

A Congenital True Teratoma With Cleft Lip, Palate, and Columellar Sinus

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Teratomas, the most common extragonadal germ cell tumor of childhood, involve at least two of the ectodermal, mesodermal, and endodermal layers. Of the teratomas seen in the first 2 months of life, 82% are sacrococcygeal. The head and neck region is the second most common location for teratomas in early infancy, accounting for five (14%) of those cases. We describe a female neonate with a teratoma of the nasopharyngeal area, bilateral cleft palate/lip, and columellar sinus pathologies. The mass, which was 8 × 5 × 7 cm and soft in consistency, blocked the airway and prevented oral feeding. On macroscopic examination of the excised mass, there was a notable typical cilia arrangement and lower eyelid appearance. The patient, who was diagnosed with a well-differentiated teratoma after the pathologic examination, did not have any complications in the postoperative period.

Key Words: Teratoma, cleft palate, cleft lip, columellar sinus

Teratomas, the most common extragonadal germ cell tumor of childhood, involve at least two of the ectodermal, mesodermal, and endodermal layers.¹ These tumors, which generally have paraxial and midline locations (sacrococcygeal region, ovaries), occur in approximately one in every 4000 births.²

There are various theories about its etiology: traumatic implantation in the tissue, failure of somatic regions to fuse during embryogenesis, or implantation of pluripotential cells that disorganize growth in the tissue.³⁻⁹ The main differential diagnosis is

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encephalocele or meningoencephalocele, and it is important to know before deciding on the operation.¹⁰ Other differential diagnoses are cystic hygroma, cyst of the branchial cleft or thyroglossal duct (all non-calcified and mainly cystic), rhabdomyosarcoma, and hemangioma (calcified and mainly solid).

In 6% of all cases depending on the localization and size of the teratomas, they may often be associated with other malformations, for example, cleft palate,¹¹ bifid tongues and noses,^{12,13} duplication of the pituitary gland, and so forth.^{14,15} Our patient had a bilateral cleft palate, cleft lip, and columellar sinus.

CLINICAL REPORT

A baby girl was who was born in 2006 as one of triplets was intubated and followed in the pediatric intensive care unit of Kocaeli University as a result of frequent hypoxia attacks. Her physical examination revealed a mass located in the nasopharyngeal area in the mouth. The mass, which was 8 × 5 × 7 cm and soft in consistency, blocked the airway and prevented oral feeding. In addition to the mass, she also had bilateral cleft palate/lip and columellar sinus pathologies. Besides, the cleft palate had an asymmetric appearance as a result of the mass (Fig 1). Her systemic magnetic resonance image, ultrasonography scans, and karyotype analyses did not demonstrate any other pathology. After the necessary anesthesia and pediatrics consultations, the patient was operated on when she was 94 days old. During the operation, which was performed under general anesthesia, the mass was totally excised from its intact boundaries. A lip reparation operation aiming at the cleft lip was conducted, and the operation was ended (Fig 2). In the macroscopic examination of the excised mass, there was a notable typical cilia arrangement and lower eyelid appearance (Fig 3). The patient, who was diagnosed with a well-differentiated teratoma after the pathologic examination, did not have any complications in the early postoperative period (Fig 4). The patient, who no longer needed intubation in



Fig 1 Preoperative image of the case. The columellar sinus is cannulated.

the postoperative period, was discharged and followed for necessary cleft palate and columellar sinus treatments.

DISCUSSION

The classification of teratomas consists of four general types: 1) dermoid, which is also called hair polyps, containing the epidermal and mesodermal elements, is the most common type; 2) teratoid, which consists of ectoderm, mesoderm, and ectodermal elements but poor differentiation (the case reported here mostly fits this classification); 3) true teratoma, which also contains all three germ layers like the teratoid but organizes or differentiates into a recognizable early organ (cartilage, teeth, and so on); and 4)

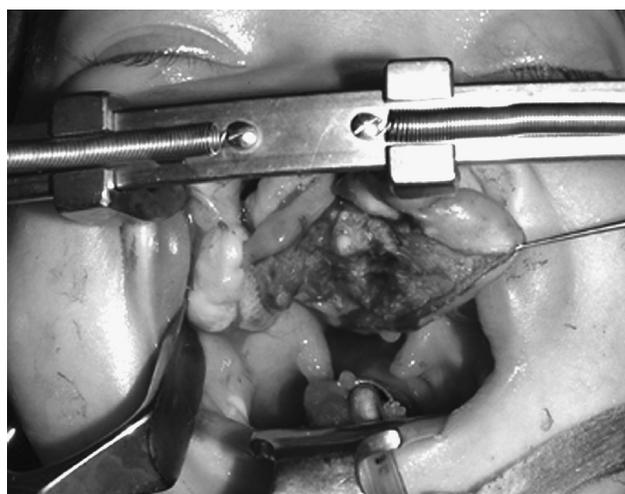


Fig 2 Image of the surgical area during the operation.

