Ectopic atypical lymphoid tissue of the infundibulum/hypothalamus

Case illustration

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This 38-year-old woman was diagnosed with a pituitary stalk lesion when she presented with diabetes insipidus and primary amenorrhea as a teenager. She has since been on replacement hormones, and the lesion has been monitored with serial MR imaging. She had a negative workup for granulomatous disease (including sarcoidosis and tuberculosis), and lumbar puncture protein and cell count were unremarkable. Over time the lesion exhibited little propensity for growth and even regressed at one point. Imaging between 2006 and 2008, however, demonstrated noticeable growth (Fig. 1). Because of the unknown cause of the lesion and its growth on serial imaging, we performed a transcranial pterional biopsy. The mass densely adhered to the hypothalamus and was subtotally resected. Pathological analysis revealed lymphoid aggregates composed mainly of T cells and rare B cells, representing atypical lymphoid aggregate (Fig. 2).

This entity has not been described; it appears to be distinct from lymphocytic infundibuloneurohypophysitis and is thought to be distinct from lymphocytic adenohypophysitis,1,2 which primarily occurs in young women and is generally thought to be a self-limited process. Lymphocytic infundibuloneurohypophysitis rarely affects the optic pathways and hypothalamus.1 Typically, it exhibits significant plasma cells. In contrast, our lesion had no inflammatory or lymphocytic response but rather atypical lymphoid aggregate. Additionally, this aggregate primarily comprised small lymphocytic cells without significant plasma cells. A neuropathologist and hematopathologists concurred with the diagnosis. The histological and immunohistochemical findings support a benign lymphoid process. Further testing, including T-cell gene rearrangement and other lymphocytic markers, was deemed unnecessary for definitive diagnosis. There was no histological evidence of germinoma or histiocytosis. This is the first known case of such an entity with the propensity to demonstrate noticeable size reduction and growth in over 20 years of follow-up.

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References


Fig. 1. Coronal MR image obtained in 2008 showing the mass enlarged to 12.6 × 10.6 × 7.9 mm.

Fig. 2. A: High-magnification photomicrograph of a representative area showing a diffuse infiltrate of benign lymphocytic cells. H & E, original magnification × 100. B: The lymphocytic cell infiltrate is predominantly composed of T cells. Immunoperoxidase with anti-CD3, original magnification × 100. C: A minority of the lymphocytic cells are B cells. Immunoperoxidase stain with anti-CD20, original magnification × 100.

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