Case Report - Congenital Deformity

Congenital facial teratoma in a neonate: Surgical management and outcome

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ABSTRACT

Teratomas are among the most common tumors of childhood, but craniofacial teratomas are rare. They can be diagnosed antenatally. Craniofacial teratomas may cause airway obstruction in the newborn. We present a case of a newborn male child who was diagnosed to have a facial tumor in the 8th month of gestation. He was delivered normally and had no respiratory or feeding difficulties. He was also found to have a cleft palate. Serum alpha fetoprotein levels were normal. He underwent excision on day of life 9. At 11 months follow-up, he is well with no evidence of recurrence and good functional outcome.

Keywords: Antenatal, cleft palate, excision, facial teratoma, neonate

INTRODUCTION

Teratomas are among the more common tumors of childhood with an incidence of 1:4000 live births.[1] They are most commonly located in the sacrococcygeal region followed by the gonads (ovaries or testes), anterior mediastinum, retroperitoneum, and the head and neck in that order of frequency. Craniocervical teratomas (which include facial teratomas) account for only 2–5% of all childhood teratomas with an incidence of 2.5–5/100,000 live births.[2] Within this group, cervical teratomas are more common and can cause neonatal airway obstruction.[2,3] Lateral facial teratomas are rare but can cause deformities of the zygoma and the orbit, and may involve the facial nerve. However, they are usually benign and malignant transformation is extremely rare.[1-3]

CASE REPORT

A 3.1 kg male child born by full-term normal vaginal delivery at our institute was referred to the pediatric surgery service at birth in view of a large mass lesion of the left cheek. The lesion had been detected on antenatal ultrasound scanning in 8th month of gestation (28.3 weeks). There was no polyhydramnios in the mother. The child cried immediately at birth and did not show any signs of airway obstruction. He was also able to feed normally. He had a cleft palate. The mass was seen to extend from the temporal fossa superiorly to the submandibular region inferiorly, causing deformity of the lateral canthus and the nasolabial fold anteriorly [Figure 1]. Posteriorly, the ear was not involved. The mass was variegated in consistency, being cystic in most areas and firm in others. There was no other mass lesion in the head and neck, and no clinically evident lymph nodes.

The child had a normal hemogram. Serum alpha fetoprotein (AFP) on day 2 of life was 40,000 IU/ml, which was within the normal range for age. A contrast-enhanced computed tomography (CT) scan was performed which showed a variegated lesion with solid and cystic areas and areas of calcification, impinging the temporal

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and infratemporal fossae causing scalloping of the adjacent bones but with no bony invasion [Figure 2].

The patient underwent surgery on day 9 of life. The tumor was exposed via a preauricular and temporal incision (lateral facial approach). The facial muscles were thinned out and stretched over the mass, being incorporated into the mass in some areas. The branches of the facial nerve were also stretched over the mass. The parotid gland was displaced posteriorly. The mass was excised *in toto*, taking care to preserve as much muscle as possible as well as the nerves. The skin was closed primarily over a suction drain, with a plan for reconstruction at a later date [Figure 3].

The postoperative course of the patient was uneventful. He did not require ventilatory support, and feeding was started through a nasogastric tube on the same evening. Oral feeds were started after 48 h. Regular breast feeding was possible by day 3. The child developed a minor superficial wound infection which healed well. The drain was removed on day 5 and the patient was discharged home on day 7.

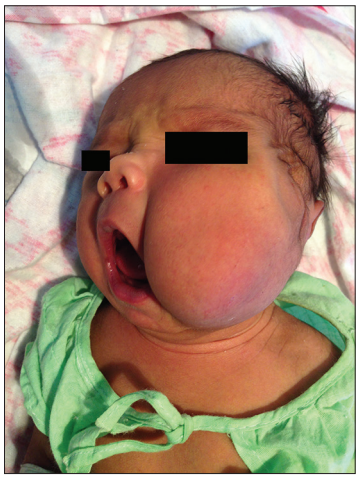

Figure 1: A clinical photograph of the tumor

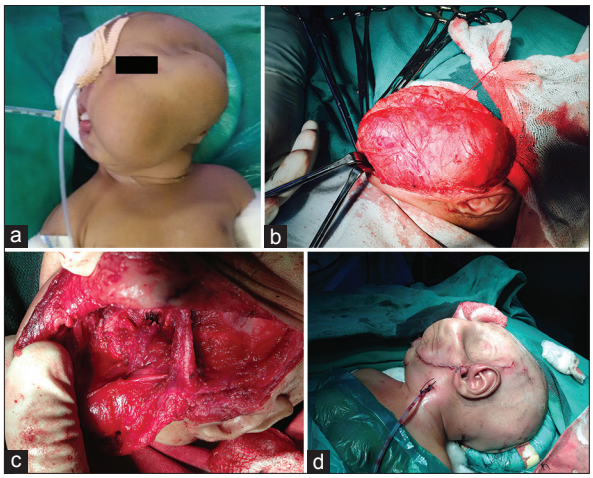

Figure 3: (a) The full extent of the tumor appreciated at surgery. (b) The exposed tumor with the stretched muscles and nerves. (c) The tumor bed. (d) The wound was closed primarily over a suction drain

Histological examination of the mass revealed a mature tridermal teratoma with no immature or malignant component. The tissues identified included-mature stratified squamous and respiratory epithelium (ectoderm); muscle, cartilage, adipose tissue, pancreatic tissue (mesoderm), and salivary gland tissue (endoderm).

