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MR Demonstration of Wegener Granulomatosis of the Infundibulum, a Cause of Diabetes Insipidus

Edward J. Czarnecki and Eric M. Spickler

Summary: We report a case of Wegener granulomatosis causing hyperprolactinemia followed by central diabetes insipidus. The initial T1-weighted MR image showed an isointense heterogeneous sellar mass. After the onset of diabetes insipidus, repeat sagittal and postcontrast T1-weighted images showed marked infundibular thickening, enlargement of the sellar mass, and enhancement of both the infundibulum and hypothalamus. Follow-up MR after marked clinical response to corticosteroids showed nearly complete resolution of the aforementioned abnormalities.

Index terms: Granuloma; Diabetes insipidus; Pituitary gland, inflammation

Central diabetes insipidus and hyperprolactinemia are rare complications of Wegener granulomatosis. We recently encountered a patient with Wegener granulomatosis in whom secondary amenorrhea and galactorrhea followed by severe polydipsia and polyuria developed. Magnetic resonance (MR) imaging of the pituitary and juxtasellar structures showed lesions with characteristic features of granulomatous disease. Repeat MR imaging after treatment with high-dose corticosteroids showed resolution of these abnormalities. Although pathologic proof was not obtained, the clinical history and response to corticosteroids establish Wegener granulomatosis as the cause of the diabetes insipidus and hyperprolactinemia.

Case Report

A 34-year-old woman was referred for evaluation of hyperprolactinemia (headaches, secondary amenorrhea, and galactorrhea). Wegener granulomatosis had been diagnosed 3 years earlier on the basis of clinical history, autoimmune serology, and nasopharyngeal biopsy. Pertinent clinical history included arthralgias, sinusitis with saddle nose deformity, tracheobronchitis, and hearing loss related to otitis media. Nasopharyngeal biopsy showed chronic granulomatous inflammation compatible with Wegener granulomatosis. Initial MR imaging (Fig 1) showed an isointense sellar mass with discrete suprasellar extension. One month later, before any intervention, the patient noted the gradual onset of polydipsia and polyuria. A diagnosis of central diabetes insipidus was made, and the patient was started on desmopression. Repeat MR imaging (Fig 2A and B) showed enlargement of the sellar mass accompanied by new enlargement of the infundibulum. Postcontrast images showed homogeneous enhancement of the pituitary stalk and hypothalamus. In light of the clinical history and marked progression of MR abnormalities, a diagnosis of Wegener granulomatosis was made. The patient was treated with high-dose corticosteroids, and her clinical condition improved dramatically. MR imaging (Fig 3A and B) 2 months after the beginning of treatment showed nearly complete resolution of the intrasellar mass, pituitary stalk enlargement, and hypothalamic enhancement.

Discussion

Wegener granulomatosis is a disease characterized by necrotizing vasculitis and granulomatous inflammation of the upper and lower respiratory tracts together with glomerulonephritis (1, 2). It is unusual for the disease to be restricted to the upper respiratory tract (incomplete or localized granulomatosis, as in our patient) (2). The cause is unknown, but most studies suggest a hypersensitivity disorder. The mean age of onset is around 40 years. There is a 2:1 male-to-female ratio. The spectrum of clinical signs and symptoms is broad. Patients can present with arthralgia, upper airway syndrome with persistent rhinorrhea, sinusitis, and otitis media complicated by hearing loss. Patients usually do not present with renal problems. The prognosis for this otherwise lethal disease has improved since cyclophosphamide and corticosteroids have been used in treatment (1).

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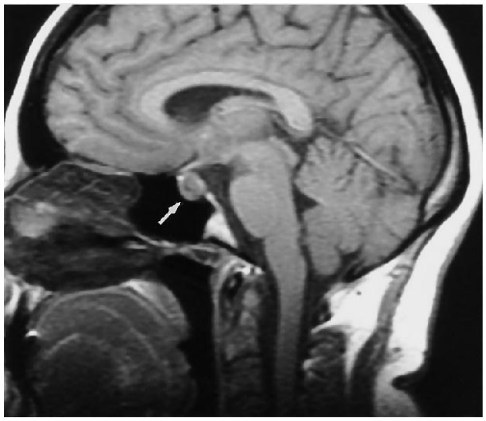

Fig 1. Initial evaluation for hyperprolactinemia. Unenhanced sagittal T1-weighted MR image (370/11/2 [repetition time/echo time/excitations]) shows an isointense heterogeneous sellar mass (*arrow*) that projects slightly into the suprasellar cistern.

Involvement of the nervous system is uncommon and may be from vasculitis of the vasa vasorum resulting in peripheral or central neuropathy, extension from underlying sinus disease, or primary nectorizing granulomas. Hyperprolactinemia and diabetes insipidus are very rare complications. We found only a few cases of Wegener granulomatosis causing diabetes insipidus and one case causing both hyperprolactinemia and diabetes insipidus (3, 4, 5). The presumed mechanism of involvement in these cases was thought to be either hypothalamic vasculitis or direct granulomatous involvement or both.

