Congenital primitive neuroectodermal tumour presenting as obstructed labour

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Abstract

Brain tumours presenting at delivery are extremely rare. A case of primitive neuroectodermal tumour (PNET) that presented intrapartum with failure to progress due to hydrocephalus is reported. Diagnosis required imaging with magnetic resonance and computed tomography in addition to open biopsy.

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Brain tumours presenting in the first year of life are often described as congenital and account for 1.4 to 8.5% of all childhood brain tumours.[1–4] Current nomenclature differentiates between truly congenital tumours (presenting before or at delivery), neonatal tumours (presenting between birth and 2 months of age), and infantile (presenting between 2 and 12 months of age). Only 18% of those tumours presenting in the first year of life are diagnosed before or at delivery.[5] The commonest modes of presentation of these truly congenital tumours are with raised intracranial pressure, dystocia, preterm delivery and stillbirth.[6 7]

In 1950 only 14 cases of brain tumours in the first two months of life had ever been reported.[8] By 1984, 191 such cases had been reported[6] and now the phenomenon, though rare, is well recognised. Diagnosis is aided by an increased index of suspicion, antenatal and neonatal ultrasonography, and computed tomography and magnetic resonance image scanning. We present a case of infratentorial congenital primitive neuroectodermal tumour (PNET) that presented intrapartum with obstructed labour.

Case report

A 34 week gestational age boy was delivered by emergency caesarean section for obstructed labour. His mother, a 25 year old caucasian, had had two previous normal pregnancies. The current pregnancy had been uncomplicated before labour. She had booked at 12 weeks of gestation and a full anomaly scan, including examination of cerebral anatomy, had been normal at 18 weeks.

She was admitted in preterm labour on the day of delivery and proceeded normally to full dilatation. The head failed to descend on active pushing and a repeat ultrasound scan was performed by the obstetrician. This revealed hydrocephalus and cephalo-pelvic disproportion was diagnosed. The infant was delivered by emergency caesarean section, was in good condition at delivery, and did not require resuscitation. Examination showed that the baby had gross hydrocephalus with widely spaced sutures and a bulging fontanelle. Head circumference was 46 cm (far above the 97th centile) and birthweight was 3.32 kg (97th centile). There was marked hypotonia in all four limbs but no other abnormal findings or dysmorphic features.

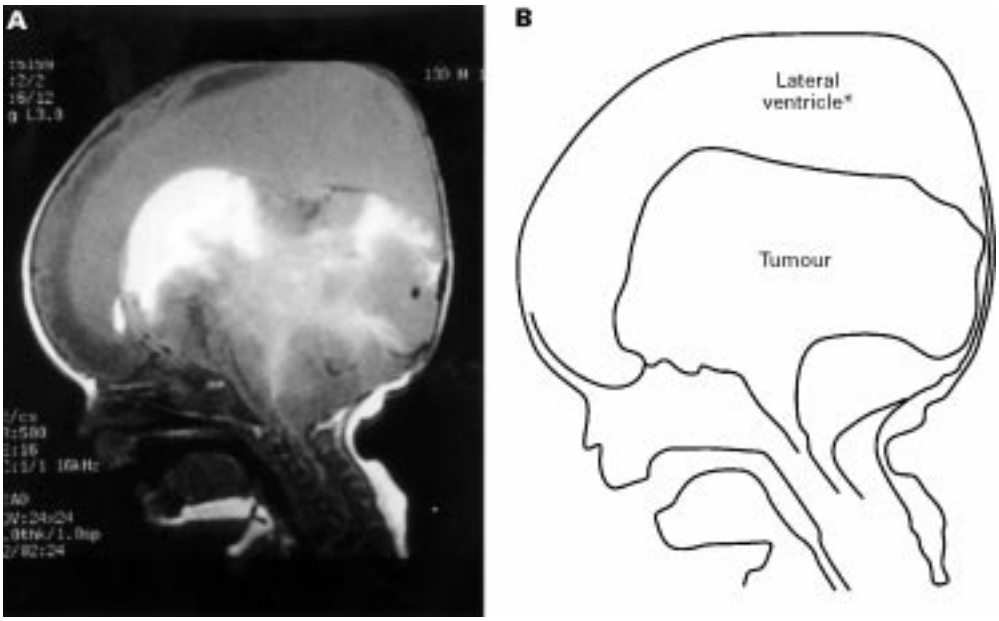

Figure 1 T1 weighted MRI scan (sagittal section): CSF looks grey due to high protein content.