At 11 months follow-up, the child has no signs of recurrence. He is growing normally and is feeding well. There are no complaints with respect to feeding and closure of the ipsilateral eyelid [Figure 4].

DISCUSSION

The head and neck is an uncommon site for teratomas.[1] Although these tumors rarely show malignant transformation, they pose a challenge in terms of achieving complete resection and in achieving good functional and cosmetic outcome. Cervical teratomas are often a threat to the survival of the neonate as they can cause severe airway compromise.[2,3] It is important to diagnose these lesions antenatally, ensure *in utero* transfer to a higher center and intervene immediately at birth. Many of these children are treated by *ex utero* intra partum therapies (EXIT procedures) that may include aspiration of the cystic component or resection of the mass.[2,3] Mortality

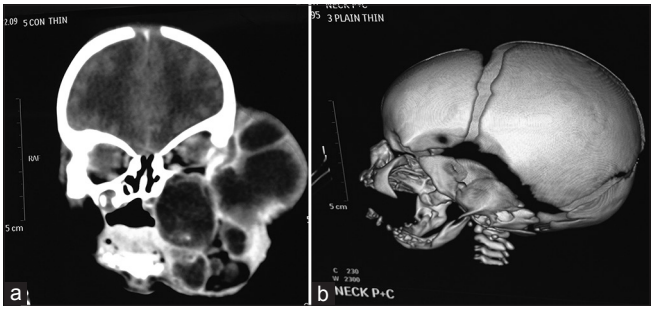

Figure 2: (a) The computed tomography scan showing a variegated lesion with solid and cystic areas and areas of calcification, impinging the temporal and infratemporal fossae causing scalloping of the adjacent bones but with no bony invasion. (b) Three-dimensional reconstruction of the computed tomography scan showing scalloping of the bones without invasion

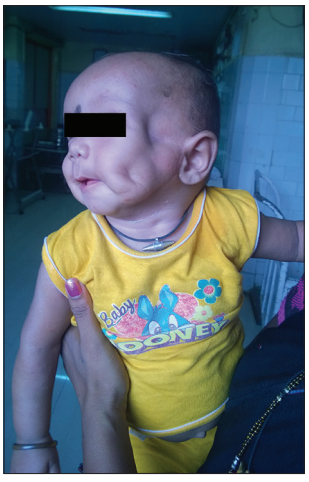

Figure 4: The patient at 6 months of age

however remains high in these cases. Neonatal compromise can be predicted antenatally by the presence of polyhydramnios and by the tracheo-esophageal index (the sum of the lateral and ventral displacements of the trachea and the esophagus from their normal anatomic positions.).[3] Head and neck teratomas also carry a significant risk of intra uterine death, mainly as a result of high output cardiac failure in extremely vascular lesions.[3]

Facial teratomas are often misdiagnosed antenatally as cystic hygromas, lymphangiomas, and even encephalocoeles, especially when they are predominantly cystic.[2] A magnetic resonance imaging is the investigation of choice to delineate these lesions *in utero* and plan surgical excision.[2,4] Postnatally, a CT scan can give the required information regarding components, surgical margins and involved structures.[2] These tumors are usually isolated, but cleft palate is a common association when the tumor develops before 8 weeks of gestation. Nasopharyngeal teratomas have been reported to occupy the area of the cleft, causing feeding difficulties.[5,6] Other associated anomalies include bifid tongue and uvula, and mandibular hypoplasia.[2] Le Saux *et al*. report a case of a lateral facial teratoma associated with Pierre Robbins sequence.[7]

The role of AFP in the diagnosis and follow-up of these tumors remains unclear.[2,3] As in our case, most patients with mature teratomas have normal serum AF*P* values for their age. Some authors propose that the AFP kinetic, even within the normal range, rather than a single value, may be helpful. Although AFP is considered a marker of malignant transformation, it has low sensitivity.[2]

Our patient had a tridermal mature teratoma. Teratomas may also be bidermal, i.e., they may show components arising from only two of the three germ layers. They are neoplastic tissue that may be mature, immature, or malignant.[8]

We excised the tumor in our patient via a lateral facial approach. This gave us adequate exposure and an esthetic scar. In the months of follow-up, the patient has shown some degree of remodeling. Paulus *et al*. have reported spontaneous remodeling of the dental arches and the maxilla following excision of a large facial teratoma in a 4-month-old girl.[4] However, our patient will require reconstruction of the zygomatic arch and the soft tissue.

CONCLUSION

Facial teratomas are uncommon tumors that can cause significant functional and cosmetic morbidity. Careful and early complete surgical excision with preservation of vital structures is desirable. Some degree of remodeling may occur after removal of the tumor. However, most patients go on to require reconstructive procedures at an appropriate age. Treatment of associated anomalies must be carried out in parallel.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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