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CASE REPORT

Congenital glioblastoma multiforme presented with intracranial bleeding: a case report

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Introduction

Congenital brain tumours constitute 0.5–4% of all paediatric brain tumours (Manoranjan and Provias 2011). Although they are not easily diagnosed before birth, the detection rate has been increasing due to prenatal screening tests. The congenital brain tumour incidence is 1.1–3.6 in every 100,000 live births, consisting of teratomas, astrocytomas, primitive neuro-ectodermal tumours, choroid plexus papillomas and glioblastoma multiforme (GBM) (Shimamura et al. 2003). Congenital GBMs are the rarest of the congenital brain tumours (Brown et al. 1997). Infants with these tumours may be stillborn or exhibit a tendency towards bleeding and haemorrhage in their first week of life (Manish et al. 2007). The majority of congenital GBMs are located in the cerebral hemispheres, cerebellopontine angle and the cerebellar hemispheres (Nakayama and Nakamura 2002). During the postnatal period, these patients may present with macrocrania, intracranial pressure symptoms, hydrocephaly and haemorrhage (Podskalny et al. 1993).

Case report

A 22-year-old gravida 2, parity 1 female was referred to our clinic because it was suspected that her foetus had a tumour with intracranial bleeding in an ultrasound examination. There were no risk factors in her medical history, and no pathological findings were recorded in her ultrasound examinations. The 37th gestational week’s ultrasound revealed the findings; the biparietal diameter was 107 mm, head circumference was 374 mm (>97% for gestational age), there was a 9.8 cm sized tumour supratentorially in the right cerebral hemisphere, causing shifting and herniation of the midline structures (the internal blood supply illustrated via Doppler ultrasonography, Figure 1(A)). A male weighing 2295 g was delivered via caesarean section at 37 weeks of gestational age. The 1 and 5 minute Apgar scores were 6 and 8, respectively, and the head circumference was 39 cm. Macrocephaly and bilateral setting-sun eye phenomena were observed. He was extremely floppy, with no active movements. The cranial magnetic resonance imaging (MRI) revealed a 9.8 cm tumour with a haemorrhage located in the right cerebral hemisphere (Figure 1(B)), shifting to the left of the midline, with a subfalcine herniation (Figure 1(C)). Unfortunately, this patient died while undergoing surgery on the second postnatal day. The pathological examination revealed a grade IV, isocitrate dehydrogenase-1 (IDH1) negative glioblastoma. His parents refused an autopsy.

Discussion

Brain tumours are extremely rare in infants younger than six months old. GBMs constitute 2–9% of all congenital brain tumours (Shimamura et al. 2003), and their locations, histology and treatment responses are different to paediatric patients. Neonatal intracranial tumours have been divided into three groups by Solitaire and Krigman (1964), which are as follows: definitely congenital with symptoms present at birth, likely congenital with symptoms manifested within the first week, and presumably congenital with symptoms manifested within the first two months. Our case falls into the first group. Most GBMs are located in the supratentorial area, as in our case. In one recent study of 488 children with central nervous system neoplasms, 43 (8.8%) had GBMs, of which 22 (51.1%) were in the cerebral hemispheres, two were in the cerebellum, and three were in the spinal cord (Farwell et al. 1977). In our case, the tumour was located in the right cerebral hemisphere.

Foetal MRI and prenatal ultrasounds have increased the congenital GBM diagnosis rate (Kasliwal et al. 2007). As the ultrasound showed an increased vascularity, it was difficult to differentiate a tumour from an intracranial haemorrhage in our case. However, the diagnosis was confirmed by MRI in the postnatal period.

The symptoms of GBMs are related to an increased intracranial pressure; for example, macrocephaly, hydrocephaly and setting-sun eye phenomena were observed in our case. The mean life expectancy is two years. According to a review (Anestis et al. 2017), the stillborn percentage was found in 29% on the first day, 38% in the first week and 56% in the first two months, which had the highest mortality as in our case.

Neonates should be monitored during surgery because even a small blood loss may cause hemodynamic instability.
Postoperative complications such as a massive intracranial haemorrhage may also occur. Our patient died from hypovolaemia, which developed secondary to a massive haemorrhage during the surgery. Adjuvant chemoradiation forms the modality of treatment choice for GBM. Nevertheless, a younger age and lack of guidelines prevent the use of adjuvant radiotherapy.

The World Health Organization classifies central nervous system tumours according to the histological characteristics. Wild-type IDH1 is responsible for 90% of all cases, and it is considered to be a de novo GBM. IDH1 mutations are common in grades II and III astrocytomas and in secondary GBMs, establishing a link with the adult patient survival rates (Louis et al. 2016). Our patient’s prognosis was poor because the tumour was IDH1 negative.

Congenital brain tumours should be considered in the differential diagnosis of an intracranial haemorrhage. Moreover, foetal MRI is required for the diagnosis and treatment planning. The onset of most of the congenital brain tumours occurs in the third trimester, and they cannot be diagnosed in the earlier weeks.

**Disclosure statement**

No potential conflict of interest was reported by the authors.

**References**


