Fetal and Pediatric Pathology

ISSN: 1551-3815 (Print) 1551-3823 (Online) Journal homepage: www.tandfonline.com/journals/ipdp20

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To cite this article: Emine Turkmen, Jack Raisanen, Metin Dogan, Ayse Sandikkaya, Derya Gumus Dogan, Nurhan Sahin & Ahmet Karadag (2011) A Newborn with Massive Congenital Astroblastoma, Fetal and Pediatric Pathology, 30:5, 325-328, DOI: 10.3109/15513815.2011.564718

To link to this article: https://doi.org/10.3109/15513815.2011.564718

Published online: 14 Apr 2011.

Fetal and Pediatric Pathology, 30:325–328, 2011

ISSN: 1551-3815 print / 1551-3823 online
DOI: 10.3109/15513815.2011.564718

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Letter to the Editor

A Newborn with Massive Congenital Astroblastoma

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Astroblastoma is a rare glial tumor that occurs most frequently in children, adolescents, and young adults [1]. Congenital examples are rare with only a few reports in the literature [2–4]. We present a 1-day-old girl with a massive astroblastoma in the right cerebral hemisphere. At 33 weeks gestational age, an ultrasound revealed macrocephaly and the baby was delivered by cesarean section at 35 weeks. She was 2950 g (25–50 centil) with ahead circumference of 39 cm (97 centil) and soon developed respiratory distress. She was tachypneic (rate of 72/min) and dyspneic and had diffuse crackles in both lungs. Apgar scores were 6 and 8 at 1 and 5 min, respectively. Nasal continuous positive airway pressure was applied.

A post-natal ultrasound, CT (Figure 1), and MRI showed a 110 × 72 × 65 mm (by MRI) right cerebral hemispheric tumor causing a significant mass effect. The tumor contained cystic/necrotic areas and exhibited contrast enhancement. On day two, the baby required mechanical ventilation. On day five, she underwent resection but, postoperatively, suffered cardiorespiratory arrest and died.

Gross examination showed a soft, brownish-gray, hemorrhagic tumor. Hematoxylin and eosin-stained sections showed the cytologic and architectural features of astroblastoma. In most areas, the tumor was pseudopapillary with cells with relatively short, stout, unipolar cytoplasmic processes radiating around vessel walls (Figure 2). Mitotic figures were rare. Tumor cells were strongly stained immunohistochemically with glial fibrillary acidic protein (GFAP) (Figure 3), vimentin and S100 protein. The stains failed to demonstrate expression of high and low molecular weight cytokeratins (LMWCK

Note: Although some features of astroblastoma may resemble those of ependymoma, distinguishing features are GFAP immunoreactivity pattern, broad-based cytoplasmic footplate directly on basal lamina of the capillary. Ependymomas often have interspersed GFAP reactivity with stronger reactivity in the ependymal glial filaments near the central vascular core. Astroblastomas have diffuse GFAP reactivity. Reticulin, phosphotungstic acid-hematoxylin (PTAH), laminin, and GFAP may be useful as adjunct studies. Astrocytomas, especially anaplastic astrocytomas and glioblastoma multiforme, may focally have an "astroblastic pattern," but the term astroblastoma is reserved for tumors that have the astroblastoma morphologic pattern throughout (courtesy of Dr. Kondi Wong).

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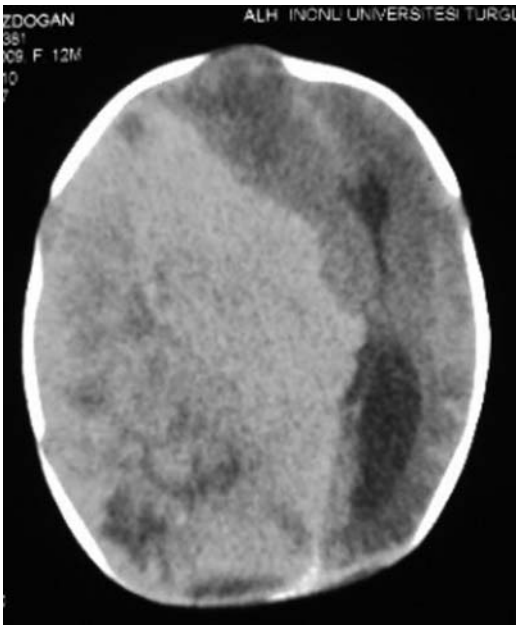

FIGURE 1 Head CT showing a very large tumor in the right cerebral hemisphere with significant mass effect.

and HMWCK), synaptophysin and neuroflament. Neuron specific enolase (NSE), and epithelial membrane antigen (EMA) stained tumor cells focally.

