A very rare cause of recurrent apnea: congenital nasopharyngeal teratoma

Zülal Ülger[1], Ayten Egemen[1], Bülent Karapınar[1], Ali Veral[2], Fazıl Apaydın[3]

Departments of [1]Pediatrics, [2]Pathology, and [3]Otorhinolaryngology, Ege University Faculty of Medicine, İzmir, Turkey

SUMMARY: Ülger Z, Egemen A, Karapınar B, Veral A, Apaydın F. A very rare cause of recurrent apnea: congenital nasopharyngeal teratoma. Turk J Pediatr 2005; 47: 266-269.

A three-month-old girl patient born at the 37th week of gestation as one of twins was admitted to our hospital with cardiac arrest. There was past medical history of multiple hospitalizations with symptoms of cyanosis, respiratory distress, and frequent and severe attacks of apnea since her birth. On nasopharyngeal computerized tomography a mass (25x24 mm) occupying the right side of the nasopharynx was detected. The pathological evaluation of the excised mass revealed benign teratoma. After the removal of the nasopharyngeal mass under general anesthesia, respiratory distress and attacks of apnea disappeared completely. In every neonate with unexplained stridor and recurrent apnea, beside the common causes like respiratory distress syndrome, sepsis, and asphyxia, the possibility of oropharyngeal and nasopharyngeal mass should be kept in mind.

Key words: apnea, respiratory distress, stridor, nasopharyngeal teratoma.

Teratoma is a quite rare congenital neoplasm, especially the one originating from the nasopharynx. Teratomas arise from pluripotent stem cells and include elements from all three embryonic germ layers. Most are benign, but in a critically placed lesion, because of local mass effect, they demonstrate high mortality and morbidity. Tumor localization and size rather than histological grading are the most significant features affecting the immediate clinical course. In that sense, congenital teratoma of the nasopharynx and oropharynx is a unique clinical entity that possesses immediate threats to the neonate. Establishment of secure airway is the primary goal and then complete surgical resection is the subsequent principle of management.

In this case report, a patient admitted with severe respiratory distress and frequent attacks of apnea requiring mechanical ventilatory support and finally diagnosed as nasopharyngeal teratoma is presented. In any child with unexplained stridor and attacks of apnea, the pediatrician should consider this possibility in the differential diagnosis.

Case Report

A three-month-old girl patient born at the 37th week of gestation as one of twins with a birth weight of 2250 g was admitted to our hospital with cardiac arrest. She had been discharged from another hospital two days before. In her past medical history there were symptoms of cyanosis, respiratory distress, loud breathing and frequent attacks of apnea since the third day of birth. She was hospitalized four times in different hospitals with these symptoms and diagnosed as laryngomalacia and convulsive apnea and given phenobarbital treatment, with no beneficial effects.

The parents were nonconsanguineous and her twin sister was healthy.

On physical examination after cardiopulmonary resuscitation, weight was 2.5 kg (3-10p), length was 50 cm (50p), heart rate: 130/min, and respiratory rate: 55/min. Severe stridor, and intercostal and subcostal retractions were present. Cardiac and respiratory auscultations and neurological examinations were normal. Micrognathia, retrognathia, high-arched palate, and mild exophthalmos with frightened facial appearance were also observed.

On laboratory examination, hemoglobin was 12.2 g/dl, Hct: 30%, white blood cell count: 8000/mm^3; blood ions, and liver and renal function tests were normal; blood and urine cultures were negative. Serum IgA level was < 5.78 mg/dl, IgG: 229 mg/dl and IgG 3:

0.07 mg/dl, and all these immunological values were less than normal according to sex and age. However, lymphocyte panel was normal. These immunoglobulin levels will be repeated at six months of age to differentiate transient hypoglobulinemia of infancy and IgG subgroup-IgA deficiency. Karyogram was normal.

On follow-up, frequent attacks of apnea were observed and she was intubated and ventilated with mechanical ventilator due to hypoxia and respiratory acidosis. She was fed with nasogastric tube and extubated four days later, but unfortunately attacks of apnea repeated. To exclude central apnea, cranial magnetic resonance imaging (MRI) and electroencephalography (EEG) were done, but all these were normal. Central apnea was excluded and phenobarbital treatment was stopped. Tracheoesophageal fistula and laryngomalacia were absent on rigid bronchoscope and barium graph of esophagus.

Gastroesophageal reflux was detected on gastrointestinal scintigraphy and even though treatment of ranitidine, domperidone and anti-reflux formula had been started, respiratory distress and apnea did not regress.

