A rare case of congenital nasopharyngeal teratoma presenting with respiratory distress

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Abstract Teratomas of the head and neck are rare, benign neoplasms, usually presenting in the neonatal period and are often malignant with regard to their location. Teratomas occur with a frequency of one in 4,000 live births with head and neck teratomas accounting for <5% of the total. Extensive literature search has shown that there are only two cases congenital nasopharyngeal teratoma reported in India, and we report the third such case.

Keywords Congenital · Nasopharyngeal · Teratoma

Introduction

Teratomas are true neoplasms composed of tissues from all three germinal layers. They have an unknown origin and a varied microscopic appearance. Teratomas occur with a frequency of one in 4,000 live births with head and neck teratomas accounting for <5% of the total [1]. Head and neck teratomas are mostly cervical with the nasopharynx being the second most common location accounting for approximately 2% of all pediatric teratomas [2].

We describe a case of congenital nasopharyngeal teratoma with an intraoral component presenting with acute respiratory distress in a neonate.

Case report

A 6-day-old female was admitted with severe respiratory distress and stridor associated with pooling of secretions in the mouth. Oral examination revealed a polypoidal mass bulging between the tongue and palate. A computerized tomography was suggested a diagnosis of a benign oral vascular neoplasm possibly a lymphangioma. Multiplanar MRI of head and pharynx illustrated a 2.8×2.5 cm multiloculated cystic mass with heterogeneous density. The tumor was seen to have an intraoral and parapharyngeal component with infiltration and loss of planes with nasopharyngeal wall; however, no intracranial extension could be seen.

A surgical excision was done to relieve respiratory distress. Peroperatively the mass was soft well circumscribed and seen to extend into the cheek and soft palate. There were no associated orofacial anomalies.

The resected tissue was received in multiple irregular grey white soft pieces altogether measuring $3 \times 2 \times 1.8$ cm. Microscopic examination revealed stratified squamous epithelium lined tissue with foci of superficial ulceration.

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The subepithelium showed multiple types of haphazardly arranged tissues comprising predominantly of neuroglial elements. Also seen were intermixed adipose tissue, ependymal lining, blood vessels, cartilage, choroids plexus, retinal pigment layer and ciliated columnar epithelium (Fig. 1). A microscopic diagnosis of teratoma, nasopharynx was given. To confirm the completeness of excision, a postoperative MRI was done. A postoperative diagnosis of nasopharyngeal teratoma with residual mass in the right masticator and buccal spaces extension was suggested.

Fig. 1 Shows intermixed adipose tissue, ependymal lining, blood vessels, cartilage, choroids plexus, retinal pigment layer and ciliated columnar epithelium (H&E $\times 200$)

The subsequent course was uneventful. The patient was discharged 4 days after treatment with no evidence of labored respiration and oral secretions.

Discussion

Teratomas, being the most common neoplasms in the newborn, occur at a rate of one in 4,000 live births [1]. They are most commonly located in the sacrococcygeal region, followed by the ovaries, testes, anterior mediastinum, retroperitoneum, and finally the head and neck region, which account for <5% [3, 4].

Teratoma is a true neoplasm composed of an assemblage of tissue which are alien to the site in which it arises viz-a-viz a hamartoma in which the tissue types are indigenous to the parent organ involved. These components tend to be arranged in a haphazard manner with asynchronous maturation of the involved tissue types. They grow independently of the host and can destroy adjacent tissue.

Teratomas of the head and neck are interesting because of their obscure origin, bizarre microscopic appearance, unpredictable behavior and often dramatic clinical

presentation. Dr. Arnold derived the classification system for teratoid malformations. The system consists of four groups: dermoid cysts, teratoid cysts, true teratomas and epignathi. In 1940, this classification of oral teratomas was modified by Ewing et al. as [5]:

- Dermoids-consisting of epidermal and mesodermal germ layers, attached to soft/hard palate/pharynx near midline
- Teratomas-consisting of all three germ layers in different degrees of disorganization, showing greater structural complexities, size and early development
- Epignathi-consisting of teratomas with higher degree of organizational and recognizable structures. There is presence of formed organs.

There are several theories regarding the histogenesis of teratoma formation. Ahfeld first proposed the included twin theory in the late 1800s. Schwalbe later suggested that teratomas arise, during embryogenesis, from displaced germ cells during migration to the genital ridge [5]. The most popular theory speculates that teratomas arise from totipotent embryonic tissues that are somehow displaced during ontogeny. For head and neck teratomas, the theory postulates that embryonic tissue in proximity to the primitive streak and notochord somehow loses its governing influence over the totipotent cells, allowing unorganized differentiation. This is presumably due to alterations in cellular membrane chemistry. The significance of immature elements in the surgical specimen is unclear since malignancy and recurrence have not been reported after local excision of nasopharyngeal teratomas. It is hypothesized that the teratoma physically interferes with the normal fusion of embryonic tissues in the early developmental period [5].

The clinical presentation depends on the size and location of the mass. A marked female predominance of six to one has been cited [2]. Clinical manifestations can range from failure to thrive and mild positional dyspnea to severe airway compromise. Nasopharyngeal teratomas may be sessile or pedunculated, and have been reported to protrude from the mouth and nares. Diagnosis is usually made early in infancy and rarely after 2 years of age. Most of the teratomas are diagnosed at birth with asphyxia neonatorum [6, 7]. Large teratomas may lead to still birth on delivery where as smaller tumors present after with less dramatic symptoms such as excessive nasal discharge, excessive snoring, failure to thrive, recurrent cough, fever and dysphagia [2]. Teratomas of the nasopharynx are often associated with cranial structural anomalies. Palatal fissures are common and hemicrania and anencephaly have been reported [2].

The diagnosis can be made with oral palpation, plain radiographs and tomographs. CT scans are better for bony evaluation while MRI is more helpful in assessing the anatomical location, size, relation to adjacent vital structures, resectability and any intracranial extension. Histopathology

is, not as a rule helpful in establishing the diagnosis, however, is the gold standard for confirming the diagnosis. Only rare teratomas of the nasopharyngeal region have been reported to have immature or malignant tissues [2].

A wide spectrum of teratoid lesions arises from the nasopharynx. Encephalocele, cephalic herniation through congenital defects in the skull, is probably the most important preoperative differential diagnosis. Ten percent of encephaloceles are located in the pharynx. Despite CT scanning, teratomas have been misdiagnosed for meningoencephaloceles and intraoperative aspiration has been recommended, with chemical evaluation of the fluid [8]. Differentiation from glial tumors often requires excisional biopsy since nasopharyngeal teratomas can be predominantly composed of neural tissue. Heterotopic nasopharyngeal brain tissue may be further differentiated from the more common encephaloceles and nasal gliomas. Oral lymphangioma/cystic hygromas which are predominantly in the lateral and posterior neck regions are also an important differential diagnosis to be considered in a congenital head and neck teratoma and as seen in the present case, this diagnosis needs to be confirmed with a magnetic resonance scan or histological examination.

Complete extirpation is the ultimate goal of treatment of head and neck teratomas. Surgical resection may however be laborious and incomplete because of the complex structure and extension into multiple structures.

Conclusion

In conclusion the authors describe the third case of congenital nasopharyngeal teratoma in India. Teratomas of the head

and neck are rare, benign neoplasms, usually presenting in the neonatal period and are often malignant with regard to their location. They can enlarge rapidly; diagnosis needs to be made promptly, usually with the aid of tomographic or magnetic resonance scans, followed by confirmation with excisional biopsy.

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