

Nasopharyngeal teratoma associated with cleft palate in a newborn

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Abstract Teratomas of the head and neck are rare congenital lesions comprising less than 10% of reported cases. Nasopharyngeal teratomas (NPT) are even rare. A case presented of a newborn with NPT associated with cleft palate. The most common presenting symptom of NPT is respiratory distress. The management of choice for NPT is surgical excision. Overall, the prognosis for NPT is excellent. Recurrences are rare, and are felt to be due to incomplete surgical resection.

Keywords Nasopharynx · Newborn · Congenital · Teratoma

Introduction

Teratoma defined as a true neoplasm composed of multiple tissues foreign to the part in which it arises. Neonatal head and neck teratomas are extremely rare with the sacrococcygeal region being the most common site. An incidence of 1 in 20,000 to 1 in 80,000 live births has been reported and these tumors account for 1.6–9.3% of all neonatal teratomas [1]. Nasopharyngeal teratoma (NPT) are, however, much rarer and account for only 2–3% of all teratomas [2].

Teratomas include tissue from all three germ layers. Grossly, they are heterogenous masses composed of both

solid and cystic components. The vast majority of head and neck teratomas are benign, especially in the pediatric population.

Nasopharyngeal teratomas are rare congenital tumors. They most commonly arise from the midline or lateral nasopharyngeal wall and display a female predominance of six to one. Histologically, they are classified into four types: dermoids, teratoids, true teratomas, and epignathi [3]. True teratomas are congenital neoplasms of tridermal origin. They are larger and have more complete structure than teratoids and dermoids and are differentiated to the degree that organs can be recognized by histologic structure. The exact origin of teratomas is unknown and is a matter of dispute [3].

This paper reports a unique case of NPT associated with cleft palate that was excised using the micro-debrider. Our aim is to draw the attention of physician for such an association to provide an early diagnosis and proper management.

Case report

The following report describes a full term 39-week-female born to a Gravida 3 Para 1 + 1 woman who had a history of having received antibiotics for urinary tract infection. The neonate was delivered by spontaneous vaginal delivery with birth weight of 3 kg; her apgar score were 8 and 9 at 1 and 5 min, respectively. Her respiratory rate was 51 breath/min. Prenatal ultrasound and follow up records are not available at our hospital since it took place at a different hospital.

The newborn was noted to have mild respiratory distress with evidence of grunting and subcostal retraction. She was taken to the neonatal intensive care unit (NICU) where she was attached to a nasal canula.

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Clinical examination revealed: (1) Small, soft right sided cleft palate; (2) Micrognathia; (3) Hard mass seen behind the soft palate most of it in the right side, pink in color and firm in consistency. The neonate had been investigated for the nasopharyngeal mass by fiberoptic nasolaryngoscope, which showed and confirmed obstruction of the nasopharyngeal space by a mass. A head computed tomography (CT) scan was performed which showed a large $5 \times 4 \times 3.5$ cm heterogenous mass with solid and cystic components arising from the right nasopharynx and extending to the oropharynx. Areas of calcification were also noted (Fig. 1). A magnetic resonance imaging (MRI) confirmed the CT scan findings and showed no evidence of encephalocele. Genetic screening including fluorescent in situ hybridization (FISH) and hearing assessment with auditory brainstem response test (ABR) were within normal.

The neonate was taken to the operating room for examination under anesthesia of the postnasal space and excision of the nasopharyngeal mass. Examination showed a large nasopharyngeal mass filling the nasopharynx and coming down through a cleft of the soft palate to the oral cavity (Fig. 2). The mass was attached to the eustachian tube and to the soft palate from the nasal side. The mass was excised completely using a micro-debrider after taking multiple biopsies for histopathology. Results revealed the presence of a mature NPT with no evidence of malignant changes. Postoperatively the baby did well, tolerating oral feeds without vomiting or regurgitation and was discharged 1 week after surgery. The patient was then followed up

