

## CONGENITAL MEDULLOBLASTOMA

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### SYNOPSIS

**Congenital medulloblastomas are very rare tumours and only 14 cases have been reported in the literature. A case report is presented on another case where hydrocephalus was present at the time of delivery resulting in dystocia. In a review of the literature, it is found that progressive head enlargement and hydrocephalus were the commonest modes of presentation. There is a high female incidence and familial cases have also been described. The importance of intra-uterine brain tumours as a cause of hydrocephalus is pointed out.**

### INTRODUCTION

Medulloblastoma accounts for about 20% of intracranial tumours in infancy and childhood, with a 2:1 male preponderance and a peak frequency between 3 and 8 years of age (Koos and Miller, 1971).

Congenital medulloblastoma, however, is a rarity. There were fourteen cases of medulloblastoma reported to occur within the first six months of life. Five of these cases had neurological symptoms and signs at birth.

The present report is on a case of congenital medulloblastoma presenting with hydrocephalus causing obstructed labour.

### CASE REPORT

C. K. K. a 26 year old gravida 2 para 1 Chinese at 40 weeks gestation was admitted to Kandang Kerbau Hospital in early labour. Her antenatal history was unremarkable except for a persistent non-engagement of presenting parts. The first pregnancy was uneventful and a normal female infant was born at term.

Physical examination confirmed a non-engagement of the presenting head. The cervix was effaced and the os was 4 cms. dilated. Artificial rupture of the membrane was done and moderately meconium stained liquor was drained. The head failed to descend into the pelvis. Radiological examination showed a hydrocephalic foetus (Fig. 1). The broadest

diameter of the head remained above the pelvic brim in spite of strong uterine contractions. A 3,070 gm. male infant was delivered after 250 ml. of blood stained cerebrospinal fluid was tapped from the hydrocephalic head. He died 2 hours later.

### Autopsy Findings

Postmortem examination showed a grossly hydrocephalic male infant. A fresh ragged perforation 2 cms. in diameter was noted at the vertex. The collapsed cranium measured about 18 cms. in diameter.

The cerebral hemispheres were haemorrhagic, collapsed and incomplete. There was obvious hydrocephalus as the remaining cerebral cortex was thin, ranging from 1 to 2 cms. in thickness. The entire brain weighed 444 gms. At the roof of the fourth ventricle, a grayish soft friable growth of 2×1 cm. in size was noted. The growth appeared to have destroyed the inferior vermis, the inferior medullary velum and the cerebellar tonsils. It extended into the fourth ventricle, growing along its roof both upwards and downwards.

No other abnormalities were noted in all the other systems.

Routine histological sections revealed a tumour consisting of compact sheets of dark round or oval cells with scanty cytoplasm (Fig. 2). Occasional pseudorosettes were present (Fig. 3). The tumour did not produce any significant amount of reticulin fibres.

A small piece of tumour tissue fixed in 10% formalin was further processed for electron microscopic examination. The tumour consisted of oval or polygonal cells arranged in compact sheets with little intercellular space. The cytoplasm contained a few mitochondria, some free and membrane-associated ribosomes and scanty endoplasmic reticulum (Fig. 4). Structures re-

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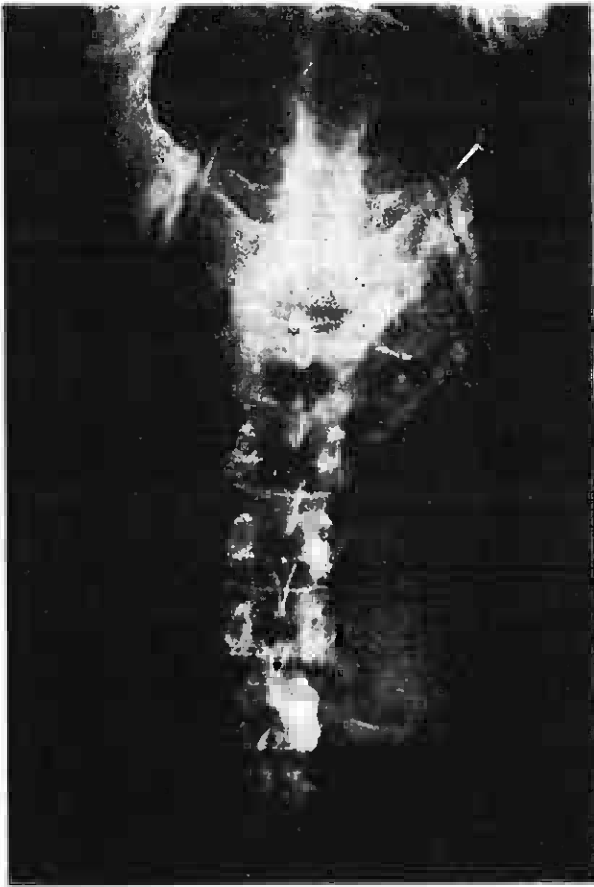


Fig. 1. Abdominal radiograph showing presence of grossly hydrocephalic foetus.

