Case Report

Iran J Pediatr
Jun 2011; Vol 21 (No 2), Pp: 249-252

Congenital Nasopharyngeal Teratoma in a Neonate

Alireza Mirshemirani*, MD; Ahmad Khaleghnejad, MD; Leila Mohajerzadeh, MD; Majid Samsami, MD, and Shaghayegh Hasas-yeganeh, MD

Pediatric Surgery Research Center, Mofid Children's Hospital, Shaheed Beheshti University of Medical Sciences, Tehran, Iran

Received: *Apr 08, 2010*; Final Revision: *Jun 05, 2010*; Accepted: *Jun 30, 2010*

Abstract

Background: Congenital germ cell tumors are uncommon. The most common site of teratoma is in the sacrococcygeal region. Teratoma arising from the head and neck comprises less than 10% of reported cases and of these, nasopharyngeal lesions are rare. Teratomas are generally benign, and have a well recognized clinical and histopathological entity. We present a case of nasopharyngeal teratoma (NPT) associated with a wide cleft palate.

Case Presentation: A 20 day old female neonate with a teratoma of the nasopharyngeal area, and wide cleft palate was referred to our center. The protruded mass which measured 6×4×3cm, was of soft consistency, blocked the airway, and prevented oral feeding. Preoperative evaluation and imaging was performed and mass was excised 2 days after admission. Pathology revealed a well-differentiated mature solid teratoma (hairy polyp). The patient had no complication in the post-operative period. Cleft palate was surgically repaired when 2 years old. She is now a six year old girl with normal development.

Conclusion: Congenital nasopharyngeal teratomas are usually benign. Surgery is the treatment of choice, and should be undertaken on an urgent basis, especially in a patient who presents with signs and symptoms of airway obstruction.

Iranian Journal of Pediatrics, Volume 21 (Number 2), June 2011, Pages: 249-252

Key Words: Teratoma; Nasopharyngeal Tumor; Cleft Palate; Neonate; Airway Obstruction

Introduction

Teratomas of the head and neck are rare congenital lesions comprising less than 10% of reported cases, and nasopharyngeal teratoma (NPT) is even rarer[1]. Teratomas are benign tumors containing cells from ectodermal, mesodermal and endodermal layers [2], and they involve at least two of the above layers[3].

*** Corresponding Author;**
Address: Pediatric Surgery Research Center, Mofid children's Hospital, Shariati Aven. Tehran, Iran.
E-mail: almirshemirani@gmail.com

Although they are rare neoplasms composed of tissue elements derived from the germinal layers of the embryo, they may originate anywhere along the midline, and clinical behavior varies significantly by site and size[4]. Although NPTs are histologically benign, but can cause considerable morbidity and mortality because of their location[5]. Nasopharyngeal teratoma represents one of the most unusual causes of respiratory distress during the neonatal period[6]. The imaging of choice in head and neck lesions is based on a number of factors, several of which are unique to the pediatric population.

Although the bulk of disease entities are adequately evaluated by CT-scan, Magnetic Resonance Imaging (MRI) can provide additional vital information in many cases. MRI provides better soft tissue characterization than CT scan, and has multiplanar capabilities[7].

In this article we describe a case of congenital mature teratoma of nasopharynx in a 20-day old neonate.

Case Presentation

Our patient was a 20-day old neonate, the product of a full term pregnancy from a 30 year-old mother G3P2, via a normal spontaneous vaginal delivery with the birth weight of 2900 grams. The pregnancy and delivery were uncomplicated and the intrapartum ultrasound examinations were normal. The patient was referred to our out-patient clinic due to a large pedunculated mass (Fig 1) with gradual enlargement which blocked the airway and prevented oral feeding.

Examination revealed defective palate with a large 6×4×3 cm mass protruding through the defect. Head and neck CT-scan demonstrated a soft tissue tumor in nasopharynx with a focus of calcification of a tooth; brain CT scan was noted normal. Alpha-fetoprotein (AFP) was 1403mg/ml. the mass was completely excised surgically two days after admission. The patient had a wide cleft palate after mass resection (Fig 2), surgical repair of which was planned for later. Postoperative course was uneventful with improvement of respiratory and feeding problems.

