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Congenital nasopharyngeal teratoma (epignathus) with airway obstruction

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Introduction: Teratomas of the nasopharynx are rare in neonates, usually presenting in the neonatal period with frequency of the total 1 in 4000 live birth and derived from the upper jaw, palate and sphenoid bone. It usually protrudes through the mouth, leading to obstruction of the upper airway .Teratoma of the head and neck are mostly benign neoplasm. In this article we are reporting a case of Epignathus associated with airway obstruction.
Case report: A 40 day old male neonate was admitted with severe respiratory distress and nasopharyngeal mass protrude to the oral cavity prevented oral feeding. History of mild hydroaminious was positive. No other abnormalities were noted on physical examination. MRI of the head and neck revealed solid cystic mass with nasopharyngeal origin without intracranial extension. Mass was resected surgically and pathology revealed a mature teratoma dominantly composed of mature neuroglial elements and choroid plexus.
Conclusion: Nasopharyngeal teratoma cases are interesting because of their obscure origin and unusual clinical presentation.surgical resection is the treatment of choice and should be done urgently in the case of airway obstruction.

Keywords: Epignathus,Teratoma, Nasopharyngeal tumor

Background

Teratoma is a true neoplasm that consists of tissues from all three embryonic germ layers; ectoderm, mesoderm and endoderm (1). It is the most common extragonadal germ cell tumors of childhood (2,3). The incidence of teratoma is 1:4000 births, with the sacrococcygeal area being the most common site (2, 4). Teratoma of the head and neck account for less than 2% of reported cases of congenital teratoma and are most commonly found in the cervical neck (2, 4, 5). Teratoma originating from the oral cavity are named as epignathus (6).Epignathus is a rare cause of upper airway obstruction of the newborn that can grow rapidly in the neonatal period (7). While adult teratoma tend to be histologically and oncologically malignant ,pediatric teratomas of the head and neck tend to be benign (8, 9). We present a case of nasopharyngeal teratoma associated with airway obstruction.

Case presentation

Our patient was a 40 day old male neonate which was referred to Imam Komeini Hspital because of severe respiratory distress and nasopharyngeal mass. The mass protruded in the oral cavity and prevented oral feeding. The intrapartum ultrasound examination revealed mild hydramnious without any malformation. No other abnormalities were noted on physical examination. Head and neck MRI demonstrated solid and cystic mass with nasopharyngeal origin, without intracranial extention (figure 1). The mass was completely excised surgically. Surgical repairing was planned for later. Histological examination revealed mature teratoma dominantly composed of mature neuroglial elements and choroid plexus. No immature component was present and on the basis of microscopic examination, the diagnosis is mature congenital nasopharyngeal teratoma (Figure 2).

Discussion

Teratoma refers to a neoplasm that recapitulates all three germ layers (8, 10). It is the most common extragonadal germ cell tumors of childhood (2, 3).The incidence of teratoma is 1:4000 births, with the sacrococcygeal area being the most common site (2, 4). Teratoma of the head and neck are extremely rare and usually seen during the neonatal period (8, 9). There are less than 300 reported cases of neonatal head and neck teratomas in the literatures (2, 11). Nasopharyngeal teratoma arises from the skull base or the posterior pharyngeal wall and extend inferiorly to cause obstruction of the upper aerodigestive tract .Although some of these tumors are small, others can be large enough to protrude from the oral cavity (2, 12).Symptoms are dependent on the size and location of the lesion and include respiratory distress, dysphasia, failure to thrive, difficulty in feeding or even intermittent symptoms of cough and cyanosis (2, 4). Our case is presented with respiratory distress and difficulty in feeding.

According to some reports sometimes there are associated abnormalities with nasopharyngeal

