Case Report

"Giant Epignathus" - A Case Report

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ABSTRACT

An epignathus is an extremely rare form of oropharyngeal teratoma that arises from the oral cavity, most commonly from the palate and is associated with a high mortality secondary to airway obstruction in the neonatal period. We present a preterm baby with a giant epignathus in whom airway was secured by an emergency tracheostomy after a caesarean section. The baby expired eleven and a half hours after birth. We discuss the multidisciplinary planned approach in the management of such an extremely rare teratoma.

KEYWORDS: epignathus, neonatal, oropharyngeal, teratoma

INTRODUCTION

Oropharyngeal Teratomas are very rare congenital neoplasms that distort facial anatomy and often cause acute respiratory embarassment at birth. They are associated with a high mortality secondary to severe airway obstruction and rapid asphyxia in the newborn period. Early establishment of a reliable secure upper airway is mandatory for a favorable outcome. Ideally it needs meticulous prenatal planning and prompt surgical intervention after birth by a team of experienced medical and surgical personnel.

CASE REPORT

A single live preterm baby boy was born by lower segment Caesarean Section to a p3+0+1+3 Kuwaiti mother at 32 weeks' gestation. The mother had antenatal care in a private clinic and came for admission shortly before the delivery, already in labour.

She had an ultrasound film (Fig. 1) that depicted the neck to be extended and a huge pedunculated mass measuring about 11 x 6.7 x 9.7 cm emanating from the mouth associated with polyhydramnios. The parents were non-consanguineous with no history of any congenital malformations in the family. As great difficulty was anticipated in securing the airway, the otorhinolaryngologist team was requested to be present at the operation. This team was waiting in the adjacent room with all necessary equipment laid out and ready including a pediatric rigid endoscopy set and a tracheostomy set. The baby was born deeply asphyxiated with an Apgar score of 2, 4 and 7 at 1, 5 and 10 minutes respectively. The weight was 3 kg (with the tumour), the length was 46 cm and the head circumference was 33 cm. There was a huge grotesque tumour of varying consistency from hard to soft measuring about 11 x 6.7 x 9.7 cm attached to the hard palate and coming out of the mouth (Fig. 2). There was a second polypoid mass protruding from the right nostril measuring 5 x 3 x 2 cm attached to the nasopharynx. There was no other obvious external congenital malformation.

An emergency tracheostomy was done. Resection of the tumor was postponed and a CAT Scan was requested to visualize tumor attachment to the base of the skull and also to rule out an intracranial extension. Assisted ventilation was started through the tracheostomy tube and the baby was given intensive care .The chest skiagram showed 'white out' appearance of Grade IV respiratory distress syndrome. Very high ventilator parameters were needed to ventilate the baby. At four hours of age, there was extensive bleeding from the tumour surface, the tracheostomy site and all over the body. The prothrombin time and the partial thromboplastin time were prolonged. Disseminated intravascular coagulation was diagnosed and treated with fresh frozen plasma and multiple fresh blood transfusions. As the bleeding was persistent, the baby was taken to the operating theatre at eight hours of age and the tumors were resected sub- totally (Fig. 3). The baby had systemic hypotension needing high doses of dopamine and dobutamine infusion. The baby deteriorated in spite of supportive care and expired at eleven and a half hours of age.

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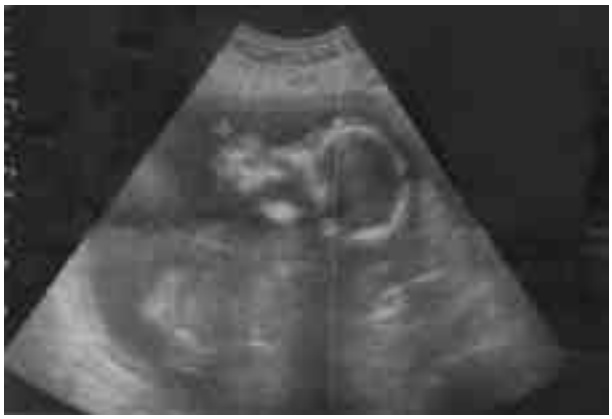

Fig. 1: The antenatal ultrasound showing the giant epignathus

Fig. 2: The epignathi *in situ* after the tracheostomy

Pertinent investigations:

Haemoglobin 10 g/dl, 7.3 g/dl. Haematocrit 0.29, 0.22. Platelet Count 25 ($x10^9$/l). Bedside Cranial Ultrasound was normal.

