Cerebral Teratoma Associated with Epignathus in a Newborn Infant*

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The association of an intracranial teratoma with an epignathus is uncommon in the newborn infant and only 2 cases have been reported previously.2,6 This is the report of an additional case.

A teratoma is both a tumor and a congenital malformation, composed of adult and embryonic tissues of different types, which may or may not be foreign to the parts where it is found. An epignathus is a teratoma attached to the roof of the mouth or pharynx.

In a review of the literature on the subject of intracranial teratoma of the newborn, Greenhouse and Neuburger5 found 25 cases reported. They divided these into three groups (Table 1): 1) The largest consisted of stillbirths and all except in the case reported by Kraus,6 were hydrocephalics. 2) Hydrocephalics who died shortly after birth. 3) Infants apparently normal at birth who, within weeks, presented a gradual enlargement of the head. The sex ratio was 12 females to 6 males; the sex was not recorded in the other 7 cases.

This report is concerned with a case of epignathus associated with a cerebral teratoma. The latter revealed itself as a calcified intracerebral zone on radiological examination of the foetal head.

Case Report

Clinical History. The mother, aged 36, was known to have a patent ductus arteriosus. She had one healthy boy, aged 3 years. During this pregnancy she suffered from subacute hydramnios which appeared during the 7th week, and the onset of labour was spontaneous at 31 weeks. There was a precipitate delivery following a second stage lasting only 5 min. The infant never really established respiration and died 55 min. after delivery.

Post mortem Findings. The body was that of a small, premature male infant, weighing 1842 g., with a crown-heel length of 39 cm. An epignathus protruded through the mouth (Fig. 1). On dissection it was found to fill the mouth and nasopharynx and was attached to a very narrow area, about 4 cm. long, in the nasopharynx, just below the posterior nares. Two polypoid prolongations of the tumour passed upwards through the nares into the nose. This tumour, weighing 50 g., was lobulated, and some of its tissue was soft and hemorrhagic; elsewhere it was very firm and appeared to be bony or cartilaginous. The circumference of the enlarged skull was 32 cm., and the bones were thin. The anterior fontanelle was wide. On opening the skull a large amount of slightly bloodstained cerebrospinal fluid escaped. On removal of the brain, which weighed 290 g., there was a large saccular dilatation of the leptomeninges over the right frontal pole. On radiological examination of the brain an area of calcification was seen in the right frontal pole which corresponded to a mass of tumour found on slicing the brain. The tumour, 80 mm. by 40 mm., was sessile and lay loosely in the lateral ventricle. On cutting, it was gritty and contained cartilage. No abnormality was found in the cardiovascular, renal, alimentary, endocrinologic or genitourinary systems.

Histology. Representative blocks of the brain, cerebellum, brain stem and somatic organs were processed in paraffin and celloidin, and stained with hematein-eosin and haematoxylin-van Gieson, for general purposes.

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TABLE 1

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* Reported by Greenhouse and Neuburger.5
Fig. 2. Rosette formed by ependymal cells. Haematoxylin and eosin.

Fig. 3. Intestinal mucosa in teratoma of brain. Haematoxylin and eosin.
with Nissl stain for the cells, Heidenhain’s stain for myelin and Mallory’s phosphotungstic acid-haematoxylin stain for glia.

Central Nervous System. The gyri and cortex of the parietal, frontal, occipital and temporal lobes were normal. Dense aggregates of glial and neural cells were present around the dilated lateral ventricles, particularly the inferior horns. An outer granular layer was situated on the surface of some parts of the cortex. There was little myelin present in the hemispheres, as one would expect for this age. The cerebellum showed some myelin, a normal dentate nucleus, a normal Obersteiner or external granular layer but no Purkinje cells. The brain stem showed myelination of the medial longitudinal bundle, medial lemnisci and pontine fibers and its cellular structure appeared normal. The general development of the brain was consistent with 31-weeks gestation, but the myelination was less well advanced than would be expected for that age.