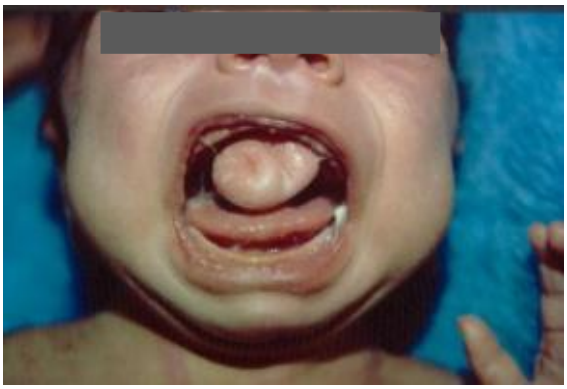
Pathologic Findings:

A firm mass with elastic consistency measuring 6×4×3 cm, with smooth external surface containing hair and a 2×1.5×0.8 cm nodule containing tooth and mucinous cyst.

Microscopic examination showed a disorganized combination of mature adipose tissue, mucin secreting glands, tooth structure, skin adnexes, neural tissue, skeletal muscle and bone. No immature component was present, and the diagnosis was congenital mature solid teratoma (hairy polyp) of nasopharynx, well-differentiated type.

Follow-up:

She was readmitted in our hospital at the age of 2 years (Fig 3). The AFP level was normal and palate defect was reduced in size; CT scan revealed no evidence of residual or recurrent tumor (Fig 4). Cleft palate was repaired surgically when 2 years old, and patient was discharged uneventful. She is now a 6-year-old girl and is under observation.



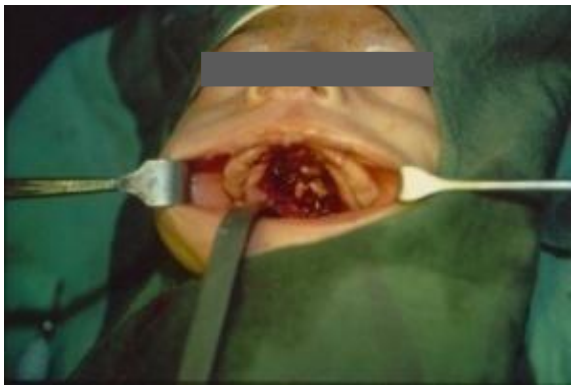


Fig. 1: The patient with a large pedunculated mass

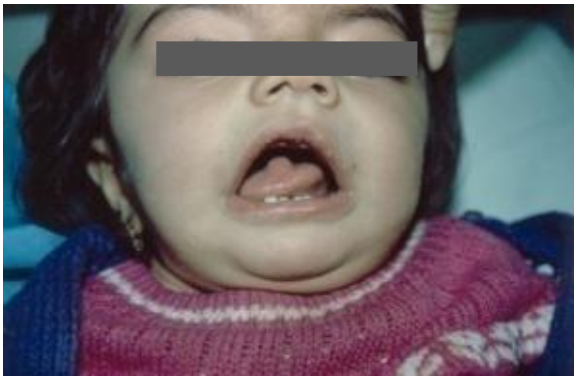


Fig. 2: The patient had a wide cleft palate after mass resection

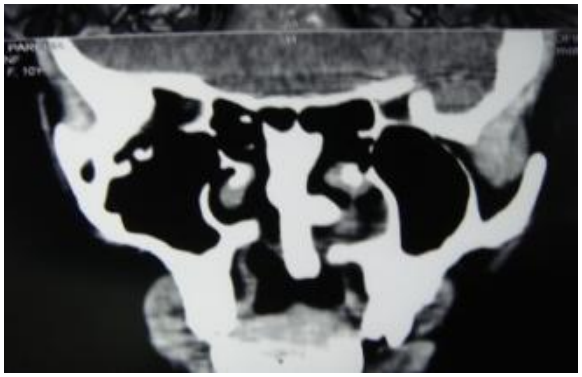


Fig. 3: The patient at the age of 2 years

Fig. 4: CT scan revealed no evidence of residual or recurrent tumor at the age of 6 years

Discussion

Teratomas occur in 1 out of 4000 live births. Head and neck occurrence is generally localized in the neck and nasopharynx and comprises 1-10% of the cases[8]. Teratomas are congenital tumors that are composed of tissues derived from all three embryonic germ cell layers. Four basic histological classifications are generally recognized: 1) dermoid cysts (epithelial lined with skin elements, composed of ectodermal and mesodermal cells), 2) teratoid cysts (all 3 germ layers but poorly differentiated), 3) true teratoma (3 germ layers differentiated into specific tissues or organs), and 4) epignathi (oral tumors with develop-mental fetal organs and limbs, which is very rare, with a high mortality rate)[9].