Astroblastoma (Figures 4A–4D) is a rare central nervous system (CNS) neoplasm accounting for 0.4–2.8% of gliomas [2, 5]. Since its original description by Bailey and Bucy in 1930 [6], both the histogenesis of astroblastoma and its validity as a distinct entity have been controversial. The finding of expression of GFAP and S100 without definite evidence of neuronal, or in most cases, ependymal differentiation has suggested astrocytic derivation. Ultrastructural observations have raised

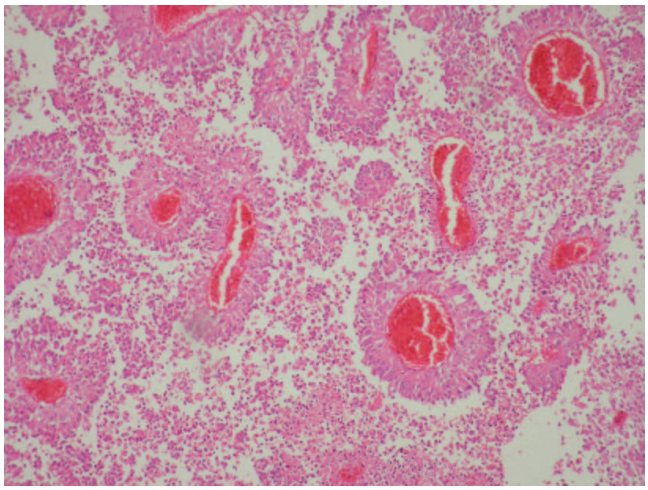

FIGURE 2 The tumor was composed of pseudopapillae with glial cells with relatively short, broad-based processes arranged around thin-walled vessels (H&E × 200).

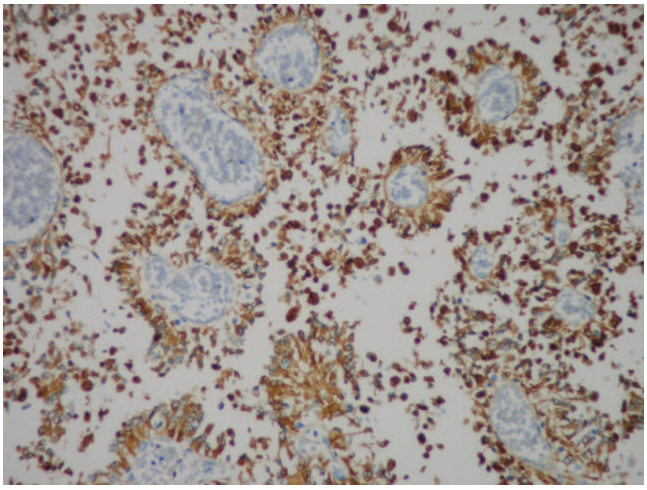

FIGURE 3 GFAP expression of the tumor cells (Immunperoxidase × 200).

the possibility of tanycytic differentiation [1]. Because of its rarity, the World Health Organization has considered it premature to establish tumor grade, though well-differentiated and anaplastic examples may be distinguished by cytologic atypia, mitotic figures, microvascular proliferation, and pseudopalisading necrosis [1, 7].

In their review of 225 CNS tumors that were present at birth, Raisanen and Davis [8] found no astroblastomas. Since then, only four newborn have been reported [2–4, 9]. Although there are few reports, congenital astroblastomas appear to have clinicopathologic features similar to other fetal CNS tumors. They are usually supratentorial and often massive with large head or tense fontanel as the presenting sign.

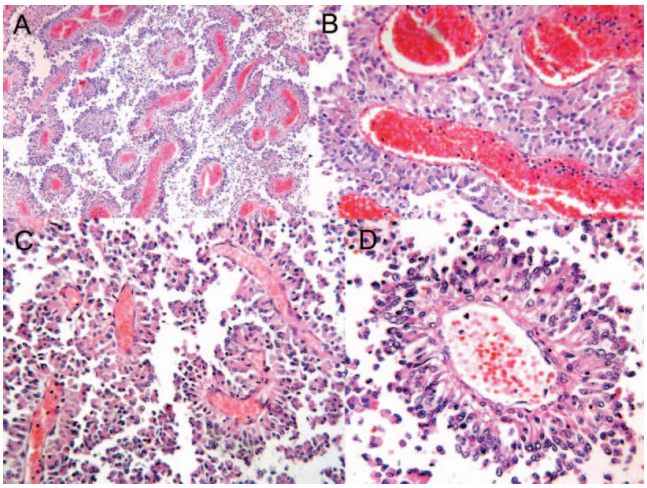

FIGURE 4 **A,** Perivascular, papillary rosette pattern pattern on low power (H&E, ×2 magnification lens). **B,** Astroblaastoma tumor cells layer directly upon capillary. Note dilated capillary lumen. (H&E ×100 magnification). **C,** Each astroblastoma tumor cell has a broad cytoplasmic footplate that broadly contacts the basal lamina of the capillary vessels. A few endothelial nuclei are somewhat plump, but most are flattened (H&E × 200 magnification). **D,** Perivascular papillary rosette with central dilated capillary lumen, astroblastoma tumor cell nuclei extending in a radial array away from the lumen (H&E × 200 magnification).

Prognosis is dismal and fetuses are often stillborn. Indeed, in descriptions of 225 congenital brain tumors, only 25 patients were alive at the time of the reports [8].

Declaration of Interest

The authors have no conflict of interest. The authors alone are responsible for the content and writing of this article.

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