Due to signs of upper airway obstruction like inspiratory stridor and loud snoring, consultation was sought from the Ear, Nose and Throat Division. But repeated fiberoptic nasopharygoscopic examinations could not reveal the underlying pathology. Thus, nasopharyngeal computerized tomography (CT) was done, which revealed a mass (25x24 mm) with necrotic tissue occupying the right side of the nasopharynx and oropharynx (Fig. 1). The patient was operated under general anesthesia. The upper part of the tumor was dissected from the nasopharynx using the 2.7 mm 30° telescope and small endoscopic instruments through the nose. The patient was then placed into the Trendelenburg position. The mouth was opened using Davis-Boyle retractor. When the soft palate was retracted, the remainder of the tumor could be removed totally through the mouth. Histopathological examination revealed a benign teratoma (Figs. 2 and 3). After extraction of this mass, the respiratory distress, stridor

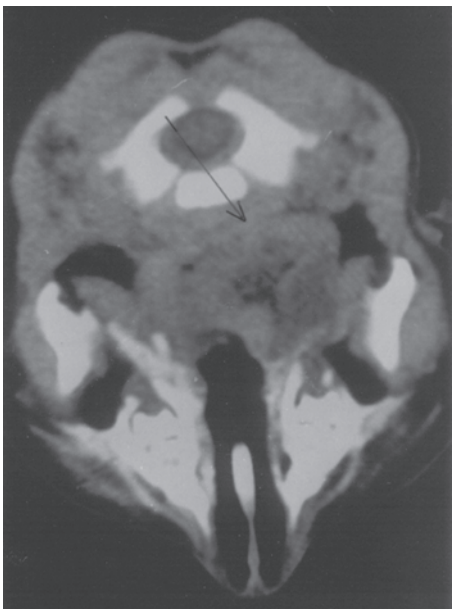

Fig. 1. Nasopharyngeal computerized tomography demonstrating a mass (25x24 mm) with necrotic tissue occupying the right side of the nasopharynx and oropharynx.

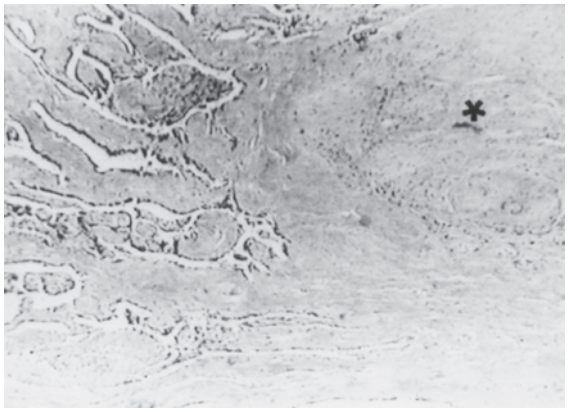

Fig. 2. Histopathological evaluation of extracted mass demonstrating epithelial component forming clefts and papillary structures and neuroglial components (*) (H&E X 40).

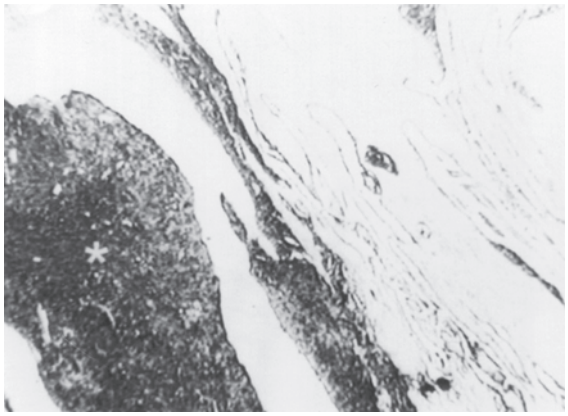

Fig. 3. Glial fibrillary acidic protein (GFAP) positivity in neuroglial tissue (*) (Anti-GFAP X 40).

and attacks of apnea abruptly regressed. The frightened facial expression of the patient was totally changed; retrognathia and exophthalmos were reduced. The postoperative follow-up was done by fiberoptic nasopharygoscopy and direct palpation of the nasopharynx including the operative field. The patient's development was normal during the two-year follow-up period.

Discussion

Apnea is the absence of respiratory gas flow for a period of 20 seconds, with or without a decrease in heart rate. Apnea is due to many primary diseases that affect neonates: central nervous system disorders (intraventricular hemorrhage, seizures, hypoxic injury), respiratory diseases (hyaline membrane disease, pneumonia, obstructive airway lesions, pneumothorax), infectious diseases (sepsis, meningitis), gastrointestinal disorders (gastroesophageal reflux, intestinal perforation), metabolic disorders (hypoglycemia, hypocalcemia, hyperammonemia, hypothermia), cardiovascular diseases (heart failure, hypotension, hypertension, vagal tone) and immaturity of the respiratory center. Apnea may be obstructive, central or mixed. Approximately 50-60% of preterm infants have evidence of apnea. The most common pattern of apnea among preterm neonates has a mixed etiology.

In this case report, we presented a preterm infant admitted with apnea and severe respiratory distress. An obstructive airway lesion, nasopharyngeal teratoma, was found to be the cause of the recurrent apnea.

In our case, the detection of the tumor could not be done during flexible nasopharygoscopy since the dynamic movements of the lateral pharyngeal bands and the adenoid tissue itself were disguising the pathology. After the nasopharyngeal CT evaluation, the mass was detected by careful inspection and could be extirpated under general anesthesia. No recurrence was seen during the two-year follow-up.