The tumour in the right frontal lobe was a typical teratoma composed of mature and immature tissues. There were neuro-epithelial structures forming tubes with distinct mantle zones, formation of rosettes by ependymal cells (Fig. 2), aggregates of neuroblasts strewn throughout the tumour, islands of choroid plexus, intestinal mucosa (Fig. 3), cartilage and bone, and haemangiomatous tissue—all this set in a stroma of connective tissue. The most frequent cellular components of the tumour were glial cells. Some areas showed concentration of oligodendroglia with the “box” appearance seen in some tumors of such cells. Other parts contained many astrocytes, sometimes packed closely together or grouped loosely in a connective-tissue stroma; their appearance was normal, gemistocytic or with a hypertrophied nucleus.

Epignathus. The teratoma here was much more mesodermic in type, with very few glial cells present. There were islands of cartilage and bone set in a meshwork of haemangiomatous and connective tissues, intestinal mucosa, a few rosettes composed of ependymal cells, rudimentary eyes (Fig. 4), and immature pulmonary tissue. But there were no hepatic, renal or muscular tissues in this tumour.

Discussion

Breslau and Rindfleisch described a stillborn hydrocephalic female infant born spontaneously during the 24th week of gestation with a large tumour projecting out of the mouth. The presentation was by breech. It was felt that the site of origin of the growth was in the vicinity of the pituitary gland, with extensions into the brain and roof of the mouth. The cortex was greatly thinned and the ventricles were dilated. There were rudimentary eyes, umbilical-cord tissue, as well as muscle, cartilage and renal tissues and various glands.

Kraus reported a stillborn premature female infant with a teratoma in the brain and an epignathus. There was no enlargement of the head. The intracranial tumour, capped by the flattened hypophysis, occupied the pituitary fossa and was connected to the epignathus through an open craniopharyngeal canal. These tumours were similar and consisted of areas of bone or cartilage set in a stroma of connective, vascular, and neural tissues, striated muscle, cylindrical and cuboid epithelium, and islands of choroid plexus. There were also neuro-epithelial tubes and ependymal-cell rosettes. Kraus felt that these tumours were of common origin arising from a site near the pituitary fossa. This is in accord with the findings of Breslau and Rindfleisch and with the opinion of Willis who felt that these associated tumours are derived from a common source in the vicinity of Rathke’s pouch and oral membrane anterior to

![Fig. 4. Rudimentary eye in epignathus. Haematoxylin and eosin.](image-url)
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the end of the notochord. However, our own case differs, for the 2 tumours seemed more distinct than in the above-mentioned cases, and careful dissection failed to establish any sort of anatomical link between them. Moreover the histological structure of these 2 teratomas was not completely similar for, as we have seen, the cerebral teratoma was rather neural in type, whereas the epignathus was mesodermal. This may, however, be ascribable to hazard of the random sections taken from the specimens.

The pathogenesis of these teratomas is unknown. Masson believed that they are malformed foetuses. Willis disagreed with this view. But it is interesting to note that Aird, discussing conjoined twins, described a baby to whom was attached at two separate points, lower abdomen and peritoneum, a teratoma containing a peritoneal cavity, pelvic colon, rectum and two lower limbs.

The calcification noted on radiological examination of the brain could be of diagnostic value. Congenital malformations are not infrequently associated with hydramnios during pregnancy. If a roentgenogram is taken of the mother's abdomen, because of hydramnios, and intracranial calcification is found in the foetus, the presence of a teratoma may be suspected. Cushing reported seeing intracerebral calcification in the radiograph of the skull of a 2-month-old hydrocephalic child. At necropsy these calcified areas were found to be part of an intracranial teratoma.

The recent National Survey of Perinatal Mortality noted the markedly higher incidence of females among hydrocephalics and anencephalics. This is of interest when comparing the findings of Greenhouse and Neuburger in cases of cerebral teratomas, in which the sex ratio is 12 females to 6 males.

Summary

An intracranial teratoma associated with an epignathus in a newborn male is described together with a review of the reported cases. It would appear that these associated tumours show a similar histological picture, and may have a common origin near the pituitary fossa, although our findings do not completely support this. The foetal malformation may be associated with hydramnios during pregnancy. Radiographs of the mother's abdomen may reveal intracranial calcification in the foetus, and the possibility that these form part of a teratoma should be considered.

References

7. MASSON. Cited by Auger.