Pathology report: Gross:

(A). The oropharyngeal tumour (423 grams) consists of multiple nodules with varying consistency from hard to soft. There was an area of hairy skin covering. The nodules are solid and cystic brownish grey in colour measuring about 80 x 100 x 70 mm. The cut section shows hair, cartilage and bone structure with soft tissue.

(B). The nasal tumour consists of a well-defined nodule covered by soft skin weighing 17 grams and measuring 50 x 30 x 20 mm. The cut section shows bone, soft tissue. Microscopic: multiple sections from both specimens show features of a benign teratoma. Tissues from all three germ layers are seen. Ectodermal tissue in the form of stratified keratinising squamous epithelium with dermal appendages and brain tissue, mesodermal layer in the form of adipose tissue, cartilage and minimal endodermal elements are seen.

Diagnosis:

Oropharyngeal and nasal masses-Benign Teratoma.

Thus, this 32-week preterm male baby with a giant oropharyngeal teratoma and a nasal teratoma had severe asphyxia neonatorum, Grade IV respiratory distress syndrome, respiratory failure, severe hypotension, intractable bleeding and disseminated intravascular coagulation.

DISCUSSION

In 1869, Virchow coined the term "teratoma", teraton meaning "monster" 'onkoma' meaning "swelling" to describe mammoth sacrococcygeal growths[1]. Teratomas are composed of multiple tissues foreign to the organ or site in which they arise. They are poorly organized tissues derived from elements of all the three embryonic germ cell layers. Their aetiology is obscure. They are thought to arise from totipotential primordial germ cells. These cells develop among the endodermal cells of the yolk sac near the allantois and migrate to the genital ridges during the 4th and 5th weeks of gestation. Some cells may miss their target destination and give rise to a teratoma anywhere from the brain to the coccygeal area usually in the midline or para axial location. Another theory is that they are remnants of the primitive node or primitive streak. Yet another theory is that teratomas may be the result of an abortive effort of twinning[2] .

Teratomas occur with an incidence of 1:4000 live births[3]. Sacrococcygeal teratomas are the most common type (45-65%) followed by teratomas of the gonads (10-35%), anterior mediastinum (10-12%), retroperitoneum (3-5%), cervical (3-6%), presacral (3-5%), central nervous system (2-4%) and other sites (<1%)[3,4]. Oropharyngeal teratomas are extremely rare lesions and comprise less than 2% of all teratomas[3,5].

Polyhydramnios is an associated occurrence when there is extrinsic obstruction and impaired fetal swallowing[6]. The mother of this baby had polyhydramnios.

Epignathus is a term used to describe a teratoma protruding from the mouth. These tumors arise from the soft or hard palate in the region of the Rathke's pouch or in the nasopharynx in the region of the basisphenoid, tongue, sinuses, mandible or tonsil. On rare occasions multicentric tumors have been noted in more than one site[7]. Our patient had two tumors, one arising from the hard palate and the other from the nasopharynx. The clinical picture is that of dyspnea, suffocation, and difficulty in sucking, swallowing and vomiting. They distort facial anatomy and often cause respiratory embarrassment at birth. Mortality at birth, from obstruction to respiration is very high