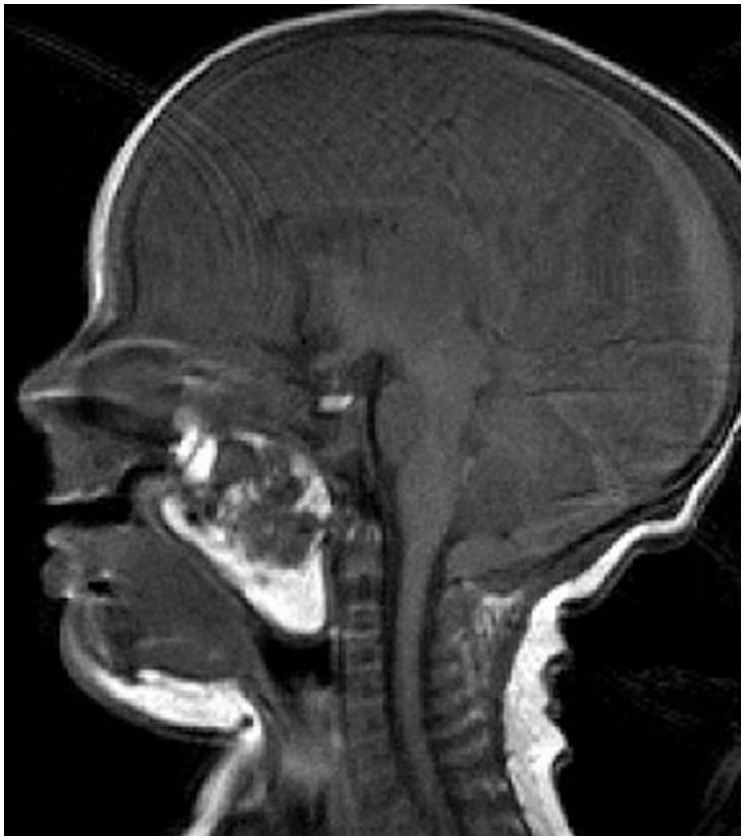

Figure 1. MRI of patient

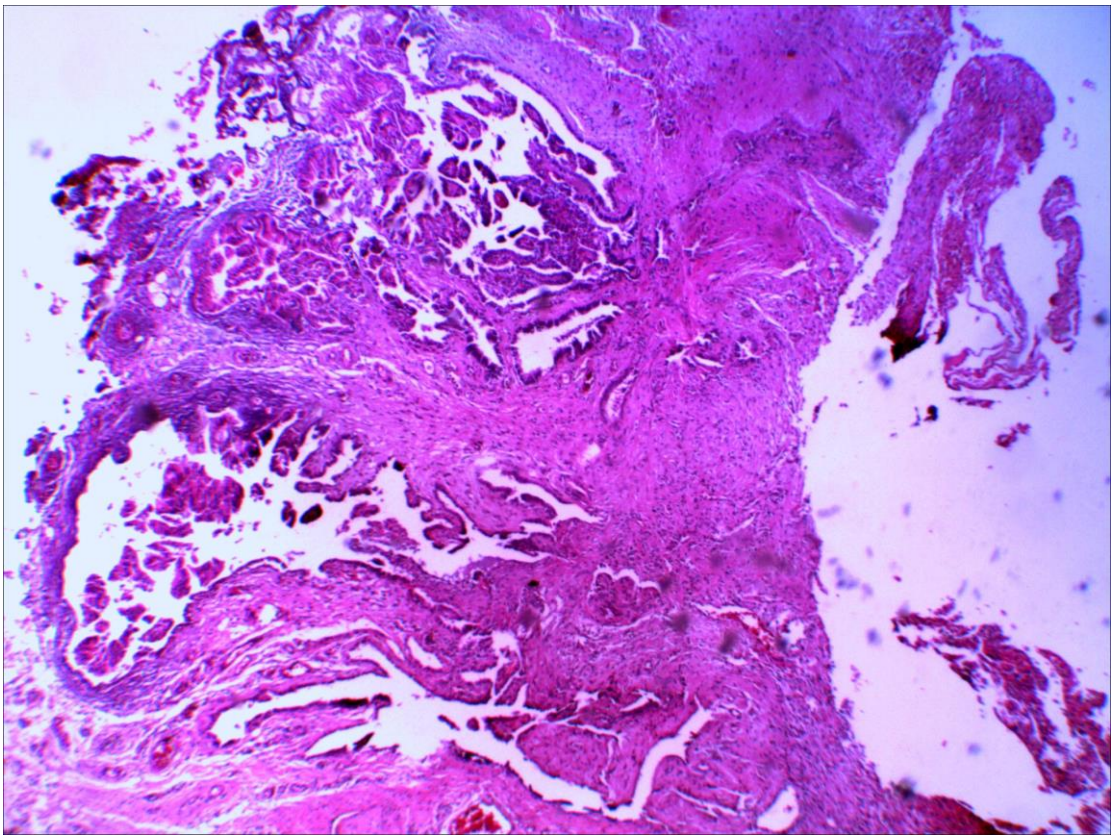

Figure 2. H & E Pathology of specimen

teratoma (2), but in our case no evidence of associated congenital malformations were detected.

The major cause of morbidity and mortality is from the size and location of the teratoma, causing airway obstruction and respiratory distress. Respiratory distress was the most common presenting symptoms (2, 4).

The differential diagnosis of nasopharyngeal masses in the infant include intranasal glioma, meningoencephalocele, encephalocele, congenital rhabdomyosarcoma, hemangioma, neurofibromatosis and lymphatic malformation (2, 4).

CT scan and MRI play a key role in differentiating neonatal nasophryngeal teratoma from other cause of neonatal neck mass (13, 14).

In our case microscopic examination revealed benign teratoma mainly composed of mature neuroglial component and choroid plexus with no evidence of immaturity.

The main therapy of teratoma is complete excision which depends on the site of tumor. The prognosis is excellent and recurrences are rare (13).

Conclusion

Congenital nasopharyngeal teratoma (epignathus) is a rare neoplasm and usually benign. Because of the most common symptom which is respiratory distress and airway obstruction urgent diagnosis and treatment with excisional surgery is required.

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