EPIGNATHUS: REPORT OF A CASE

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SYNOPSIS

Epignathus is a rare congenital teratoid tumor which arises from the cavity of the mouth or upper part of pharynx and often possesses recognisable foetal parts.

A case presenting with dystocia is described.

Epignathus is a monster consisting of a more or less normal foetus with a mass which often arises from the base of the skull and protrudes through the mouth.

The earlier records of these rare tumors were fully reviewed by Hill (1885) and Windle (1899). Only about 60 cases have been documented up to 1899 (Windle). Since then, few cases have been recorded (Strachan, 1930; Hankins and Harding, 1932; Kesson 1954; Bennett, 1970). The following is the report of another case complicated by dystocia in the second stage of labour.

Case Report

Madam T.S.L., a 27 year old Chinese female, Gravida 6 Para 5 was admitted to hospital on 1st of September 1970 with premature labour, at 28th week of gestation. Her previous 5 pregnancies and deliveries had been uneventful.

Physical examination revealed that her general condition was satisfactory. She was not anaemic. The B.P. was 120/80 and there was slight ankle oedema. Heart and lungs were clinically normal. The uterus was unduly large with abundant liquor, the fundus almost reaching the xiphisternum. Foetal heart sounds were not heard.

Vaginal examination showed that the cervix was fully dilated and the membranes were intact and buldging. Artificial rupture of membranes released about 4 pints of brown colored liquor. The foetus presented as a breech with flexed legs. The lower limbs, the body and the arms were delivered easily but there was obstruction to the delivery of the after-coming head. Abdominal palpation revealed that a large irregular foetal mass was still within the uterus and possibly attached to the foetal head. The after-coming head was finally delivered by traction employing rocking manouver. During the manipulation, 2 soft cystic masses were separated from the main tumor and these were manually delivered.

The male stillbirth weighed 2 lbs. 12 ozs. and measured 16½″ in length (Fig. 1). The limbs and body did not present any abnormality, but protruding from its widely opened mouth was a large lobulated mass measuring 10″ × 8″ × 6″ and attached to the roof of the mouth by a stalk measuring 1 inch in diameter. When viewed from the front, it almost entirely covered the face.

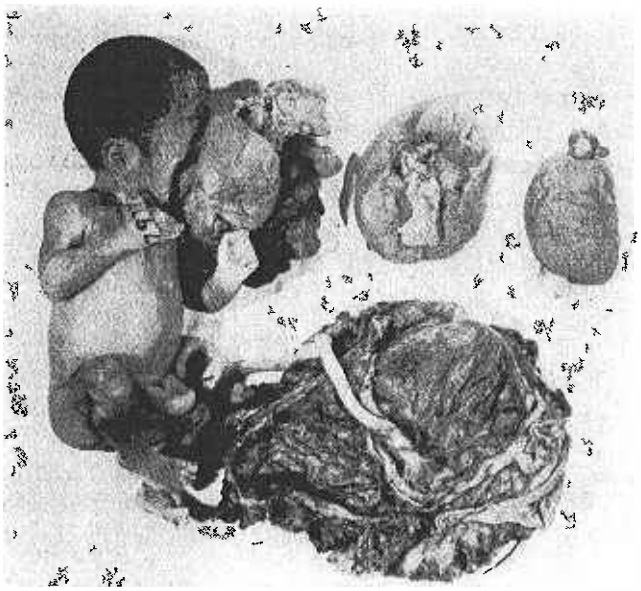

Fig. 1. Gross specimen of the stillbirth showing a large lobulated mass protruding from the mouth and two primitive limbs.

The lobules on the tumor varied from ½″ × ½″ to 8″ × 7″ in size. Some areas were cystic containing yellowish or greyish jelly like material. Some areas were solid containing pieces of bone and cartilage. Skin with fine hairs was seen in certain lobules. The 2 cystic masses which were separated during delivery of the foetus were definite primitive limbs with digit buds.

The placenta and membranes appeared normal but weighed 1 lb. 12 ozs. X-ray report of the tumor

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stated "the giant teratoma seems to originate from the pharyngeal cavity producing marked depression of the mandible. It consists of 3 components—the proximal being the largest and lobulated and shows a bizarre pattern of bone structure. The second component shows a few osseous structures whereas the third shows some primitive limb buds with bone formation" (Fig. 2).