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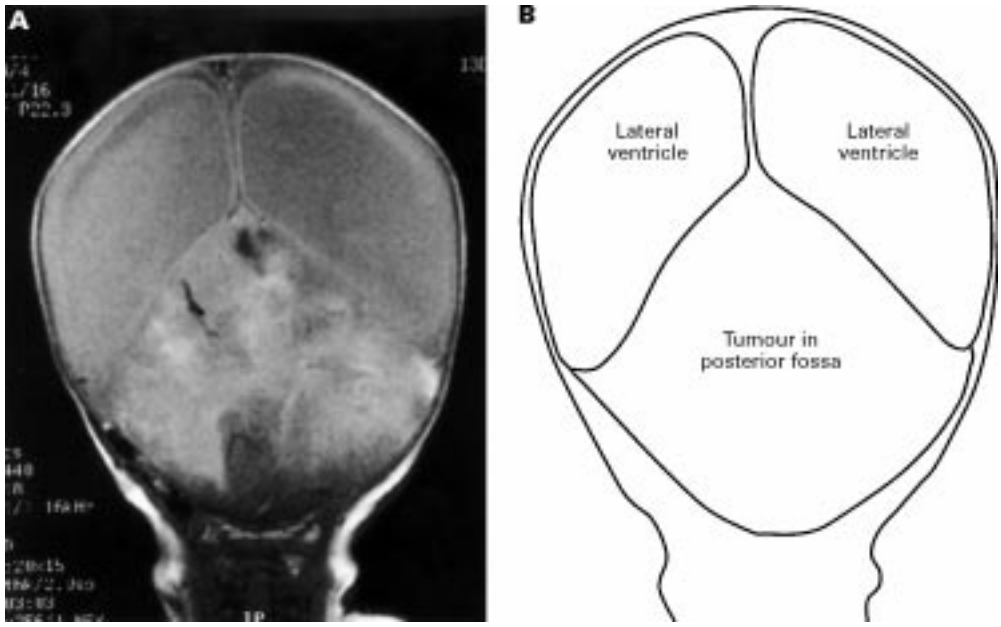

Figure 2 T1 weighted MRI scan (coronal section).

The infant was transferred to the neonatal intensive care unit for further management. Initial ultrasound scanning confirmed the diagnosis of hydrocephalus with massive dilatation of both lateral ventricles and no midline shift. An inhomogeneous mass was noted in the region of the thalamus and midbrain and no third or fourth ventricle were seen. A thin rim of cortex was noted under the cranium.

Further imaging included a computed tomogram and magnetic resonance imaging, which revealed a large midline mass occupying most of the posterior fossa. The mass was of mixed density but had minimal contrast enhancement. Extreme dilatation of both lateral ventricles was noted, with marked hypoplasia of both cerebral hemispheres (figs 1-3). The differential diagnosis was a PNET or a germinoma.

The infant required intubation and mechanical ventilation on the fourth day of life due to hypoventilation. Neurosurgical review was arranged and needle biopsy of the mass and insertion of an external ventricular drain were performed. Unfortunately, histological examination of the biopsy specimen and cytological examination of cerebrospinal fluid were not diagnostic. Most of the cells seen were necrotic. Intracerebral pressures were not raised and the drain was removed five days later.

Craniotomy and open biopsy were performed to obtain a full tissue diagnosis. Histological examination revealed a malignant embryonal tumour composed of primitive undifferentiated cells with high mitotic activity. The parents were advised that intensive care should be withdrawn and following discussion,

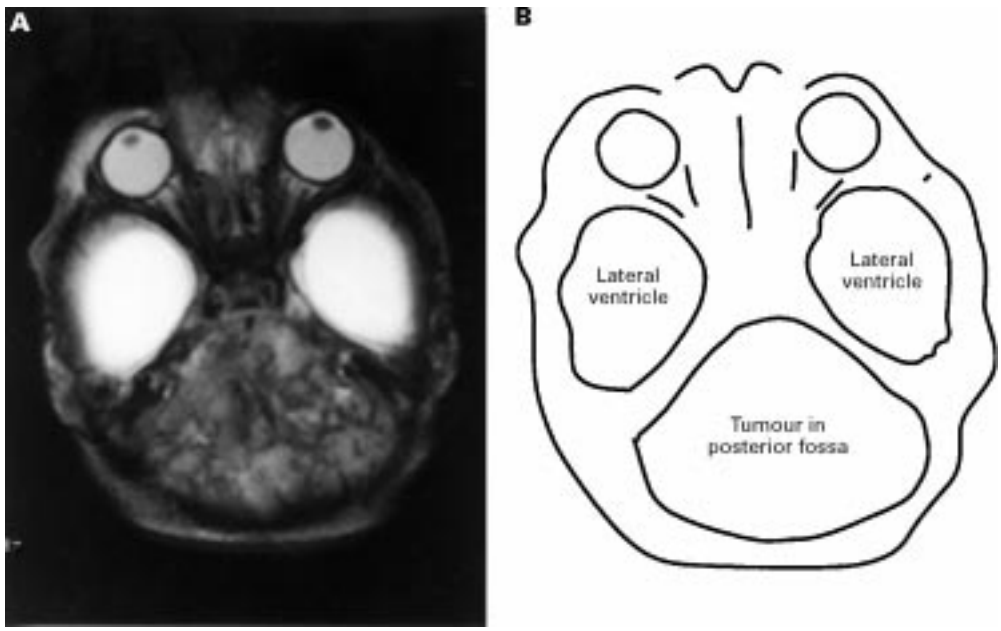

Figure 3 T2 weighted MRI scan (transverse section).

