45) negative fat, vascular tissue, & muscle fiber bundles [Fig. III]. The morphological picture was pretty in favour of benign fibrolipomatous hamartoma. Child was discharged the following day and the post-operative period was

uneventful [Fig IV]. Follow-up visits showed no signs of relapse or any other complication.

DISCUSSION & CONCLUSION

Tumors in the sinonasal region are predominantly epithelial in origin. Whether of epithelial or mesenchymal origin, presentation of nasal masses in infants is very rare. These hamartomatous lesions may be angiomatous, chondroid, neurogenic, lipomatous or epithelial depending on the dominant tissue present [5,6]. Unlike teratomas and dermoids, hamartomas are proliferations of normal mature tissue which occur within the normal tissue of origin. The growth stops after cellular maturation is achieved [7].

An initial clinical diagnosis may become daisy. Although radiological investigations including CT and MRI are crucial for finding the tumor's site of origin, texture, and extension into surrounding structures but a detailed histopathological examination remains the mainstay. The diagnosis of fibrolipomatous hamartoma was arrived at because of presence of both fibrous components and mature fat in the mass. Enzinger in 1965 coined the term fibrous hamartoma of infancy although the original description of fibrous proliferation in infants was given by Reye in 1956 [8,9]. A study on 197 cases of fibrous hamartomas showed a male preponderance of 70% with 23% cases presenting at birth. The pre-calcaneal region constitutes the most common presentation site for congenital fibrolipomatous hamartomas [10]. They may also be present in axillary region, forearms, inguinal region and in external genital regions. Nasal presentation has a reported frequency of 0.5% only [11].

Thus, nasal fibrolipomatous hamartomas constitute a less commonly encountered but well known clinical entity which can be easily treated with surgical excision without any recurrence.

Figure I – A 7 day full term infant presenting with a mass in left nostril

Figure II – Histopathological slide showing hair follicles, sebaceous and sweat glands embedded in the dermal layer with normal looking epidermis.

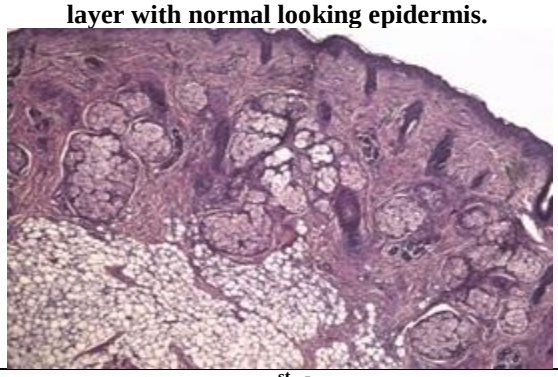

Figure III – Underlying layer consisted of anti-human melanoma (HMB-45) negative fat, vascular tissue, & muscle fiber bundles.

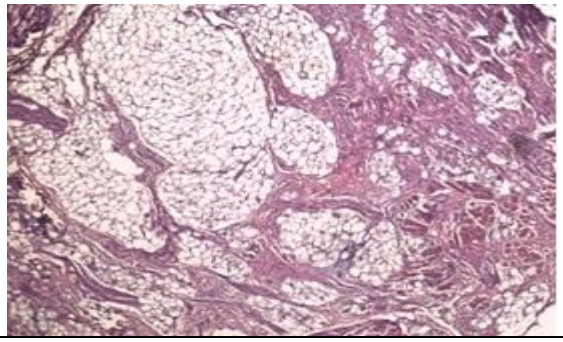

Figure IV – 1st day post-op

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CONFLICT OF INTEREST: NIL

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

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