Bir Yenidoğanda Epignatus – Orofaringeal Teratom

Epignathus – Oropharyngeal Teratoma in a Newborn

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ABSTRACT

Epignathus also known as oropharyngeal teratoma is a very rare tumor in newborns, which has a female predominance. The treatment is simple excision of the mass surgically which should not be delayed. We operated a male newborn due to intraoral mass causing episodic deglution and respiration problems. The histopathology was reported as epignathus and this case is discussed based on the current literature because of its rarity.

Key Words: Epignathus, Newborn

ÖZ

Epignatus ya da orofaringeal teratom yenidoğanın nadir bir tümörü olup daha çok kız cinsiyette görülür. Tedavisi cerrahi eksizyondur. Ağız içi kitleye bağlı aralıklı beslenme ve solunum problemleri nedeniyle postnatal ikinci gününde opera edilen erkek yenidoğan olgu nadir görülmesi sebebiyle literatür bilgileri eşliğinde tartışıldı.

Anahtar Kelimeler: Epignatus,Yenidoğan

INTRODUCTION

Teratomas are the most common congenital tumors in newborn infants (1).Intraoral teratom or epignathus is a rare congenital tumor that originates from the upper jaw, palate, and sphenoid bone (2). Also the confusion of epignathus is a mature teratoma or a parasitic twinning process, has yet to be clearly answered. Both of the theories are concluded with surgical excision because they are locally invasive and can be destructive (3-5). Tumors of the oral cavity other than cystic and hamartomatous lesions are rare and include ranula, lymphatic malformation, dermoid cyst, epidermoid cyst, heterotopic gastrointestinal cyst and duplication foregut cysts (6). The epignathus consist of 2% of all congenital tumors and is generally solitary and may protrude out of the mouth. It can cause life-threatening airway obstruction resulting in asphyxiation at birth. Here we report the case with epignathus was managed successfully and followed up for 3 years with minimal facial deformity.

CASE REPORT

One day old male term infant was reffered to our department for evaluation of malformation in his oral cavity. A 19 years old mother underwent cesarean section delivery at 38 weeks after an uneventful pregnancy although she had no regular prenatal follow-ups. A mass had been noticed in his mouth after birth (Figure 1). Apgar scores were 9 at 1 min and 10 at 5 min, showing no signs of respiratory distress. On clinical examination, a pedunculated, smooth, fleshy, skin covered mass of about 4 × 3 cm in size was present in oral cavity arising from the palate. The mass was mobile and filling almost the whole oral cavity causing intermittent respiratory distress but did not required immediate intubation or tracheostomy. No other co-morbidity was evident. For diagnostic complementation, beta-human chorionic gonadotropin (B-hCG) and alpha-fetoprotein (AFP) were evaluated. The concentrations were in normal limits for age. Surgery was urgently required because of intermittent breathing and feeding difficulties on postnatal second day. The

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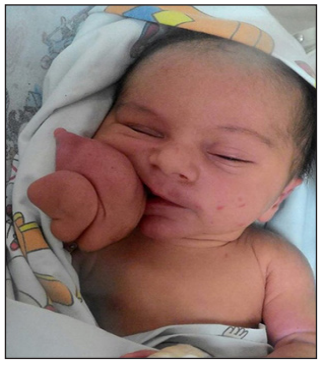

Figure 1: Two days old boy with a tumour protruding through the mouth.

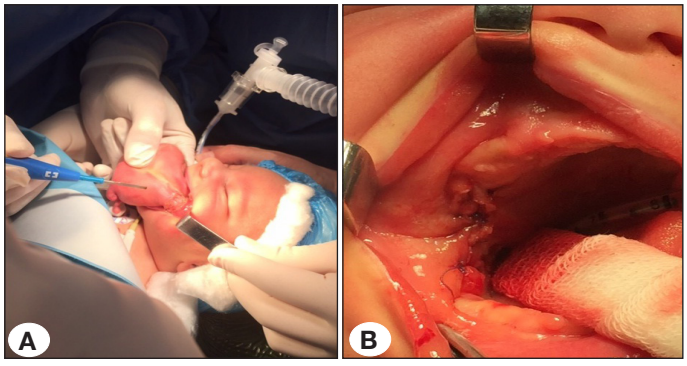

Figure 2: (A) Hard palate the base of the mass. **(B)** Excision of the mass by the use of cautery.

