A Unique Case of Cerebellar Choroid Plexus Carcinoma

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A 3-year-old male, complaining of 4-month headache and vomiting, was evaluated at Children's Hospital 2. His medical profile showed no abnormalities. During clinical evaluation, no signs of neurological deficits were identified, and routine blood tests were within normal limits. Brain magnetic resonance imaging, with a contrast agent, revealed the absence of hydrocephalus and supratentorial lesions. A heterogeneous-signal-intensity mass ($48 \times 41 \times 38$ mm^3), with surrounding edematous parenchyma, was identified in the right cerebellar hemisphere, on the T1-weight image (Figure 1A), T2-weighted image (Figure 1B), and the fluid-attenuated inversion recovery image (FLAIR) (Figure 1C). Hemosiderin deposition was identified inside the mass, from old hemorrhage. The mean apparent diffusion coefficient (ADC) values for the parenchyma and the solid lesion component were 0.63 and 0.86×10^{-3} mm^2/s, respectively (Figure 1D). The relative enhancement (%), peak enhancement, peak relative enhancement (%), time to peak (s), wash-in rate (s^{-1}), wash-out rate (s^{-1}), and area under the curve values of the parenchyma compared with the solid mass component, as calculated from the T1-perfusion map, were 1.76 vs 34.64, 37.21 vs 153.26, 2.21 vs 7.36, 43.68 vs 168.47, 12.25 vs 24.00, 5.98 vs 3.14, and 392.47 vs 3632.18, respectively (Figure 1E and F). The preliminary diagnosis was medulloblastoma, and the patient underwent radical tumor excision. The histopathological assessment of the excised tissues revealed a choroid plexus carcinoma (CPC) (Figure 2A and B). The patient was discharged after 2 weeks and continued to receive adjuvant chemotherapy at a different oncological hospital.

According to the World Health Organization Classification, CPC is a particularly malignant, type III tumor that represents fewer than 1% of all intracranial tumors. CPC originates from the choroid plexus epithelium (CPE) and is most frequently found in the ventricular system, particularly the lateral ventricles (50%) and the fourth ventricle (40%), although less common in the third ventricle (5%).[1] CPC can occur in all ages of patients but is commonly diagnosed in children.[1] Extraventricular, intraparenchymal CPC is exceedingly rare. To the best of our knowledge, only two prior reports have described an intraparenchymal CPC.[2,3] Carter et al.[2] reported a 6-year-old female patient with an intraparenchymal CPC situated in the left frontal lobe, and Stevens et al.[3] presented a 6-year-old female patient with an intraparenchymal CPC located in the right frontal lobe. Thus, our case represents the third report of an intraparenchymal CPC but may be the first reported case of infratentorial, intraparenchymal CPC. Our case was initially misdiagnosed as medulloblastoma or

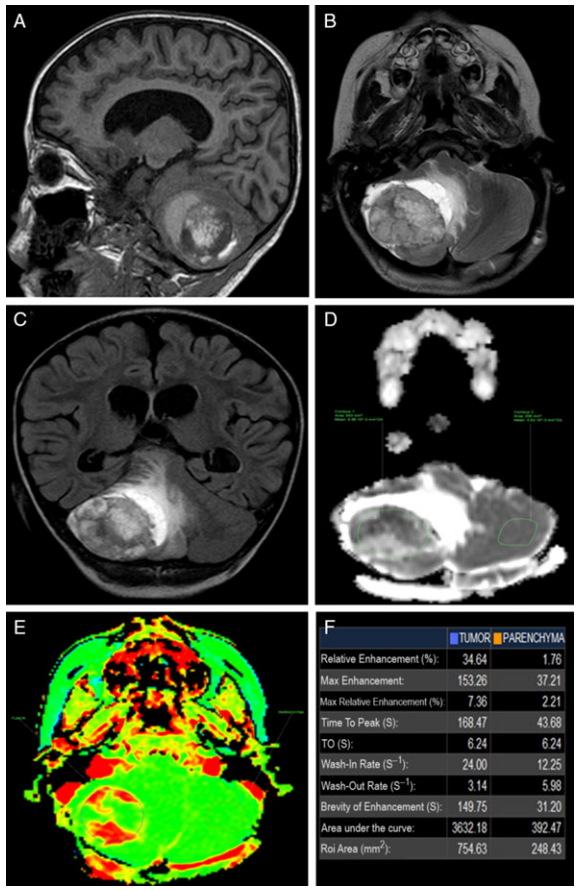

Figure 1: *A mass, located in the right cerebellar hemisphere, was heterogeneous signal intensity on the sagittal T1-weighted image (A), axial T2-weighted image (B), and coronal FLAIR image (C). ADC map of the lesion and the normal-appearing parenchyma (D). T1-perfusion map of the normal-appearing parenchyma and the lesion (E). The detailed perfusion parameters of the normal-appearing parenchyma and the lesion (F). Roi, region of interest.*

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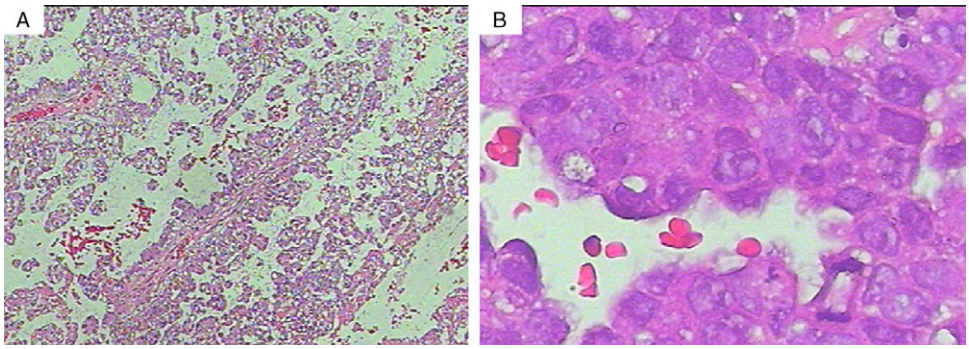

Figure 2: *Photomicrographs, showing typical CPE, with nuclear pleomorphism and high mitotic activity, accompanied by some necrotic areas (hematoxylin and eosin staining); original magnification (A) and × 400 (B).*

atypical teratoid rhabdoid tumor (grade IV), a very common pediatric, malignant tumor of the cerebellum,[3,4] due to the atypical position of the CPC, which was situated in the right cerebellar hemisphere.

Two theories have been suggested to explain CPC development in unusual locations. First, CPC may extend from primitive ectopic CPE within the brain parenchyma, beyond the ventricular system. Second, CPC may develop from migrating CPE that becomes isolated during brain development.[5] Because most extraventricular intraparenchymal CPCs are adjacent to the ventricular region, CPC may originate from the proximate CPE, and become separated during development.[2,3,5,6]

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Statement of Ethics

This study was approved by the Institutional review board of Children's Hospital 2 (Ref: 352/ND2-CDT). Written informed consent from the patient's legal guardian was obtained for the publication of this case report and any accompanying images.

Statement of Authorship

LTD and NMD contributed equally to this work as co-first authors.

Conflict of Interest

The authors have no conflicts of interest to declare.

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