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, Nfk2-1, Notch2, Pdgfra, Tnfaip3).

Postoperatively, the patient was found to have left-side hemiplegia 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 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 (5400cGE whole ventricular with a boost to the tumour bed totalling 5940cGE) following the second resection and has remained in remission for 5 years.

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.

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).

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.

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. Extraventricular locations may occur in children and are generally associated with younger age, seizures at presentation and worsened overall survival. 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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**REFERENCES**