A RARE CASE OF EPIGNATHUS- CASE REPORT

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Abstract:
An Epignathus is a very rare form of oropharyngeal teratoma arising from the oral cavity. Most common site of origin is the palate. We report a rare case of Epignathus in a 23 week still born baby. It was associated with cleft palate and there was no intra cranial extension which was confirmed by post mortem MRI and pathologic examination.

Keyword: Epignathus, Teratoma

INTRODUCTION:
Teratomas are tumors composed of tissues derived from all the three germ layers -ectoderm, mesoderm and endoderm. The incidence of teratoma is about 1:4000 live births(1). Epignathus is a very rare oropharyngeal teratoma arising from the palate, sphenoid and upper jaw(2),(3),(4),(8). The incidence of epignathus is 1 in 35,000 to 1 in 2,00,000 live births. Epignathus is more common in females than in males, with a ratio of 3:1(5),(6),(8). We report a rare case of epignathus in a 23 week still born female baby.

CASE REPORT:
A 30 year old woman, G2 P1 L1, with a 1 1/2 year old alive, healthy girl baby was admitted for the complaint of polyhydramnios at 22 weeks of gestation. She had no history of consanguineous marriage. There was no history of drug intake or infection in the present pregnancy. Her USG revealed a large oropharyngeal teratoma. So hysterotomy was done and a still born baby with a huge mass arising from the mouth was delivered.

PATHOLOGIC EXAMINATION:
We received a fetus with wide opened mouth and a mass protruding out from the oral cavity. The fetus measured 34 cm in length and weighed 540 gms. The mass measured 16x9x4 cm (Figure 1). The mass was seen to originate from the hard palate. There was a cleft in the palate. External surface of the mass was lobulated and variegated. A soft tissue was seen to cover around the base of the mass for about 5 cm. Consistency of the mass was variable from hard to soft.
Postmortem MRI was done and the mass was attached to the hard palate and there was no intra cranial extension (Figure 2). There was associated cleft palate.

Microscopic examination showed tissues derived from all the germ cell layers. Primitive neuroepithelial tubules, choroid plexus, glial elements, cartilage, bone, squamous epithelium, hair and immature mesenchyme are present (figure 3 and 4). Among these, neural elements predominated. Sections from the placenta and the umbilical cord showed normal histology.

Figure 3: Photomicrograph of the tumor showing cartilage, bony trabeculae and squamous epithelium.

Fetal autopsy was performed. There was no other associated anomaly apart from cleft palate.

MICROSCOPY:
Figure 4: Photomicrograph of the tumor showing choroid plexus, neural tubules and cartilage.

DISCUSSION:
The term teratoma was coined by Virchow in 1869, teraton meaning monster, onkoma meaning swelling to describe mammoth sacrococcygeal growth (7). Teratomas are derived from all the three germ cell layers. Totipotential primordial germ cells are thought to give rise to teratoma. Oropharyngeal teratomas are very rare lesions and they comprise less than 2% of all teratomas. On gross examination, teratomas are usually multinodular. Cut surface varies from solid to multicystic. Nodules of translucent, white cartilage may be seen. Cysts are filled with clear or mucoid fluid or keratinous material. In areas of immature tissue, fleshy, encephaloid character is present. 

The microscopic complexity of teratoma is an immediate clue to its germinal nature. Usually a mixture of mature squamous epithelium (often with cutaneous appendages), foregut type columnar epithelium, bone, cartilage, fat, neuroglial elements and fetal or mature striated muscle tissue are present. Other potential histologic components are islands of hepatocyte, pancreas, choroid plexus and pigmented neuroepithelium resembling retinal tissue.

Immunohistochemical staining of teratoma yields the expected results for the specific element examined. Thus chromogranin is present in neuroendocrine cells, neural markers in neural tissue, cytokeratins in epithelia, vimentin in stromal tissues and desmin in muscle. Tumor size, nasopharyngeal involvement and intracranial spread are poor prognostic factors, since they cause mechanical airway obstruction and feeding difficulties. These factors should be considered in deciding the management. At birth due to obstruction of airways, mortality is very high. When prognosis is favourable, ex utero intrapartum technique (EXIT) with intact materno-fetal circulation and gas exchange is an option (4).

REFERENCE: