REAH : Unusual Case of Unilateral Nasal Blockage

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Introduction

Hamartomas are non-malignant malformations or inborn errors of tissue development. They result from excessive proliferation of otherwise normal local tissue components. Hamartomas of the nasal cavity and paranasal sinuses are relatively rare, especially respiratory epithelial adenomatoid hamartomas (REAH). This benign lesion is characterized by a glandular proliferation originating from the surface of the respiratory epithelium [1].

Case Report

A 52 year old male patient reported with history of unilateral nasal blockage of left side of one year duration with left sided frontal headache. There was no history of discharge from nose or epistaxis. Examination revealed a 2 x 1 cm firm swelling with smooth surface 0.5 mm behind the mucocutaneous junction of lateral wall of left nasal cavity. Endoscopic examination of the nasal cavity as well as the nasopharynx was normal. Radiograph of paranasal sinuses showed soft tissue density in the left nasal cavity, the sinuses were normal and there was no evidence of bony erosion. The mass was excised completely under local anaesthesia.

Gross examination of the specimen showed a 1.5 cm pale coloured fleshy rounded firm mass with smooth surface. There were no areas of haemorrhage or altered texture on the cut section. Histopathological examination by routine haematoxyline and eosin stained sections showed a well encapsulated mass with compressed parenchyma. It comprised of lobular adenomatous proliferations with tubular glands showing ciliated columnar epithelium at places. Some of the glands showed large amounts of secretions. Goblet cells were seen dispersed among the lining epithelium of glands. The stroma showed hyalinization and a focal chronic inflammatory infiltrate. No atypia or increased mitotic figures were observed (Fig.1). The lesion was diagnosed as respiratory epithelial adenomatoid hamartoma (REAH).

Discussion

Hamartoma is a rare, non-neoplastic lesion

Fig. 1 : REAH showing adenomatous proliferations surrounded by hyalinized stroma (H&E X 100)

characterized by an abnormal mixture of tissues, which are indigenous to the region. Hamartomas commonly originate from the lung, kidney and intestine. Glandular hamartomas involving the sinonasal tract have received only limited documentation in the literature.

In the upper aerodigestive tract, respiratory epithelial adenomatoid hamartoma (REAH) is described as a polypoid proliferation of glands lined by ciliated respiratory epithelium that seem to invaginate downward into the submucosa while maintaining direct continuity with the surface mucosa [2]. Wernig et al [3], reported 31 cases of what they called “Respiratory epithelial adenomatoid hamartoma of the sinonasal tract”. The median age was 58 years (range 27-82 years) and men were primarily affected.

The etiology of the lesion is unclear and may be secondary to either sinonasal inflammation or developmental error. Respiratory epithelial adenomatoid

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hamartomas of the nasal cavity are usually associated with nasal polyposis. This association supports the hypothesis that inflammation is one of the factors that induces the development of a hamartoma [4].

Within the nasal cavity, the most common site is nasal septum along its posterior aspect [2,3,5]. Other areas in the sinonasal tract where the lesion has been documented are maxillary sinus [6-8], ethmoid sinus [6] and the nasopharynx [9]. The lesion in our patient was found to have arisen from the anterior part of lateral nasal wall a few millimeters behind the mucocutaneous junction. Involvement of the anterior part of lateral wall of nasal cavity is an unusual localization and no other report of a respiratory epithelial adenomatoid hamartoma involving this area was found in the literature.

Histologically the lesions are characterized by a prominent tubular glandular proliferation lined by ciliated respiratory epithelium, originating from the surface epithelium, stromal hyalinization which envelops the adenomatous proliferation, and mucous cell metaplasia. The basement membrane is thickened. No destructive growth is noted [3,5,9]. The differential diagnosis includes inflammatory polyps (fewer glands) and inverted schneiderian papillomas (which have a hyperplastic squamous epithelium with only a few goblet cells and a thin basement membrane) [10]. More important is the differential diagnosis of sinonasal low grade adenocarcinoma, which usually demonstrates a back to back glandular pattern with nuclear atypia, prominent mitotic activity, desmoplastic stroma and perineural invasion [5]. Diagnostic misinterpretations may result in untoward surgical intervention. Limited but complete surgical resection is the treatment of choice and surgery is curative with no recurrence [3,5].

Conflicts of Interest

None identified

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