Table 1 UK study of brain tumours in the first two years of life[9]

1971–1985 n=535	
Astrocytoma (including optic nerve glioma)	25%
Medulloblastoma (including PNET)	23%
Ependymoma (including choroid plexus)	22%
Germinoma	2%
Other	25%

the infant was extubated and died peacefully in his parents' arms.

Discussion

Brain tumours in infancy have a different biology to tumours presenting later in childhood (table 1). Supratentorial astrocytomas, medulloblastomas (including PNET), and choroid plexus tumours are the most common tumours of infancy, whereas later in childhood, the commonest tumours are infratentorial astrocytomas and medulloblastomas. Tumours of infancy are predominantly supratentorial, with a shift to infratentorial predominance at around 2 years of age.[9]

Imaging of congenital neoplasms presents its own difficulties. Cranial ultrasonography has the advantage of being performed at the cotside but computed tomography and MRI provide more detailed images and are recommended for brain stem and posterior fossa tumours where bone artefact can obscure anatomical details on ultrasonography.[10] MRI provides finer detail than computed tomography and sagittal views that computed tomography cannot provide. Before effective imaging, many congenital intracranial neoplasms were diagnosed at necropsy.[11] However, there are now 20 reports of antenatal ultrasound diagnosis.[12] The median gestational age at the time of diagnosis in these cases was 31.5 weeks. In the case presented, the presence of a normal early scan suggests tumour genesis or growth in the second or third trimester of pregnancy.

The management of congenital intracranial neoplasms is controversial, and, in the absence of definitive trial data, highly individual. There is a current trend towards initial surgical management with complete excision or maximal debulking of the tumour followed by radiotherapy or chemotherapy. Various regimens have been used,[5 7 13] with a trend towards avoiding radiotherapy if at all possible in the developing brain. The current prognosis for choroid plexus papilloma is excellent, with almost 100% disease free survival following surgical resection.[5] However, the results for other histological types of tumour are variable, with two year survivals of tumours presenting at birth ranging between 7 and 70%.[5 6 9]

We suggest that there should be a high index of suspicion for congenital intracranial tumours in any infant presenting with hydrocephalus not noted on early antenatal scans. These infants present unique challenges in diagnosis, often requiring computed tomography and MRI scanning in addition to postnatal cranial ultrasonography. Management is also complex, involving dilemmas of ethics and medical management and should be determined in conjunction with a paediatric neuro-oncologist.

1 Jooma R, Kendall BE, Hayward RD. Intracranial tumours in neonates: a report of seventeen cases. *Surg Neurol* 1984;**21**:165-70.
2 Keith HM, Craig WM, Kernohan JW. Brain tumours in children. *Paediatrics* 1949;**3**:839-44.
3 Koos WT, Witt RN, Miller MH. *Intracranial tumours of infants and children*. Stuttgart: Georg Thema Verlag, 1971.
4 Ellams ID, Neuhauser G, Agnoli AL. Congenital intracranial neoplasms. *Child Nerv Syst* 1986;**2**:165-8.
5 Haddad SF, Menezes AH, Bell WE, *et al*. Brain tumours occurring before 1 year of age: a retrospective review of 22 cases in an 11 year period. *Neurosurg* 1991;**29**:8-13.
6 Wakai S, Arai T, Nagai M. Congenital brain tumours. *Surg Neurol* 1984;**21**:597-609.
7 di Rocco C, Ceddia A, Ianelli A. Intracranial tumours in the first year of life. A report on 51 cases. *Acta Neurochir* 1993;**123**:14-24.
8 Arnstein LH, Boldrey E, Naffziger HC. A case report and survey of brain tumours during the neonatal period. *J Neurosurg* 1950;**8**:315-19.
9 Stiller CA, Bunch KJ. Brain and spinal tumours in children aged under two years: incidence and survival in Britain, 1971-85. *Br J Cancer* 1992;**66** (**Suppl XVIII**):S50-S3.
10 Buetow PC, Smirniotpoulos JG, Done S. Congenital brain tumours: a review of 45 cases. *Am J Radiol* 1990;**11**:587-93.
11 Janisch W, Haas JF, Schreiber D, *et al*. Primary central nervous system tumours in stillborns and infants. *J Neurooncol* 1984;**2**:113-16.
12 Heckel S, Favre R, Gasser B, *et al*. Prenatal diagnosis of congenital astrocytoma: a case report and literature review. *Ultrasound Obstet Gynecol* 1995;**5**:63-6.
13 Zeltzer PM. Toward a cure for infants with brain tumours: the challenge or the 1990's. *Br J Cancer* 1992;**66** (**Suppl XVIII**):S41-7.