Neoplasms of the nasopharynx are rare in children, but they are life-threatening when they do occur[1,2]. The nasopharynx tends to harbor dysontogenetic neoplasm. After classification into benign and malignant groups, nasopharyngeal neoplasm in children can be further characterized according to the age of the patients at which clinical manifestations usually appear[3]. Dermoids and teratomas are the most frequently encountered neoplasms of the nasopharynx in infants and may produce airway obstruction and dysphagia. Among the benign tumors of the nasopharynx in children, the juvenile angiofibroma deserves the most attention.

Teratomas are very rarely seen congenital neoplasms, especially the one originating from the nasopharynx (1/40,000)[4]. They are composed of three germinal layers of the embryo that form tissues foreign to the part. The most common site of origin is in the sacrococcygeal region. According to most studies, teratomas of the head and neck account for only 2-9% of all cases[5].

The teratoma is a well recognized and generally benign clinical and histopathological entity. Four histological types of nasopharyngeal teratoma exist: dermoid, teratoid, true teratoma, and epignathi. Of these, dermoid teratomas comprise the vast majority; true teratomas occur considerably less frequently. The etiology of these lesions is unknown. They probably arise from uncontrolled growth of a pluripotent cell originating in the region of the embryonic notochord.

Nasopharyngeal teratoma is a very unique condition that possesses immediate threats to the neonate. Description of these lesions in medical literature is uncommon. In the majority of these reports, the tumors are associated with stillbirth, perinatal death or significant morbidity after attempted resection. They usually present at or soon after birth with signs of upper aerodigestive tract obstruction. Although prenatally diagnosed cases are present, the reports of nasopharyngeal teratoma diagnosed as late as 10 years of age are also present. Coppit et al.[6] reported that prenatal diagnosis has made little impact on the overall diagnosis and treatment of these lesions.

The patients with nasopharyngeal teratomas have a higher incidence of maternal polyhydramnios, preterm birth, need for emergent airway management, and associated congenital abnormalities[7]. Some of the teratomas extend into the intracranial cavity during development. There was no intracranial extension of the teratoma in this case report.

Congenital teratomas of the neck and nasopharynx are unusual tumors that seldom metastasize. Due to their rarity, reviews often have relied upon complications of isolated case reports. Byard et

al.[8] reported their experience of the early outcome of 18 cases (14 cervical, 4 nasopharyngeal) to demonstrate the high morbidity and mortality that these benign but critically placed lesions have because of local mass effects. Thirty-three percent of patients were stillborn or died of disease two days after birth; of the survivors, 22% had significant respiratory obstruction requiring surgery.

Congenital nasopharyngeal masses are very rare and not assigned priority in the differential diagnosis of apnea and upper airway obstruction. Generally in a neonate with apnea, pediatricians usually consider the possibility of respiratory distress syndrome, intrauterine pneumonia, central nervous system abnormalities, sepsis, hypoxemia, hypothermia, anemia, and metabolic disturbances like hypoglycemia and hyponatremia. Although it is rare, the nasopharyngeal mass must be kept in mind in the differential diagnosis of apnea, especially when it exists together with upper airway obstruction symptoms. Otherwise, mortality and morbidity are very high even in benign lesions.

REFERENCES

1. Tharrington CL, Bossen EH. Nasopharyngeal teratomas. Arch Pathol Lab Med 1992; 116: 165-167.
2. Hong MY, Kuo WR, Wu JR, Lin HJ, Lo YS, Joan KH. Congenital nasopharyngeal teratomas: report of a case and literature review. Gaoxiong Yi Xue Ke Xue Za Zhi 1993; 9: 476-480.
3. Chaudhry AP, Lore JM Jr, Fisher JE, Gambrino AG. So called hairy polyps or teratoid tumors of the nasopharynx. Arch Otolaryngol 1978; 104: 517-525.
4. Gunkel AR, Sturm C, Simma B, Thumfart WF. Transoral CO2 laser resection of extensive nasal and oropharyngeal teratoma. Laryngorhinootologie 1996; 75: 239-241.
5. Shetty SC, Gupta S, Cherian M, Chary G, Shariff S. Mature teratoma of the nasal vestibule: a case report. Ear Nose Throat J 2000; 79: 620-623.
6. Coppit GL 3rd, Perkins JA, Manning SC. Nasopharyngeal teratomas and dermoids. A review of literature and case series. Int J Pediatr Otorhinolaryngol 2000; 52: 219-227.
7. Jarrahy R, Cha ST, Mathiasen RA, Shaninian HK. Congenital teratoma of the oropharyngeal cavity with intracranial extension: case report and literature review. J Craniofac Surg 2000; 11: 106-112.
8. Byard RW, Jimenez CL, Carpenter BF, Smith CR. Congenital teratoma of the neck and nasopharynx. A clinical and pathological study of 18 cases. J Paediatr Child Health 1990; 26: 12-16.