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Neuroimaging Highlight

Metachronous Brain Tumors: Supratentorial Ependymoma Following Polymorphous Low-Grade Neuroepithelial Tumor of the Young

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A previously healthy 41-year-old man with a first generalized tonic-clonic seizure underwent a head CT that disclosed a peripherally located, partially calcified mass in the right frontal lobe (Fig. 1a). Neurological examination was unremarkable. MRI showed a T1-mixed intensity and T2-hyperintense mass with central cystic changes, mild heterogeneous enhancement, no significant perilesional edema and no restricted diffusion (Fig. 1b–f). Gross total resection and pathological examination confirmed a polymorphous low-grade neuroepithelial tumor of the young (PLNTY). Postoperative seizures were well-controlled on antiepileptic drugs, and there was no evidence of recurrence on MR surveillance. However, 11-months post-resection, he presented with worsening headaches and right facial weakness. Repeat MRI revealed a new T1-hypointense and T2-hyperintense heterogeneously enhancing mass in the left frontal centrum semiovale with different MR imaging characteristics from the original tumor, including mild restricted diffusion, internal necrosis and minimal edema (Fig. 2a–e). MR spectroscopy demonstrated increased choline and decreased NAA peaks within the lesion (Fig. 2f) suggesting high cellular turnover. Biopsy confirmed a *ZFTA::RELA* fusion-positive anaplastic ependymoma. Despite receiving radiation treatment and two resections, the patient ultimately passed away.

PLNTY is a rare tumor first described in 2017 that frequently presents with seizures and refractory epilepsy.[1] While most common in pediatric populations, cases have been reported in adults.[2] Imaging findings include intralesional calcifications, cystic components and well-defined margins. MRI signal characteristics include T1-iso/hypointensity and T2-iso/hyperintensity, mild heterogeneous enhancement, minimal perilesional edema and no diffusion restriction.[3] The differential diagnosis includes oligodendroglioma due to imaging similarities and calcification, and ganglioglioma due to its epileptogenic nature.[2,4] Histopathology and molecular analysis are required to diagnose PLNTY, with characteristic CD34 immunoreactivity and distinct genetic mutations.[5] Standard

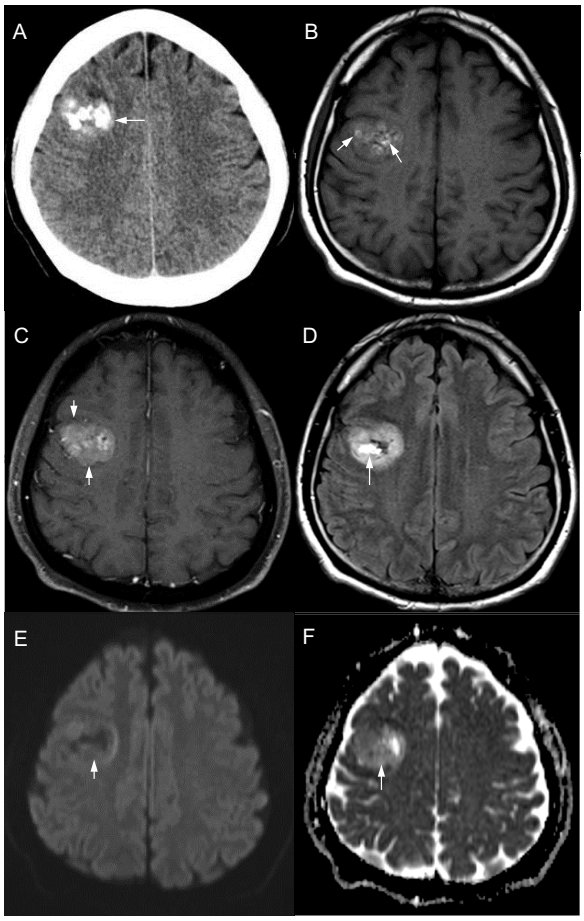

Figure 1. Right frontal PLNTY. Nonenhanced brain CT demonstrates a right frontal partially calcified mass (A, arrow). MRI confirms the mass which is predominantly t1 hypointense with t1-hyperintense calcifications (B, arrows) and mild gadolinium-contrast enhancement (C, arrows). FLAIR images show internal cystic changes (D, arrow) without perilesional edema. Diffusion-weighted imaging (E) and corresponding ADC map (F) reveal no diffusion restriction within the lesion (arrows).