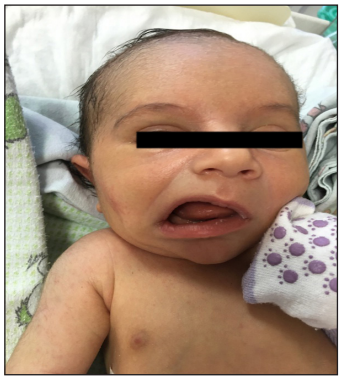

Figure 3: Fascial deformity on postoperative fifth day.

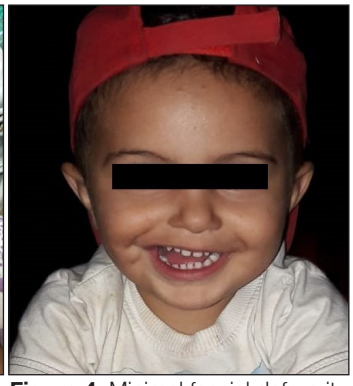

Figure 4: Minimal fascial deformity after three years follow up

lesion was pedunculated and the base was connected to the right upper gingiva, the mass was simply excised from the base by the use of cautery after orotracheal intubation (Figure 2A-C). Histopathological examination showed a well-differentiated mature teratoma, which was covered by keratinised skin containing hair follicles and epidermoid structures. Internally, there was mature fat tissue and a tooth bud. Microscopic examination showed mature epidermal and connective tissue associated with mucosal hyperplasia but no atypia. The baby was fed from nasogastric tube in the early postoperative period, he tolareted full oral feeding at the postoperative fifth day because of fascial deformity (Figure 3). He is currently in good general health and showed no clinical signs of tumor recurrence except minor fascial deformity due to prolonged intrauterin mass effect (Figure 4). With a preserved quality of life at 3 years of age.

DISCUSSION

Congenital oral tumors are commonly recognized at birth or just after birth except in instances where the tumor is very small and causing no obvious symptoms. Teratomas are rare tumors composed of tissue originating in all three embryonic layers, with an occurrence of 1:20.000 to 1:40.000. Only 5% occur in the head or neck, and, of these, malignant transformation has been reported in less than 5% of cases (7-9). The clinical manifestations depend on the size and location of the lesions. When teratomas are found in the head or neck, high perinatal mortality results, mainly caused by neonatal upper airway obstruction, which occurs because the local tumor usually impairs fetal swallowing and causes pharyngeal obstruction by its mass (10). Oropharyngeal teratoma or epignathus constitutes less than 2% cases of congenital teratoma (10-12). Its incidence ranges from 1:35.000 to 1:200.000 live births and has a female predominance (13,14). Ewing (1940) classified oropharyngeal teratoma into dermoids, teratomata and teratoids, and epignathi (12). Dermoids or hairy polyps are the most common and tend to affect adults. More than 100 cases of hairy polyps have been reported in the literature, whereas true epignathi are very rare (10). Giant epignathi are highly organized teratomas containing recognizable organs and are regarded as parasitic fetuses. Approximately 60% of epignathi originate in the nasopharynx, predominantly in females and frequently associated with cleft palate which may be severe (10,15). Approximately 6% of patients with epignathus have associated anomalies, including other facial abnormalities, branchial cleft cysts, and congenital heart disease (11). Midline abnormalities like cleft palate may accompany the epignathus (16-18). In literature, multiple localized epignathus has been reported (19).

The treatment of epignathus depends on the extent and localization of the lesion. The most important concern is the risk of asphyxiation immediately after birth, from life-threatening obstruction of the upper airway by the tumor. Antenatal diagnosis is helpful in fetus in which the mass can compress the airway causing respiratory embarrassment. Early diagnosis will help the surgeon to plan for an ex utero intrapartum treatment (EXIT) procedure (20). Local recurrence after excision due to the degree of tumor extension, permanent facial deformity, nerve damage may list as the post operative complications.

CONCLUSION

In a patient with an epignathus the treatment option is exclusively surgical, and complete resection is curative in most cases during the early neonatal period. If the mass is not resected it will continue to grow and destruct the structures by compressing.

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