Dysembryoplastic neuroepithelial tumor

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The authors have succinctly described the course of a 15-year-old boy with dysembryoplastic neuroepithelial tumor (DNET) occupying the corpus callosum and pericallosal region. They have successfully kept the patient symptom free and in remission by surgery alone. Most DNETs are situated in the brain parenchyma, with superficial cortical regions being the most common location. Occasionally, DNETs may be quite aggressive and large, and can extend from the cortex to the white matter, and then to the deep nuclei such as the basal ganglia. Very rarely, malignant features may be seen within these tumors. After 3 years, the patient is doing well without neurological issues. Although the imaging features were suggestive of DNET, the location was not. The authors have done well to draw our attention to atypically presenting DNETs, and to underscore the role of surgery in these cases.

Reference

Response

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We appreciate Dr. Rutka’s interest in our work and his thoughtful comments. As Dr. Rutka emphasized, DNETs are most frequently located in cortical surfaces but also can arise in various locations including basal ganglia, cerebellum, and even pons. The lesion in our case was uniquely located in the pericallosal region, with intraventricular and septal extensions. Dysembryoplastic neuroepithelial tumors are usually diagnosed in young patients, with medically intractable seizures being their only clinical manifestation, and they are accepted as benign lesions. These lesions can be followed radiologically if the patient is asymptomatic or seizures are well controlled. We operated on our patient mainly because the location was very atypical and tissue diagnosis was critical for the patient’s further management plan. It was imperative in this case that an exact pathological diagnosis be established so that potentially harmful adjunctive treatment modalities, such as radiation therapy, could be avoided. After obtaining a histological diagnosis we followed our patient closely with MR images because the lesion was resected subtotally. His MR imaging studies showed no tumor progression within 4 years. However, DNET is still a relatively new tumor in neurooncology; it was first described in 1988. Therefore, the data regarding its natural and postoperative course are still limited, and there is 1 reported case of malignant transformation of the lesion 11 years later. It is our belief that long-term, close radiological follow-up is still critical in the management of the disease in patients with DNET. (DOI: 10.3171/2009.2.PEDS0927)

References