Idiopathic Granulomatous Hypophysitis

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Idiopathic granulomatous hypophysitis is a chronic inflammation of the pituitary gland. It has a rare occurrence and is diagnosed only on pathologic examination. We report a case of a 60-year-old patient presenting with headache, vomiting and isolated hyponatremia. MRI study showed a space occupying lesion of the pituitary gland with a marked enhancement after gadolinium injection. Pituitary biopsy was consistent with the diagnosis of idiopathic granulomatous hypophysitis. The aim of this case report is to discuss the variable clinical and radiological aspect of the disease.

KEY WORDS: Pituitary - Idiopathic - Granulomatous - Hypophysitis.

Introduction

Inflammatory pituitary lesion are rare and represent only 0.38% of pituitary lesion. Idiopathic granulomatous hypophysitis, a rare inflammatory disease of unknown etiology, is characterized by lymphoplasmocytic infiltration of the pituitary and the presence of epitheloid cells and multinucleated giant cells constituting granulomas. Patients are misinterpreted as pituitary adenomas due to their rarity and to the nonspecific radiological feature. The diagnosis is made by histological examination if surgical resection is undertaken. We report one patient with idiopathic granulomatous hypophysitis who was diagnosed postoperatively.

Case Report

A 60-year-old woman presented with sudden, severe headache and vomiting. She had no systemic complaints. Neurological examination was normal except generalized weakness. On magnetic resonance imaging (MRI), there was a 20×20×20mm-sized intrasellar mass. The lesion was isointense on T1-weighted image, inhomogeneously high signal on T2-weighted image and was enhanced homogeneously (Fig. 1). Endocrinological finding revealed sick euthyroid state. The patient was operated upon by trans-sphenoidal route for a supposed pituitary macroadenoma. At surgery, a large, gray, and firm mass was noted. A subtotal resection was achieved. Microscopic examination of the lesion demonstrated nonnecrotising granulomatous chronic inflammation with lymphocytic infiltration and interstitial fibrosis (Fig. 2). Immunohistochemical characterization of the chronic inflammation-positive at cluster of differentiation (CD) 68 immunostaining showed predominantly macrophage. Staining for organism, including Gram-stain, acid-fast bacillus stain, and tuberculosis-polymerase chain reaction (TB-PCR) test, were negative. Culture for bacteria showed no growth. So, this case was diagnosed idiopathic granulomatous hypophysitis. The postoperative course was uneventful, but the patient was required glucocorticoid and L-thyroxine replacement therapy.
Discussion

Inflammatory diseases of the pituitary are rare. Granulomatous hypophysitis contains multinucleated giant cells. It can be a part of systemic granulomatous disease such as tuberculosis, sarcoidosis, histiocytosis X, and syphilis or an isolated pituitary disease due to a foreign body reaction to a ruptured Rathke’s cleft cyst, mycotic infection or a pituitary adenoma. If these conditions are excluded by clinical and histological examination, granulomatous hypophysitis is termed “idiopathic”.

Lymphocytic hypophysitis is distinguished from granulomatous hypophysitis by the absence of nodular aggregation of epitheloid histiocytes and multinucleated giant cells, but there are ultrastructural similarities between these two pathologies: the presence of inactive, degranulated secretory cells, focal oncocytic changes in the secretory cells and inflammatory cells within the periacinar membrane, so that it has been suggested that they have the same pathogenetic background or represent different stages of the same lesion and autoimmunity may play a role.

Clinical presentation of this disease is not always the same. There may be progressive mass effect (visual disturbance, headache), endocrinological symptom (amenorrhea, galactorrhea, diabetes insipidus), fatigue, and sudden onset headache, vomiting, generalized weakness, but no definitely abnormal endocrinological finding.

Recent reports have looked for characteristic or pathogno- mic MRI finding in inflammatory pituitary disease. The most detailed study is that of Honegger et al., who reported six cases of lymphocytic and three of granulomatous hypophysitis. In granulomatous hypophysitis, a sellar mass with a tongue-like suprasellar extension because of infundibular infiltration was a constant finding. Pamir et al. reported heterogeneous hypertrophy of the pituitary, with irregular high signal on T2-weighted images in their two cases. Vasile et al. reported heterogeneous gadolinium contrast enhancement and involvement of the cavernous sinus. However, the radiological finding of granulomatous hypophysitis has no reliable diagnostic sign. The lesion can be dumbbell-shaped, spherical or elliptical. Isointensity with the brain on T1-weighted image and heterogeneous high signal on T2-weighted images were noted. Enhancement can be homogeneous or heterogeneous. The stalk is always infiltrated in advanced lesions, and this can be tongue-like in typical case. In our case, the lesion was located in intrasellar portion without suprasellar extension, and showed isointensity on T1-weighted image and homogeneous high signal intensity on T2-weighted image. None of these radiological signs seems sufficient to differentiate hypophysitis from pituitary macroadenoma.

In general, the outcome of surgery for granulomatous hypophysitis is favorable even if the lesion is only partially removed or biopsied. Therefore, the recommendation have been to avoid major resection because of the self-limited course or to defer surgery if vision is not compromised. In view of the results reported in the literature, authors advocate early surgical intervention. It excludes the possibility of an infectious disorder and confirms the precise diagnosis. Usually, a trans-sphenoidal approach is appropriate. Abnormal tissue is removed, but normal appearing tissue is preserved to prevent pituitary dysfunction. If only an enlarged gland is encountered, one must be content with a biopsy to confirm the diagnosis. Observation alone should be recommended only in the absence of pituitary, hypothalamic, and visual dysfunction. Despite recent experience with corticosteroids therapy, surgery remains the mainstay of therapy.

Conclusion

In summary, idiopathic granulomatous hypophysitis is a rare disorder of the pituitary gland that carries a favorable prognosis. Treatment is composed of hormone replacement therapy when hypopituitarism is present, and biopsy if the presence of a systemic disorder is suspected. Radical excision of the lesion is not recommended, because it will adversely affect pituitary function and these lesions respond well to steroid therapy.

References

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