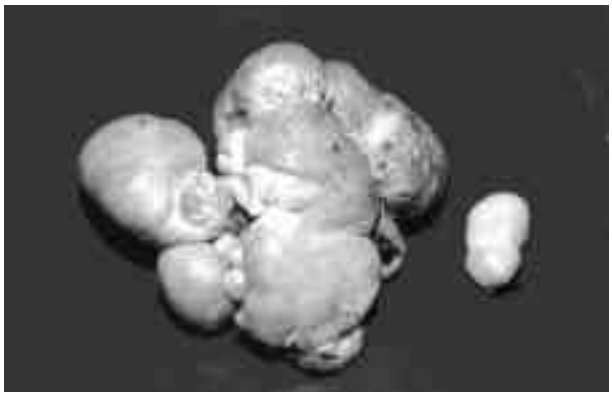

Fig. 3: The resected epignathi

(about 80to100%)[8-10]. This baby had gross distortion of the facial anatomy and very severe respiratory embarrassment needing immediate tracheostomy. In addition, this baby also had intractable hypotension and disseminated intravascular coagulation causing profuse relentless haemorrhage from all over the body including the tumor and subsequently, renal shutdown. All these complications could be the sequelae of asphyxia. There have been case reports of epignathus associated with other midline anomalies[11,12]. This baby did not have any obvious external midline anomalies. Most of the epignathi are unidirectional and protrude from the mouth but some epignathi protrude bidirectionally involving and destroying brain tissue, resulting in a poor prognosis[13]. The exclusion of midline congenital CNS lesions or intracranial extension is part of the preoperative management[3,11]. The bedside ultrasound was normal but the baby was moribund for further work up to rule out a bi-directional protrusion of the epignathus.

Recent advances in the prenatal diagnosis permit meticulous planning and team effort by a perinatal multidisciplinary team including a high-risk obstetrician, neonatologist, pediatric otolaryngologist, and an anesthesiologist[6,8,9,14-16]. Since the mother came to the hospital shortly before delivery there was no sufficient warning or time to do detailed planning and orchestrate a more elaborate team effort. Fetal airway obstruction can make it difficult if not impossible to secure the airway at birth, before hypoxia, brain injury, or death results. Hence a recommendation was issued that the multidisciplinary team should be waiting in an adjacent room to secure an airway for the baby after a cesarean section was done in an adjoining operation theatre[17]. This "adjoining room" recommendation has been replaced by yet another remarkable breakthrough. This exciting recent advance is the assembling of a multidisciplinary management team and doing an elective caesarean delivery of the fetal head and thorax, under conditions of uterine tocolysis, permitting a controlled evaluation of the airway and endotracheal intubation while oxygen supply was maintained through the placenta. Thus the EXIT procedure (EX-utero Intrapartum Technique)[8,18-20] or the OOPS procedure (Operation On Placental Support)[16] consist of securing the airway of the fetus, partially delivered and still connected to the placenta. This technique leaves an intact feto-placental circulation and guarantees a normal fetal oxygenation while fetal airway patency is secured[10,17,18]. After a cesarean section delivery of the infant from the uterus, the umbilical cord was not clamped and the feto placental circulation was left undisturbed. Then a tracheal intubation or tracheostomy was performed. Once a secure, stable airway was obtained, the cord was clamped and the infant was stabilized.

CONCLUSION

Epignathus is a potentially lethal fetal anomaly and 80 to 100% of these cases succumb to acute respiratory distress secondary to airway obstruction at the time of birth[6,8,9,10,16,19]. There is little hope of survival without a multidisciplinary team and the EXIT technique[8,9].

These lesions can be detected early by prenatal ultrasound and a strategic plan developed early in the prenatal period. The airway management should be tailored to each individual patient. A multidisciplinary team should be formed and there should be meticulous planning to coordinate the available expertise to manage these complex situations. A suitable algorithm of airway management should be clearly envisaged. The fetus should be delivered by using the extra uterine intrapartum treatment (EXIT) procedure with intact materno -fetal circulation and gas exchange[18-20]. Thus, invaluable time is redeemed to perform procedures like direct laryngoscopy, bronchoscopy or tracheostomy to obtain a stable secure airway.

With such precautions, a potentially dramatic emergent airway crisis scenario can be converted into a controlled "emergency" situation thus reducing the severe morbidity and mortality in this extremely rare congenital condition.

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ADDENDUM

The name of the co-author, Bader A Abdul Kader, Department of Medicine, Farwaniya Hospital, Kuwait was missed in the article titled "**Increased Blood Pressure Variability in Hypertensive Patients with Left Ventricular Hypertrophy**"published in the issue 2004; 36(2):98-102), which is added to the authorship.