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CORRESPONDENCE

Pediatric primary lymphoma of the pituitary stalk: A different disease entity from the adult form?

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Pediatric primary pituitary lymphoma (PPL) is a rare and challenging disease entity. We report the case of a 7-year-old girl who presented with central diabetes insipidus (DI). Brain magnetic resonance imaging (MRI) showed diffuse thickening of the pituitary stalk with extension to the hypothalamic region. A small, nodular, heterogeneous enhancement of the pineal gland was also noted (Fig. 1A). A combined anterior pituitary function test and clonidine test showed a subnormal growth hormone response although her body height and body weight were between the 50th and 75th percentiles. A peripheral blood examination and serum α -fetoprotein (α -FP) and β human chorionic gonadotropin (β -hCG) levels were within normal limits. Progressive enlargement of the lesion on follow-up imaging and the development of pan-hypopituitarism prompted a transcranial biopsy, which revealed a diffuse large B-cell lymphoma. She received chemotherapy and showed a complete response. At the time of writing, the patient was doing well with persistent pan-hypopituitarism. Follow-up MRI 16 months after surgery showed complete disappearance of the tumor (Fig. 1B).

In this patient, the initial tentative diagnosis was germinoma or Langerhans cell histiocytosis (LCH). LCH occurs

more often in children and usually involves multiple organs but may be limited to the central nervous system (CNS). The hypothalamus–pituitary region is the principal site for CNS involvement, and patients usually present with DI and other neuroendocrine abnormalities. Germ cell tumors, more common in Asian children, can involve the pituitary stalk, hypothalamus, basal ganglia, and the third ventricle. Despite the absence of the production of either α -FP or β -hCG, this is still a differential diagnosis in this case, especially considering the coexisting pineal lesion. The age of this patient would go against the diagnosis of lymphocytic hypophysitis used to describe cases of central DI in adults with a thickened pituitary stalk, which often occurs in late pregnancy or the postpartum period.

PPL is a rare disease entity and represents less than 1% of sellar masses. Even though the number of PPL cases has increased during the past decade, only 22 cases, including the present one, have been reported in the literature and only four of these patients were younger than 18 years.^{1–5} A review of the literature revealed that the clinical presentation, tumor size and location, course of the disease, histology and coexisting lesions differ between pediatric and adult PPLs. Pediatric patients with PPL tend to have a smaller tumor size, mainly located in the pituitary stalk, although pineal involvement may be observed in half of the patients. Most of the lesions progress slowly over a period of years before a definite diagnosis is made. The histology showed B-cell lymphoma without a coexisting

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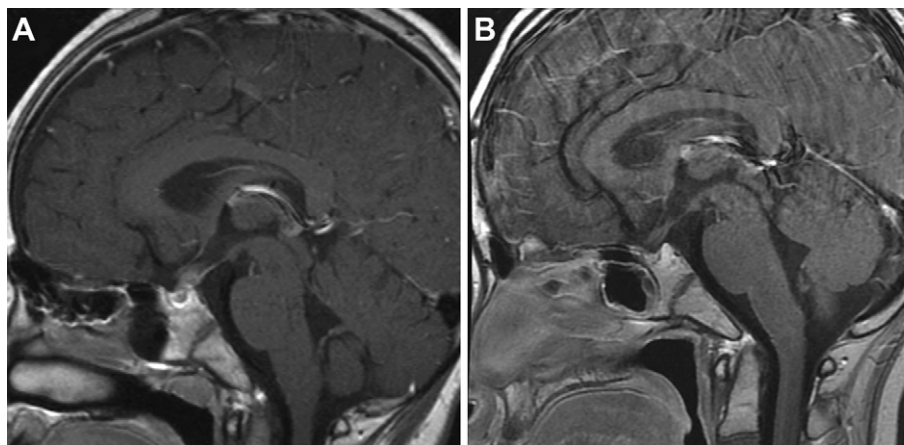


Figure 1 Sagittal T1-weighted magnetic resonance imaging with gadolinium injection. (A) Initial image showing pituitary stalk thickening (6 mm thick) with extension to the hypothalamic region, the floor of the third ventricle and loss of the physiological high signal intensity of the posterior lobe on T1-weighted imaging. A small nodular, heterogeneous enhancement of the pineal gland was also noted. (B) Follow-up image 16 months after surgery and chemotherapy showing complete disappearance of the tumor.

lesion or condition such as pituitary adenoma, lymphocytic hypophysitis, and immunodeficiency.

Although the mechanism of pathogenesis of PPL is still unknown, the data suggest that PPL in pediatric patients may be a different disease entity from that in adults. In the near future, detailed molecular and genetic studies of these rare tumors will provide insight into their pathogenesis and the apparent clinical differences observed between adult and pediatric cases.

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