Atypical central neurocytoma with aggressive features in a child

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DESCRIPTION

A 12-year-old girl presented with 3 months of headaches, blurred vision and progressive vomiting. Neurological examination revealed bilateral papilloedema and mild difficulties with tandem gait. MRI demonstrated a 4 cm mass within the right lateral ventricle that showed reduced diffusivity on diffusion-weighted sequences, and heterogeneous enhancement (figure 1). Magnetic resonance spectroscopy showed an elevated choline to creatinine ratio. The patient underwent gross total resection where neuropathology revealed a moderately cellular proliferation of monotonous polygonal cells with generally round nuclei, 'salt and pepper' chromatin and a modest amount of lightly eosinophilic cytoplasm and two mitotic figures per high powered field consistent with a diagnosis of atypical neurocytoma (figure 2). A DNA-based next generation sequencing panel consisting of 397 cancer-related genes performed on paraffin-embedded formalin fixed tumour demonstrated variants of unknown significance in seven genes (*DICER1*, *DOT1L*, *GATA6*, *NKX2-1*, *NOTCH2*, *PDGFA*, *TNFAIP3*).

Postoperatively, the patient was found to have left-side haemiplegia that slowly improved. A follow-up MRI performed 1-year postsurgery showed concern for progressive disease that was confirmed on additional follow-up MRI 2 years

Figure 2 H&E-stained section of initial tumour at 200× (A) and 400× (B) magnification demonstrates revealed a moderately cellular proliferation of monotonous polygonal cells with generally round nuclei, 'salt and pepper' chromatin and a modest amount of lightly eosinophilic cytoplasm and two mitotic figures per high powered field consistent with a diagnosis of atypical neurocytoma.

Learning points

- ▶ Central neurocytoma is an extremely rare paediatric tumour with both intraventricular and extraventricular presentations.
- ▶ Central neurocytoma is commonly associated with a favourable prognosis; however, atypical features including elevated mitotic index may be associated with worsened survival and require adjuvant therapy following surgery.
- ▶ The molecular features of central neurocytoma have not been fully elucidated highlighting the complexity of this rare tumour.

Figure 1 MRI reveals a ventricular tumour with reduced diffusivity on diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) sequences (A, B) and punctate areas of mineralisation on susceptibility-weighted images (C). The tumour showed avid enhancement on post-T1 gadolinium weighted sequences (D) with associated hydrocephalus (E). Magnetic resonance spectroscopy showed elevation of choline to creatinine ratio consistent with a higher-grade malignancy (F).

postsurgery, prompting near total resection of the mass. The neuropathology revealed a similar mitotic index as the original tumour and similar histological features. Given the rapid recurrence of tumour the patient underwent focal proton radiation therapy (5400cG whole ventricular with a boost to the tumour bed totalling 5940cG) following the second resection and has remained in remission for 5 years.

Central neurocytoma is an extremely rare paediatric brain tumour classified by WHO classification as a grade 2 neuronal or mixed neuronal-glial tumour commonly located near the foramen of Monro and lateral ventricles.^{1,2} In general, neurocytoma is associated with a favourable prognosis

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in cases of gross total resection; however, adjuvant therapy to include chemotherapy or radiation therapy may be required in cases of recurrence or progression. Central neurocytoma with a MIB-1 of greater than 2%–4% has been associated with poorer outcome.^{3,4} Extraventricular locations may occur in children and are generally associated with younger age, seizures at presentation and worsened overall survival.⁴ The molecular understanding of central neurocytoma is less understood than other childhood central nervous system tumours. In summary, central neurocytoma is a rare paediatric brain tumour with ill-defined molecular genetics and may exhibit more aggressive features that require adjuvant therapy following surgery.

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