Central diabetes insipidus can result from a number of diseases that affect the hypothalamic-neurohypophyseal axis. The causes of central diabetes insipidus include Langerhans cell histiocytosis, germinoma, craniopharyngioma, hypothalamic glioma, granulomatous inflammation, surgery, and trauma. The MR findings of central diabetes insipidus are diffuse or focal infundibular thickening and absence of the normal high-intensity signal in the posterior pituitary lobe seen on T1-weighted images (6). Although the source of the hyperintense signal in the posterior pituitary gland on the T1-weighted images remains controversial, the literature

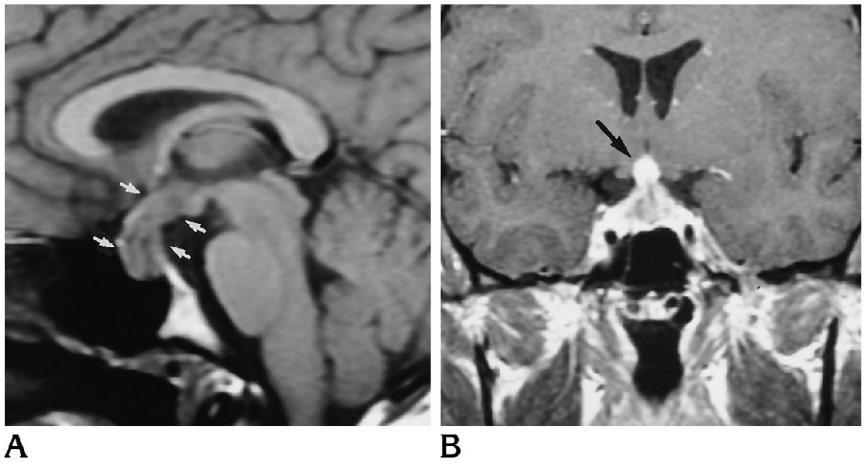

Fig 2. Diabetes insipidus developed before therapy, and a progress examination was performed.

A, Sagittal T1-weighted spin-echo image (370/11/2) shows interval development of marked infundibular thickening extending to the hypothalamus (*arrows*) with persistence of the sellar mass. The posterior pituitary bright spot is absent.

B, Postcontrast coronal T1-weighted spin-echo image (370/12/2) shows marked homogeneous enhancement of the thickened infundibulum and hypothalamus (*arrow*) as well as the periphery of the sella.

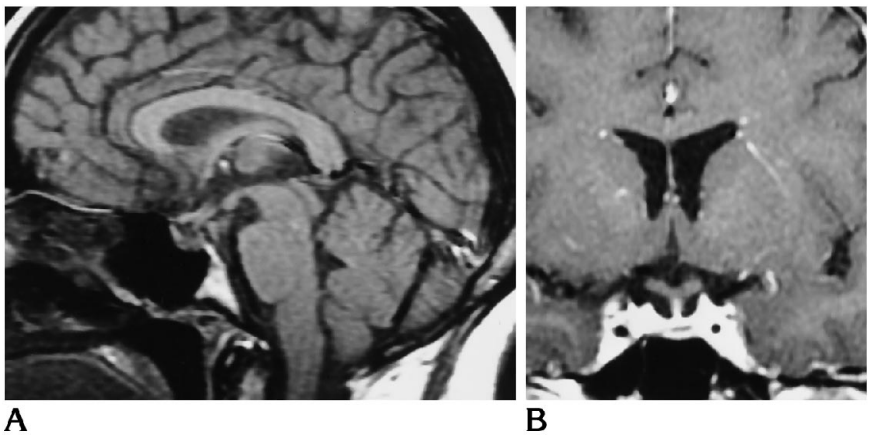

Fig 3. Sagittal (*A*) and postcontrast (*B*) T1-weighted images (370/12/2) after successful corticosteroid therapy show nearly complete resolution of sellar mass, infundibular thickening, and infundibular/hypothalamic enhancement.

agrees that it is a functional marker of the neurohypophysis and is absent in central diabetes insipidus. The posterior bright spot can be detected in nearly 100% of healthy persons when high-resolution MR imaging is used. However, most routine brain imaging protocols do not provide high-resolution sequences of the sella and, therefore, the posterior pituitary bright spot may appear absent in healthy persons (7). However, the presence of a bright signal is inconsistent with the diagnosis of central diabetes insipidus. It should also be noted that in cases of tumoral, surgical, or traumatic infundibular distortion, identification of infundibular thickening usually is not possible.

By coupling the clinical data with the MR imaging characteristics, the specific cause of central diabetes insipidus usually can be made. In this case, the patient had hyperprolactinemia; even though the initial MR findings showed isolated sellar involvement, we hypothesize that the hyperprolactinemia was caused by occult disturbance of prolactin inhibitory factor at the hypothalamic level. Development of diabetes insipidus ensued, and repeat MR showed marked abnormalities within the sella, infundibulum, and hypothalamus. A posterior pituitary bright spot was not identified. The hyperprolactinemia and diabetes insipidus resolved after corticosteroid therapy, with nearly complete resolution of MR abnormalities, thus establishing Wegener granulomatosis as the cause of the hypothalamic-pituitary disturbance.

In summary, this case demonstrates the utility of MR imaging in the evaluation of diseases of the hypothalamic-pituitary axis. In the appropriate clinical setting, Wegener granulomatosis should be considered in the differential diagnosis of abnormalities involving the sella and hypothalamic-neurohypophyseal axis.

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