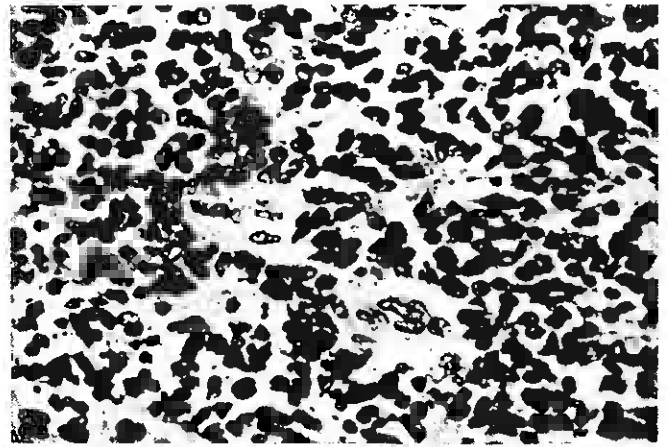


Fig. 3. Sheet of oval or round, closely packed tumour cells. Note a pseudorosette close to a blood vessel. H. & E.  $\times 500$ .

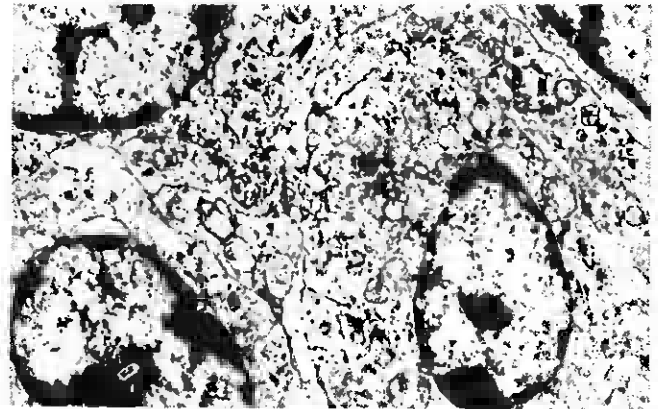


Fig. 4. Oval or polygonal cells with moderate amount of cytoplasm. The cells are closely packed with little extracellular space.  $\times 4500$ .

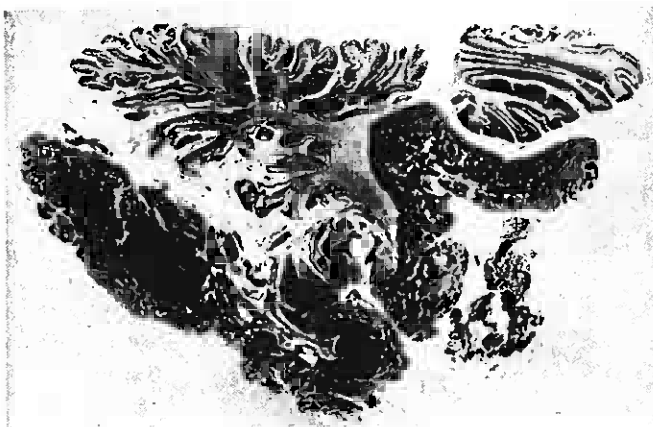


Fig. 2. Histological section through vermis showing sheets of densely packed tumour cells growing up the roof of the fourth ventricle.

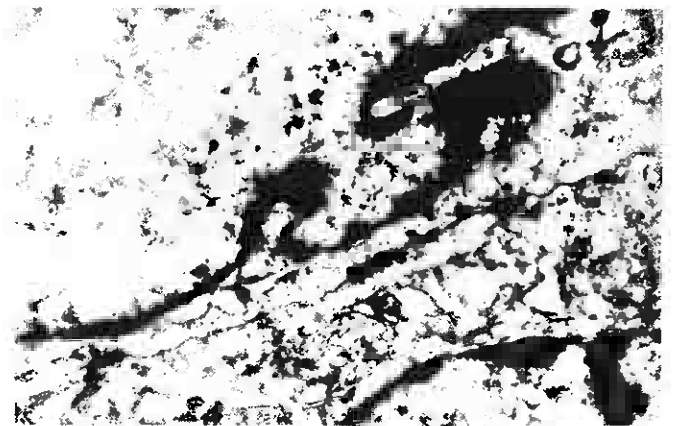


Fig. 5. Cell junction. Cytoplasmic plaques of electron dense material adjacent to opposing cell membranes.  $\times 9500$ .