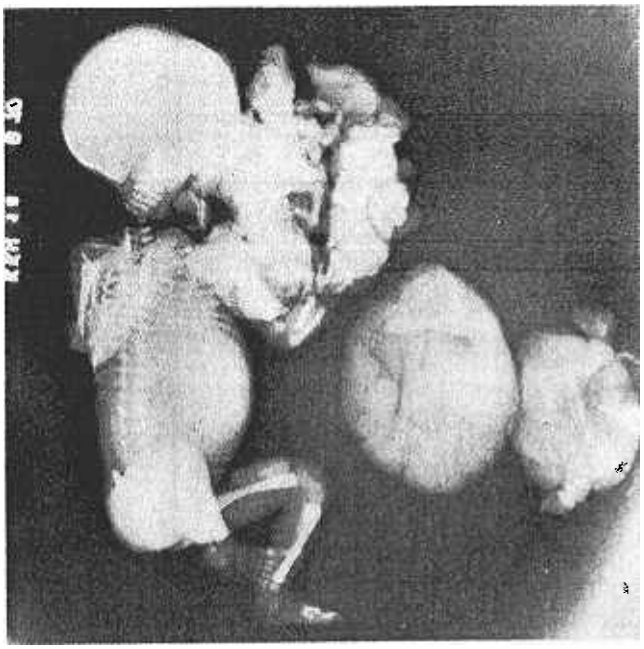

Fig. 2. X-ray of the stillbirth showing bizarre bone structure in the giant teratoma and bone formation in one of the primitive limbs.

Postmortem examination of foetus revealed no abnormality in the cardiovascular, respiratory, alimentary and urogenital systems.

Histological section of the tumor showed "presence of numerous elements including epidermis, sebaceous glands, mucosa tissue, neural tissue, cartilageous, and bony elements. Some poorly differentiated hyperplastic glandular tissues in acinar formation were also seen. The placenta was normal".

DISCUSSION

The name "Epignathus" was first given by St. Hilare, who based his description of it on a single imperfectly described case.

It is a rare congenital teratoid tumor, often possessing recognisable parts of a foetus and is attached within the cavity of the mouth or upper part of the pharynx.

The tumor is present at birth and may occur in premature infants as in the present case, or full term infant, who often dies in the neonatal period. When the tumor is small, it can be surgically excised and the patient may survive (Abraham, 1881). Recently Bennett (1970) reported a patient who survived after excision of a large epignathus at birth.

Like teratomata occuring in other sites, the etiology remains unknown. Potter (1961) states that there is a natural progression which can be traced from normal twins to conjoint symmetrical twins, asymetrical twins, parasitic foetuses, foetal inclusions and teratoma. On the other hand, Willis (1960) holds this view that a teratoma is a true neoplasm, arising from foci of pleuripotential embryonic tissue, differentiating according to its own "labile determinations" and producing a variety of tissues foreign to the part in which it grows.

Windle (1899) found that the most common instance of attachment of the tumor is to the palate, as in this case, or to the superior maxillary bone of one or other side, because the presence of the tumor often caused an almost entire absence of the palate.

Some are attached to the base of the skull or other part of the pharynx. Some even have double attachments.

The attachment to the base of the skull can either be to the lower surface of the bone or the mucous membranes which invests the upper part of the pharynx. In some cases, the stalk of the tumor is distinctly traceable through the pituitary foramen. In others, the tumor is hour-glass in form, with intracranial and extracranial parts connected by an isthmus traversing the basisphenoid.

The nature of the contents varies. The most simple form is the skin-clothed pharyngeal polyp, with a core containing usually a rod of cartilage, connective tissue and striated muscles. Next, there is a group in which the tumor is much larger, often polycotyledonous and contains pieces of cartilage, bones, muscles, but no organ of recognisable character. In a further group, bones of recognisable shape, extremities, usually the distal part, may be present. Neural tissue, liver cells, intestine, and rudimentary genitalia have all been described. To this group belongs the case described in this paper.

ACKNOWLEDGEMENT

We wish to thank Professor S. S. Ratnam for permission and encouragement to study and report on the case and Professor E. Tock for performing the post-mortem examination on the foetus.

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