Idiopathic Granulomatous Hypophysitis Presenting as Pituitary Apoplexy

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Abstract

Introduction: Granulomatous hypophysitis is an inflammatory disorder of the pituitary gland characterized by the formation of intrasellar giant-cell granuloma. In the absence of underlying systemic disease, such as sarcoidosis, tuberculosis, or syphilis, it is classified as idiopathic. Idiopathic granulomatous hypophysitis (IGH) is an extremely rare disorder with few cases reported in the literature. We report a case of IGH manifesting as pituitary apoplexy in a young woman, diagnosed preoperatively on MR imaging as pituitary macroadenoma.

Methods: This 36-year-old woman presented with the sudden onset of left-sided retro-orbital headaches, diplopia, and left ptosis. On examination, she had a partial left oculomotor nerve palsy. Magnetic resonance imaging demonstrated an enhancing sellar mass with suprasellar extension and mass effect on the left cavernous sinus. The dorsal aspect of the lesion demonstrated a smaller nonenhancing area of low signal on T2-weighted imaging, consistent with recent hemorrhage.

Results: Clinical pituitary tumor apoplexy was suspected and the patient underwent an emergent transsphenoidal removal of the tumor to decompress the optic nerves and the left cavernous sinus. Postoperatively, the patient’s diplopia and oculomotor nerve palsy resolved completely. Histologic examination demonstrated chronic granulomatous inflammation with caseous necrosis. Stains were negative for mycobacteria, fungi and bacteria. Systemic workup for sarcoidosis, tuberculosis, syphilis, and Wegener’s granulomatosis was negative.

Conclusions: IGH is a rare lesion of the pituitary gland and an uncommon presentation of pituitary apoplexy. In the presence of caseous necrotizing granulomas, a thorough work-up for tuberculosis, sarcoidosis, and other granulomatous etiologies is warranted.

Introduction

Granulomatous hypophysitis is an inflammatory disorder of the pituitary gland characterized by the formation of intrasellar giant-cell granuloma. In the absence of underlying systemic disease such as sarcoidosis, tuberculosis, or syphilis, it is classified as idiopathic. Idiopathic granulomatous hypophysitis (IGH) is an extremely rare disorder with fewer than 20 cases diagnosed antemortem reported in the literature. Of these cases, only four were reported with preoperative MR imaging and only one of these presented with acute findings mimicking pituitary apoplexy. We report the second case of IGH manifesting as pituitary apoplexy in a young woman, diagnosed preoperatively on MR imaging as a pituitary macroadenoma.

Case Report

A 36 year old woman presented with sudden onset of left-sided retro-orbital headaches, diplopia, and left eye ptosis. On neurologic examination, she had a partial left oculomotor nerve palsy. On further investigation, the patient admitted to menstrual irregularities over the preceding 8 months and intermittent galactorrhea. Magnetic resonance imaging of the brain demonstrated an intensely enhancing 1.9 x 1.1 x 1.3 cm sellar lesion with suprasellar extension and mass effect on the left cavernous sinus. The dorsal aspect of the lesion displayed a smaller nonenhancing area of low signal on T2-weighted imaging, consistent with recent hemorrhage (Fig. 1).

Endocrinological findings demonstrated a mildly elevated serum prolactin level of 46.9 ng/mL (normal 4.8-23.3 ng/mL), an elevated thyroid-stimulating hormone level of 5.88 mU/L (normal 0.27-4.2 mU/L), and normal thyroxine, follicle-stimulating hormone, luteinizing hormone and growth hormone levels. A urine pregnancy test was negative.

Clinical pituitary tumor apoplexy was suspected and the patient underwent an emergent transsphenoidal removal of the tumor to decompress the optic nerves and the left cavernous sinus. At surgery, a firm, rubbery, yellowish-white mass was noted. The mass was adherent to surrounding arachnoid and to the left cavernous sinus. Gross total resection was achieved aside from a small remnant which remained on the pituitary stalk.

Microscopic examination demonstrated chronic granulomatous inflammation with extensive caseous necrosis. Palisading of epitheloid cells around the zones of necrosis was noted, as well as multinucleated giant cells within the necrotic zones (Fig. 2). Staining for organisms, including Gram, acid-fast, and Grocott-Gomori methenamine silver nitrate, was negative for bacteria, mycobacteria, and fungi, respectively. A skin purifed protein derivative test for tuberculosis was negative, as was a QuantiFeron Gold TB assay. Serum rapid plasma reagent (RPR) test results were normal, as was a CSF RPR test. Angiotensin-converting enzyme levels were found to be normal, as part of the workup for sarcoidosis. CSF cultures for bacteria and acid-fast bacilli showed no growth. No mycobacterium tuberculosis complex DNA was detected on polymerase chain reaction assay of CSF.

Postoperatively, the patient’s diplopia and oculomotor palsy resolved completely. Visual fields and acuity were normal. She experienced an episode of transient diabetes insipidus following surgery which responded well to pitressin and oral desmopressin. An extensive systemic workup for sarcoidosis, tertiary syphilis, and Wegener’s granulomatosis was carried out; all results were negative. Endocrinological evaluation was notable for hypothyroidism. She was discharged in excellent condition on L-thyroxine replacement therapy. Postoperative imaging 18 months out demonstrates excellent resolution of the mass lesion with decompression of the optic chiasm and left cavernous sinus (Fig. 3).

Discussion

Most cases of granulomatous hypophysitis are associated with systemic granulomatous diseases, notably sarcoidosis, syphilis and tuberculosis. The largest review of granulomatous hypophysitis carried out was by Inoue and Harvey in 1954 with 115 documented cases and 23 idiopathic cases; however, all were diagnosed postmortem by autopsy.3 Brisman et al in 1996 performed a review of antemortem diagnosis of idiopathic granulomatous hypophysitis and identified 13 cases.2 However, only 3 of these cases had preoperative magnetic resonance imaging and none of these presented with acute neurologic deterioration. Rather, the prevailing presentation of idiopathic granulomatous hypophysitis in this series was hypopituitarism. Our case is a notable exception. The only other reported case of idiopathic granulomatous hypophysitis presenting as pituitary apoplexy with consistent preoperative MR findings is by Inoue et al, in a 62 year old female.

Common radiographic features in cases of idiopathic granulomatous hypophysitis include homogenous enhancement, a well-circumscribed mass lesion and infundibular thickening. At surgery, the mass is uniformly found to be firm and densely adherent to surrounding structures. Postoperative results are generally favorable for visual dysfunction but not endocrine disturbances.

Conclusions

IGH is a rare lesion of the pituitary gland and an extremely uncommon presentation of pituitary apoplexy. As it can appear identical to pituitary adenoma both radiographically and clinically, a high index of suspicion is warranted. Treatment consists of operative intervention in the face of acute neurologic deterioration and evidence of mass effect, with hormone replacement therapy common required for hypopituitarism either pre- or post-operatively. In the presence of caseous necrotizing granulomas, a thorough work-up for tuberculosis, sarcoidosis, and other granulomatous etiologies is warranted.

References


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