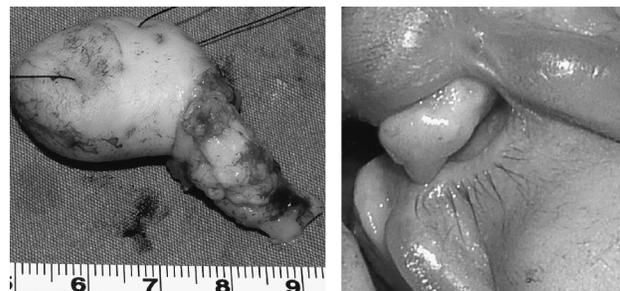


Fig 3 (A) Image of the excised mass. (B) Typical cilia localization and eyelid presentation on the mass.

epignathus, which is highly differentiated into recognizable organs or limbs, is very rare, and has a high mortality rate. Types 2 and 3 are often associated with other developmental craniofacial anomalies.^{4,12-14,16-21}

Of the teratomas seen in the first 2 months of life, 82% are sacrococcygeal.²² The head and neck region is the second most common location for teratomas in early infancy, accounting for five (14%) of those cases.² The majority of extragonadal teratomas, including those of the head and neck, manifest during childhood, although they represent less than 5% of all pediatric neoplasms.²²

Most of them include fat and cartilage as well as bone and nervous tissue. There are various theories about the etiology: traumatic implantation in the tissue, failure of somatic regions to fuse during embryogenesis, or implantation of pluripotential cells that disorganize growth in the tissue.^{7,8} The main differential diagnosis is encephalocele or meningoencephalocele, and it is important to know before deciding on the operation. Other differential diagnoses are cystic hygroma, cyst of branchial cleft

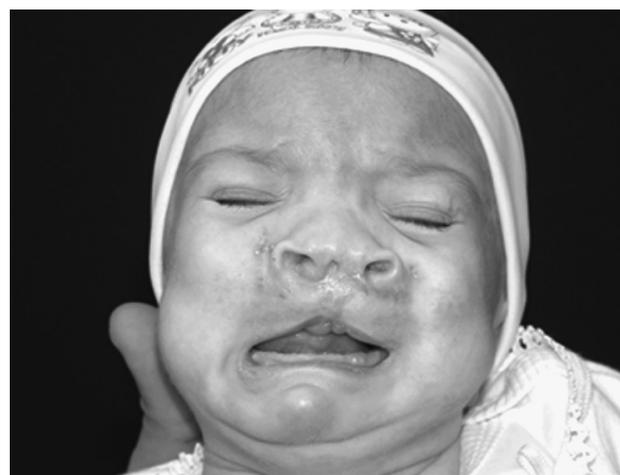


Fig 4 Early postoperative image.

or thyroglossal duct, rhabdomyosarcoma, and haemangioma.⁸

Tongue choristoma is another anomaly that can result in a cleft palate. It is difficult but not impossible to distinguish a teratoma from a choristoma. Choristoma is a tumor-like mass of tissue or an organ that is not indigenous to the organ in which the lesion is located and that results from growth to displaced primordial tissue. Simply, the tumor is more likely to be a teratoma if calcification is present.²⁴

Nasopharyngeal teratomas are usually limited to the region of the nasopharynx and oropharynx, but extensive neck involvement has rarely been reported.²⁴ They are usually sessile or have short pedicles and may completely obstruct the upper airway causing severe respiratory distress. Both teratoids and true teratomas are associated commonly with extensive skull deformities such as anencephaly, hemicrania, and palatal fissures.^{24,25}

As opposed to the cases in the literature, in the nasopharyngeal teratoma case we have presented, bilateral cleft palate/lip and columellar sinus pathology accompanied the mass. Macroscopic examination performed after the excision of the mass, which was large enough to lead to breathing and feeding problems, showed that the 8 × 5 × 7-cm mass had the typical cilia arrangement and lower eyelid presentation. The mass was diagnosed as well-differentiated true teratoma on the histologic examination.

The patient, whose first surgical treatment aimed to ensure the airway and feeding by removing the mass, additionally underwent a lip reparation operation. The patient, whose feeding and growth progress normally, is followed for cleft palate, columellar sinus reparation, and any possible recurrences.

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