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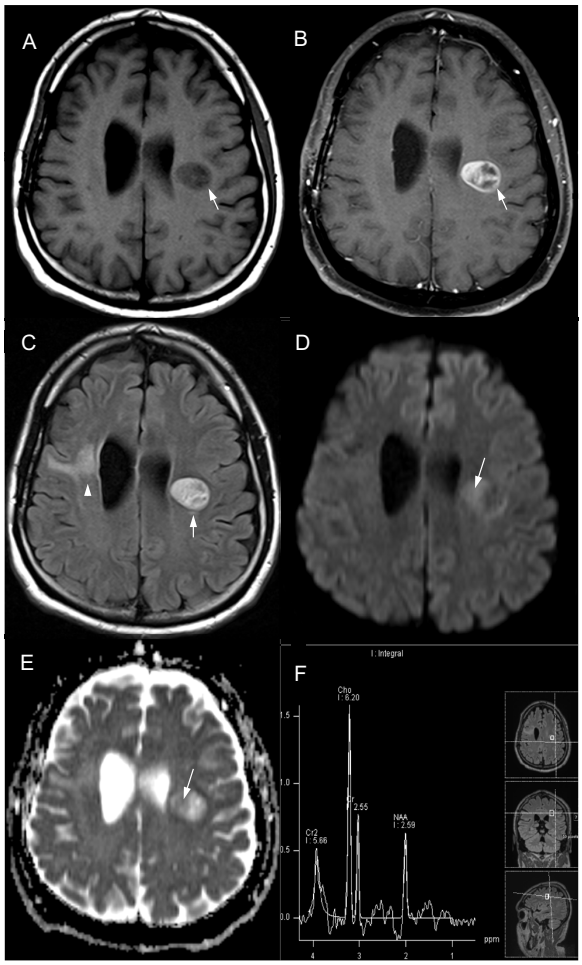

Figure 2. Left frontal supratentorial ZFTA fusion-positive ependymoma. MRI with T1-weighted imaging reveals a hypointense left frontal mass on the pre-contrast acquisition (A, arrow), that has nodular enhancement and internal necrosis post-gadolinium administration (B, arrow). FLAIR imaging shows minimal surrounding edema (C, arrow), and expected postoperative/gliotic changes at the right resection site (C, arrowhead). Diffusion-weighted imaging (D) and ADC map (E) demonstrate mild heterogenous restricted diffusion (arrows). MR spectroscopy shows an elevated choline peak and decreased NAA peak (F).

treatment is gross total resection, and recurrence is due to incomplete surgical margins.[2]

Supratentorial ependymomas (STE) are most common in the adolescent population and have varied presentations including headaches, seizures and focal neurologic deficits.[6] Molecular analysis has classified STE into two main categories, RELA and YAP1, according to fusion gene products, with the RELA subtype having worse outcomes.[5,7] Typical imaging findings include central calcification, large cystic components and central necrosis, with high-grade features such as moderate heterogeneous enhancement, perilesional edema with mass effect, abnormal spectroscopy and diffusion restriction. MR signals are T1-iso/hypointense and T2-iso/hyperintense.[8] The differential diagnosis includes oligodendroglioma due to internal calcifications and diffuse astrocytoma due to the presence of cystic components.[8] Resection and radiotherapy are mainstays of treatment.[9]

Multiple different primary brain tumors are often caused by prior radiation exposure or a genetic predisposition, but sporadic cases have been reported.[10] These most frequently involve meningiomas, gliomas or pituitary adenomas; can present simultaneously (synchronous) or sequentially (metachronous); and are often in separate brain regions.[10]

Our report of metachronous PLNTY and STE is the first to our knowledge. PLNTY and STE are more prevalent in pediatric populations but remain on the differential for primary CNS tumors in adults. Molecular analysis is necessary for accurate diagnosis. Though rare, our findings raise awareness for these entities; two metachronous primary brain tumors raise the possibility of an underlying cause. Sporadic cases remain possible but highly unusual.

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