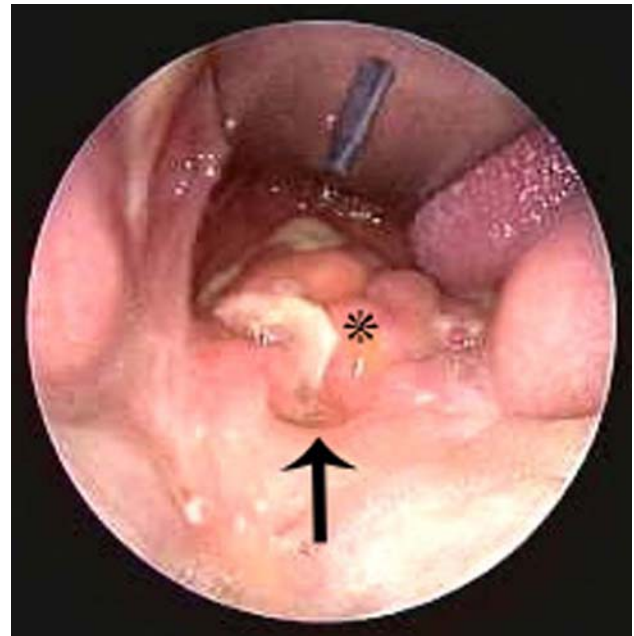


Fig. 2 Nasopharyngeal teratoma extending to oropharynx (*asterisk*) and splaying the soft palate (*arrow*)

with the plastic surgery and underwent cleft palate repair at the age of 6 months.

Discussion

Nasopharyngeal teratomas are principally solid masses composed of tissues derived from embryonic ectoderm, mesoderm, and endoderm. They are usually limited to the region of the nasopharynx and oropharynx, but extensive neck involvement has rarely been reported [4].

Embryologically, the oral cavity begins as a depression in the stomodeum. The ectoderm of the stomodeum and endoderm of the foregut are in direct contact forming the buccopharyngeal membrane. Gradually, the stomodeum and foregut merge. An out pouching of the ectoderm grows rostrally as the Rathke pouch. Occasionally, remnants of endoderm and mesoderm may migrate with the ectoderm. These germ layers can become trapped in the nasopharynx and subsequently undergo differentiation, creating a teratoma [5]. Nasopharyngeal teratomas frequently prevent fusion of the palatal processes, resulting in clefting. In our case, a cleft soft palate was detected.

Teratomas in neonates and infants contain tridermic tissue elements: ectoderm, endoderm, and mesoderm. Although the preponderant tissue is most often neuroepithelium, other tissues that can be involved include fetal cartilage, myogenic tissue, and epithelium (squamous, ciliated, columnar, enteric, and endocrine) [6].



Fig. 1 Axial CT scan at the level of nasopharynx showing a large, mixed attenuation mass (*asterisk*) with areas of calcifications (*arrow*)

The differential diagnosis of a nasopharyngeal mass in the infant includes intranasal glioma, meningoencephalocele, encephalocele, congenital rhabdomyosarcoma, hemangioma, neurofibromatosis, and lymphatic malformation. The most important alternative diagnosis is encephalocele or meningoencephalocele. Infants with nasopharyngeal teratoma may be full term, premature, or stillborn.

The most common presenting symptom of nasopharyngeal teratomas is respiratory distress. CT and MRI findings that help confirm the diagnosis include the presence of a heterogeneous cystic/solid mass, often with calcific components, with evidence of chronic mass effect on adjacent structures, especially distortion of the ipsilateral hemimandible and pterygoid plates [7].

There have been several reports of associated abnormalities occurring with NPT. These include incomplete or complete palatal clefts, cardiac abnormalities, microcephaly, and atresia of the left common carotid [8]. In our case the patient presented with a big nasopharyngeal mass filling the nasopharynx and coming down through a cleft soft palate to the oral cavity, and this was associated with micrognathia.

Ideally, the diagnosis of a head and neck teratoma is made in utero so that airway management strategies can be instigated before birth. The fetus sometimes is unable to swallow amniotic fluid owing to mass effect of the teratoma. As a result, approximately 18% of mothers have polyhydramnios on fetal ultrasound [9]. An increased alpha-fetoprotein (AFP) level in amniotic fluid has been reported in association with NPT [2]. Prenatal ultrasound (US) is useful in identifying head and neck teratomas. Since 1977, prenatal US have been used to diagnose head and neck teratomas [10]. If a prenatal US does show a head and neck mass, the likelihood of prenatal airway obstruction is high. This finding should alert the physician in charge to the potential risk and appropriate prenatal planning should occur.

The management of choice for NPT is surgical excision. When a neonate is experiencing respiratory difficulty, the first priority should be stabilization of the airway. Overall, the prognosis for NPT is excellent. Only one reported death as a direct result of a NPT has been reported in the past 20 years [11]. Recurrences are rare, and are felt to be due to incomplete surgical resection and most likely this reflects the increased difficulty of complete surgical resection of these lesions in the neonate.

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