TABLE I  
CONGENITAL MEDULLOBLASTOMA  
- Summary of reported cases

Case No.	Reference	Sex	Age at onset of signs	Age at Presentation	Presenting Features	Age at Death	Remarks
1.	Leibner (1948)	M	Birth	Birth	Head enlargement, fits	28 days	Presented with difficult instrumental delivery.
2.	Cuneo <i>et al</i> (1952)	F	1½ months	6 months	Right facial palsy, fits	6 months	
3.	King (1953)	Not Stated	2 weeks	3 months	Vomiting, head enlargement and hydrocephalus	4½ months	
4.	Griepentrog <i>et al</i> (1957)	F	Birth	Birth	Left clavicular tumour, Hemi-paresis	8 weeks	Autopsy findings: medulloblastoma in region of IVth ventricle and teratoma containing medulloblastomatous nodules pressing on the left brachial plexus.
5.	Griepentrog <i>et al</i> (1957)	F	Birth	Birth	Left facial paralysis, hydrocephalus	11 weeks	Cases 4 and 5 were identical twins.
6.	Fine (1962)	F	6 weeks	7 weeks	Head enlargement and hydrocephalus; projectile vomiting	7 months	—
7.	Russell <i>et al</i> (1963)	F	10 days	7 weeks	Head enlargement	Not stated	—
8.	Duckett <i>et al</i> (1966)	F	Birth	Birth	Multiple congenital anomalies	1 day	Pregnancy complicated by hydramnios. Of 4 previous pregnancies, 1st child died at 12 hrs., 2nd child died at 4 months with hydrocephalus. 3rd child was stillborn with hydrocephalus and 4th child normal. Present child had multiple abnormalities in abdominal wall and genito-urinary system.
9.	Takaku <i>et al</i> (1967)	F	Birth	26 days	Left facial paralysis and dysphagia	27 days	Tumour in left cerebellopontine angle, consisting histologically of elements of medulloblastoma and glioblastoma multiforme.
10.	Belamaric <i>et al</i> (1969)	F	15 days	33 days	Progressive head enlargement and hydrocephalus	68 days	Cases 10 and 11 were sisters. In this family, of the 5 other children, 4 males were healthy whilst the eldest daughter (5th child) had progressive hydrocephalus of unknown etiology and died at 4 months.
11.	Belamaric <i>et al</i> (1969)	F	Birth	Birth	Hydrocephalus	12 days	—
12.	Kadin <i>et al</i> (1970)	F	Not Stated	13 weeks	Lethargy, poor feeding, enlarging head and hydrocephalus	13 weeks	—
13.	Papadakis <i>et al</i> (1971)	M	3 weeks	4 weeks	Enlarging head and downward deviation of eyes, hydrocephalus	11 weeks	—
14.	Papadakis <i>et al</i> (1971)	F	Not Stated	3 months	Vomiting	11 months	—
15.	Present case	M	Birth	Birth	Hydrocephalus	2 hours	Presented with dystocia. Hydrocephalus diagnosed before delivery.

sembling tight junctional complexes were frequently identified between adjacent cells (Fig. 5).

## DISCUSSION

A summary of the clinical features of the 15 cases of congenital medulloblastoma is presented in Table I. Of the 14 cases where the sex of the infant was specified, there were 11 females and 3 males. This is in marked contrast to the overall male preponderance in medulloblastomas.

Most of the congenital medulloblastomas presented for medical attention within the first 3 months of life. Progressive head enlargement and hydrocephalus were the commonest modes of presentation (67%). Other modes of presentation included fits, vomiting and facial palsy.

The prognosis in all the cases was poor; the longest survivor died at 11 months old.

Familial occurrence of congenital medulloblastoma had been reported. The two cases described by Griepentrog and Pauly (1957) were identical twins. Belamaric and Chau (1969) described congenital medulloblastoma occurring in two sisters with one other girl in the family who died of hydrocephalus of unknown etiology at the age of 4 months. In the family of the case described by Duckett (1966), one sibling died at the age 4 months with hydrocephalus and there was a stillbirth with hydrocephalus. These reports of familial incidence indicate that there may be a genetic factor in the occurrence of congenital medulloblastoma. With reference to medulloblastomas occurring in older children, Kjellin (1960) reported an instance of a boy aged 10 years and his half-sister aged 11 years both developing cerebellar medulloblastoma.

In the present case, there is a congenital midline cerebellar tumour with classical histological and electron microscopical pictures of medulloblastoma. The site of this tumour is consistent with the postulation that medulloblastoma arises from germinative cells originating in the neuroepithelial roof of the fourth ventricle (Rubinstein, 1972).

The present case is the first case of congenital medulloblastoma where a diagnosis of hydrocephalus was made prior to delivery. Congenital

brain tumours though relatively rare should be considered in the differential diagnosis of congenital hydrocephalus.

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