Another pathologic variation of epignathi is fetus-in-fetus, which may be considered to be incomplete twinning of monozygotic twins at a primitive stage when axial development begins[10]. Teratomas are true neoplasms originating from pluripotent cells and are composed of tissues from all three germinal layers[5]. Hairy polyp (dermoid) is a teratoid lesion[11], which we had in our patient. Hairy polyp of the pharynx may be associated with an ipsilateral branchial sinus[12]. Teratomas are more common in female gender[13]. Teratomas are usually benign, when they present in early childhood[14] as in our case. In nasopharyngeal teratoma the most common symptoms are upper airway obstruction, dysphagia and failure to gain weight[15]. Our case was referred because of respiratory and feeding problems.

Polyhydramnios and the severity of respiratory distress correlate with the size of the teratoma. Large lesions cause hyperextension of the neck of the fetus and lead to esophageal obstruction, swallowing disturbance and polyhydramnios[6].

Clinical differential diagnoses of neonatal oral mass include: embryonic congenital rhabdo-myosarcoma, retinoblastoma, nasal glioma, heterotopic thyroid, cystic lymphangioma of the oro or nasopharyngeal regions, and sphenoid meningoencephalocele[10]. CT scan and MRI play a key role in differentiating neonatal naso-pharyngeal teratomas from other causes of neonatal neck mass[16]. These modalities were also used in our patient.

The main therapy of teratoma is complete surgical excision which depends on the site of the tumor. The prognosis is excellent, recurrences are rare, and in our understanding, it occurs due to incomplete surgical resection.

Conclusion

Congenital nasopharyngeal teratomas are usually benign. Surgery is the treatment of choice, and should be undertaken on an urgent basis, especially in a patient who presents with signs and symptoms of airway obstruction.

Acknowledgment

We thank Miss Fatemeh Jalali for her kind assistance in preparing the manuscript.

References

1. Makki FM, Al-Mazrou KA. Nasopharyngeal teratoma associated with cleft palate in a newborn. *Eur Arch Oto-Rhino-Laryngol* 2008; 265(11):1413-5.
2. Becker S, Schon R, Gutwald R, et al. A congenital teratoma with a cleft palate: report of a case. *Br J Oral Maxillofacial Surg* 2007;45(4):326-7.
3. Tonguc I, Sahin AM, Ayla G, et al. A congenital true teratoma with cleft lip, palate, and columellar sinus. *J Craniofacial Surg* 2007;18(5): 1083-5.
4. Barksdale EM Jr, Obokhare I. Teratomas in infants and children. *Current Opinion Pediatr* 2009;21(3):344-349.
5. Khoddami M, Mirchi A, Mirshemirani A. Congenital pharyngeal teratoma associated with malposed palatine teeth (a case report), *Iran J Pathol 2006;1(2):87-90.*
6. Hasiotou M, Vakaki M, Pitsoulakis G, et al. Congenital cervical teratoma. *Intern J Pediatr Otorhinolaryngol* 2004;68(9):1133-9.
7. Sachin G, Dheeraj G, Suresh M. Pediatric head and neck masses. *Topics Mag Reson Imag* 2004;15(2): 95-101.
8. Cay A, Bektas D, Imamoglu M, et al. Oral teratoma: a case report and literature review. P*ediatr Surge Int* 2004;20(4):304-8.
9. Jarrahy R, Cha St, Mathiasen RA, et al. Congenital teratoma of the oropharyngeal cavity with intracranial extension: case report and literature review. *J Craniofac Surg* 2000;11(2):106-12.
10. Hej DDS, Wang Y, Zhu H DDS, et al. Nasopharyngeal teratoma associated with cleft palate in newborn: report of 2 cases. *Oral Maxillofacial Surg* 2010;109(2), 211-6.
11. Brown-Kelly A. Hairy or dermoid polyp of the pharynx and nasopharynx. *J Laryngol Otol* 1918; 33:65-70.
12. Burns BV, Axon R, Pahade A. Hairy Polyp of the pharynx in association with an ipsilateral bronchial sinus. *J Laryngol Otol* 2001;115(2): 145-8.
13. Ulger Z, Egemen A,Karapinar B, et al. A very rare cause of recurrent apnea: Congenital naso-pharyngeal teratoma. *Turk J Pediatr* 2005;47(3): 266-9.
14. Anderson PJ, David DJ. Teratomas of the head and neck region. *J Craniomaxillofac Surg* 2003;31(6): 369-77.
15. Rotenberg B, Nasopharyngeal teratocarcino-sarcoma. *Int J Pediatr Otorhinolaryngol* 2009; 62(2):159-64.
16. Andronikou S, Kumbla S, Michelle Fink A. Neonatal nasopharyngeal teratoma: Cross sectional imaging features. *Pediatr Radiol* 2003; 33(4):241-6.