Base of Tongue Hamartoma—an unusual cause of airway obstruction

Case report

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Case report

History:

A 12 year old Yemeni girl presented to the ENT outpatient clinics with history of aspiration and choking at night with difficulty of breathing and recurrent sleep apnea for 3–4 years.

Her symptoms started as mild throat discomfort and progressed over weeks to repeated choking episodes and apneic attacks during sleep. Initial consultation at a private hospital was sought by the patient where a diagnosis of a small oro pharyngeal mass was made. Patient did not seek active treatment until after 6 months when she felt difficulty in swallowing and increased attacks of sleep apnea. There was no history of any bleeding per mouth.

On GPE, the patient looked well. ENT examination revealed nasal turbinate hypertrophy, normal ear examination and oro pharyngeal examination revealed a pinkish smooth surfaced pedunculated mass, arising from the base of the tongue, about 1.5 cm in height. An initial diagnosis of base of tongue lipoma or papilloma was entertained. All hematological examinations were within normal parameters and radiological examination of the sinuses revealed only inferior turbinate hypertrophy bilaterally. T_c 99m Thyroid scan showed a normal uptake by both thyroid lobes and the oro pharyngeal mass was not visualized.
The patient was admitted and underwent EUA of oropharynx and hypopharynx with excision of oral mass from the base of the tongue. Peroperative findings were a pedunculated mass arising from the midline of the base of tongue and going to the nasopharynx. (fig. 1). No other masses, ulcerations were seen in the area. The oral mass was excised completely and sent for histopathological examination. HPE result was ‘mild dilatation of salivary gland duct and focal non-specific inflammation with abundant benign adipose tissue’.

Discussion

Vascular tumors of the head and neck region comprise a heterogeneous group of lesions that have very different histology, presentations, clinical courses and treatment options. The difficulties inherent to the study of such a varied group are compounded by the fact that present literature contains a multitude of classification schemes and obscure terms. 

Benign oral cavity growths have been classified variously in literature and a quick review reveals that hemangiomas are the most common congenital lesion in man and occur in approximately two to three percent of infants. Hemangiomas are actually hamartomas of blood vessels, not true neoplasms, and are often divided into three subgroups based on the size of the vessels. These are Capillary (strawberry) hemangiomas, Cavernous hemangiomas and the final subgroup that of mixed hemangiomas which are comprised of blood vessels of various sizes. 

Hamartomas of the oral cavity are uncommon lesions that show a variety of clinical presentations, histological appearances and growth patterns. They have been described as benign tumor like malformations resulting from faulty development in an organ and composed of an abnormal mixture of tissue elements that develop and grow at the same rate as normal elements but are not likely to compress adjacent tissue. Hamartomas have also been defined as non-neoplastic developmental anomalies that appear as simple spontaneous growths composed of a mixture of locally derived mature tissue. 

Capillary (strawberry) hemangiomas are by far the most common and are more frequent in females. They are usually red or pink in color and are often
papular with a lobular surface. An often alarming rate of enlargement occurs until the child is ten to twelve months of age at which time the lesions usually stabilize and eventually regress with few remaining by seven years of age. Capillary hemangiomas involving the larynx usually occur in the subglottis and are found mainly in the pediatric population. These lesions typically present in a previously healthy infant who develops biphasic stridor during the first few months of life. As with other pediatric airway problems, the child may be initially referred to the otolaryngologist for a further work up of recurrent episodes of croup. At endoscopy, the lesions are usually located in the posterolateral subglottis and are submucosal, soft, compressible masses. Because the overlying mucosa is normal, many of these hemangiomas will not display the usual red or pink color of a cutaneous lesion. Pediatric subglottic hemangiomas generally behave like their cutaneous counterparts with eventual regression occurring in most cases.

Cavernous hemangiomas also are more frequently found in females and are soft, compressible, often poorly defined lesions. They tend to involve deeper structures and many do not regress.

The final subgroup is mixed hemangiomas which are comprised of blood vessels of various sizes.

A hamartoma may occur in any organ, but it most often involves the gastrointestinal tract. Its biological behavior is benign. The reason for development of the tumor is still unclear. Maybe, certain genetic factors during pregnancy might play a role.

A review of the literature indicates that epithelial and mesenchymal hamartomas are uncommon in the head and neck. They have been described in the sinonasal tract [4], nasal cavity [11, 12], nasopharynx [14, 6], oropharynx [7], hypopharynx [8, 9], cervical esophagus [10], ear [11], and eustachian tube [2]. Hamartomas composed of vascular tissue, or hemangiomas, are most common in the head and neck. Mature cartilage outgrowths have been described in the nasal cavity, larynx, trachea, bronchi, and lungs [12].

The term “hamartoma” was first used by Albrecht in 1904 (7). Hamartomas are defined as tumor-like, but primary non-neoplastic malformations or inborn errors of tissue development, characterized by an abnormal mixture of tissues indigenous to a part of the body with an excess of one or more of the cellular components (8). They do not clearly represent
either neoplastic or inflammatory disorders (4). In contrast to neoplasms, hamartomas have no capacity for continuous unimpeded growth and therefore their proliferation is self-limiting (2). In general, hamartomas have no malignant potential and no tendency to regress spontaneously (9). The mechanisms that induce hamartomas remain unknown (4); however, it has been speculated that they arise as a result of an underlying inflammatory process (2). As a rule, the treatment for hamartoma is complete removal. Recurrence caused by incomplete excision has been reported (10).

References:
11. B.M. Wenig, D.K. Heffner, Respiratory epithelial adenomatoid


References: