

Clinical Photograph: Nasopharyngeal Hamartoma

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Hamartoma is a benign developmental malformation. Nasopharyngeal hamartoma is an uncommon lesion. We describe a clinical case of nasopharyngeal hamartoma presented with airway obstructive symptoms. Apparent patent nasal and nasopharyngeal airway delayed the diagnosis. The clinical features, pathology and management are discussed.

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THE CASE

A 3 month old female patient presented with a history of recurrent attacks of choking, cyanosis and cough since birth. Sleeping and crying were normal. The patient was born full term by normal delivery.

Clinically, the patient was stable, no respiratory distress, no dysmorphic features and growth parameters were normal. Systemic and otolaryngological examination were unremarkable. The initial assessment on admission was done by the paediatrician with possible diagnosis of tracheoesophageal fistula, laryngotracheal cleft, gastroesophageal reflux and vascular ring. Nasogastric tube was inserted. Chest X-ray, barium study, nuclear study and echocardiogram were requested and were found normal. CT scan of head showed homogenous, non-enhanced soft tissue mass arising from nasopharynx. No invasion or intracranial extension. Endoscopy was normal but the examination of the nasopharynx under general anesthesia showed a pedunculated non-pulsating 2 x 3 cm mass arising from the left side of the nasopharynx with intact mucosa (Fig 1).



Figure 1. Nasopharyngeal mass. Soft palate and uvula (small arrow) pulled anteriorly for better exposure

The mass was excised. Microscopically, stratified squamous epithelium was covering the mass. There was a core of hyaline cartilage surrounded by various components of epithelial tissue in fibrovascular stroma without any adenexial structures consistent with epithelial-predominant hamartoma.

After excision, the patient improved and there was no recurrence.

DISCUSSION

The definition of hamartoma can be found in its Greek root, "hamartia" - literally, a defect or error - and the suffix - "oma" referring to a tumour-like growth. Hamartoma is used to designate a self-limited, non-neoplastic malformation composed of an excess admixture of mature cells and tissues that are indigenous to the affected part, often with one element predominating¹. Nasopharyngeal developmental masses are divided into three groups: (a) teratomas; (b) dermoids or teratoids; and (c) hamartomas. The first category, teratoma, which is the most clearly neoplastic, is derived from all the three germ cell layers and containing tissue that may be foreign to the area. In head and neck, teratomas can take the form of an "epignathus", occasionally with well formed organs and limbs of a parasitic fetus². Other congenital anomalies are usually associated with teratomas. The second category, dermoids or teratoids, contains masses that are formed from skin and its appendages, cartilage, bone muscle and salivary glands and form the group known as "hairy polyp". These occur most commonly in infancy, and have been considered hamartomatous in the area¹⁻³. The last group, nasopharyngeal hamartomas, are uncommon and can be divided into two types based on histologic presentation. The first and common type is composed predominantly of mesodermal tissues. The less common second type, the epithelial-predominant type, contains a spectrum of squamous and cuboidal epithelial ducts and serous/mucinous exocrine gland in addition to various stroma elements¹. Possible cases have been reported that included elements not indigenous to the region, these elements being considered metaplastic, such as keratinizing squamous epithelium, bone, cartilage and parathyroid^{2,4}. The growth pattern, lack of atypia and lack of expansile growth or true infiltration all support a benign diagnosis². There is no specific anatomic location from where they arise¹. Respiratory epithelial adenomatoid hamartoma is a term coined by Wenig and Haffner to describe a group of glandular hamartomas characterised by glandular proliferations which are entirely derived from the nasopharyngeal surface epithelium and not related to the minor salivary (seromucous) glands⁵.

Clinically, nasopharyngeal hamartomas present either with obstructive or infectious symptoms early in life or alternately remain asymptomatic to be discovered later in life². In the

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infant, differential diagnosis includes: enlarged adenoid, meningoencephalocele, nasal glioma, hemangioma and choanal atresia.

Birth and Knight-Jones described a similar case to ours and stressed on the point that easily passed nasotracheal or nasogastric tube through the nostrils is not a guarantee of physiological patency and careful examination of the nasal part of the airway in an infant with unexplained respiratory distress or feeding difficulties is advised⁴.

Nasopharyngeal hamartomas can be surgically excised with no history of recurrence¹⁻⁵.

CONCLUSION

Nasopharyngeal hamartoma is uncommon benign developmental lesion and should be considered in the differential diagnosis of nasal and nasopharyngeal mass. Careful examination of the nose and nasopharynx should

be applied to the patients with noisy breathing or respiratory distress symptoms to detect any abnormalities and to avoid unnecessary investigations.

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