

Sarcoidosis of the hypothalamus and pituitary stalk

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Abstract: We report a rare case of sarcoidosis of the hypothalamic and suprasellar region, with clinical course and the magnetic resonance imaging follow-up.

Key words: Sarcoidosis – Neurosarcoidosis – Magnetic Resonance Imaging – Pituitary insufficiency

Introduction

Sarcoidosis is a multisystem, noncaseating, granulomatous disease of unknown origin [1]. Involvement of the central nervous system has been observed in up to 14% of autopsies in cases of systemic sarcoidosis [2].

We present the case of a 52-year-old man with a sarcoid granuloma in the hypothalamus and suprasellar region, detected with magnetic resonance imaging (MRI). The lesion was followed up by two further MR examinations, while the patient was treated with high-dose corticosteroids and hormone replacement.

Case report

The patient had suffered only ever sports injuries. In July 1989, he had an attack of extreme pain in the left flank and high fever after an exhausting tour in the mountains. Renal calculi were diagnosed. In September 1989, he felt increasing chills and fatigue during a sailing tour and, 1 month later, had an attack of fever (39 °C), chills without respiratory infection, weakness and fatigue. On 13 March 1990, a chest radiograph showed bilateral hilar lymph node enlargement. Erythema nodosum was observed at the same time, leading to the diagnosis of sarcoidosis. The patient then recalled that he had had pain in both ankles early in 1989. In April 1989, the patient had rapidly developed diabetes insipidus with a maximum of 10 l fluid intake per day. The diabetes insipidus was aggravated

when prednisolone was given as treatment for sarcoidosis. At the same time, severe hypotension was detected.

Suspected hypopituitarism was confirmed by cortisol levels below 4 µg/dl throughout the day (3 examinations) and a response to stimulation by ACTH (32,5 µg/dl). The patient immediately felt stronger when given prednisolone. He had a low basal TSH (0,2 mu/l) with a poor response to stimulation by TRH (4,37 mu/l). In May 1990, perimetry showed a left upper outer quadrant field defect. These findings indicated a diagnosis of acute sarcoidosis with anterior and posterior pituitary insufficiency, secondary to sarcoidosis of the pituitary stalk and hypothalamus.

An MR examination in July 1990, when the patient was already receiving prednisolone, revealed on T1-weighted images a 21 × 12 × 15 mm lesion in the hypothalamus and suprasellar region (Fig. 1). The lesion was of variable signal intensity both before and after i. v. Gd-DTPA application. The hypothalamic portion was uniformly hyperintense, but the presumed cystic suprasellar portion was isointense with grey matter before injection and showed marginal enhancement. This component extended up beyond the optic chiasm, explaining the visual defect.

Corticosteroid therapy was continued and, in September 1990, a follow-up showed a minor reduction in size of the hypothalamic portion of the lesion (Fig. 2). The suprasellar portion was unchanged in size; neither showed any change in signal intensity with Gd-DTPA.

The corticosteroids were gradually reduced to 10 mg/d by December 1990. The visual defect remained unchanged and a second follow-up MR examination in November 1990 (Fig. 3) showed a slight increase in size of the hypothalamic portion, which was now isointense with grey matter on T1-weighted images and showed only minor enhancement. The suprasellar part was unchanged in size and signal intensity. The patient developed a complete temporal hemianopia in the left eye during summer 1990, while the right eye showed an upper outer quadrant defect, which remained unchanged. The patient declined surgery for these

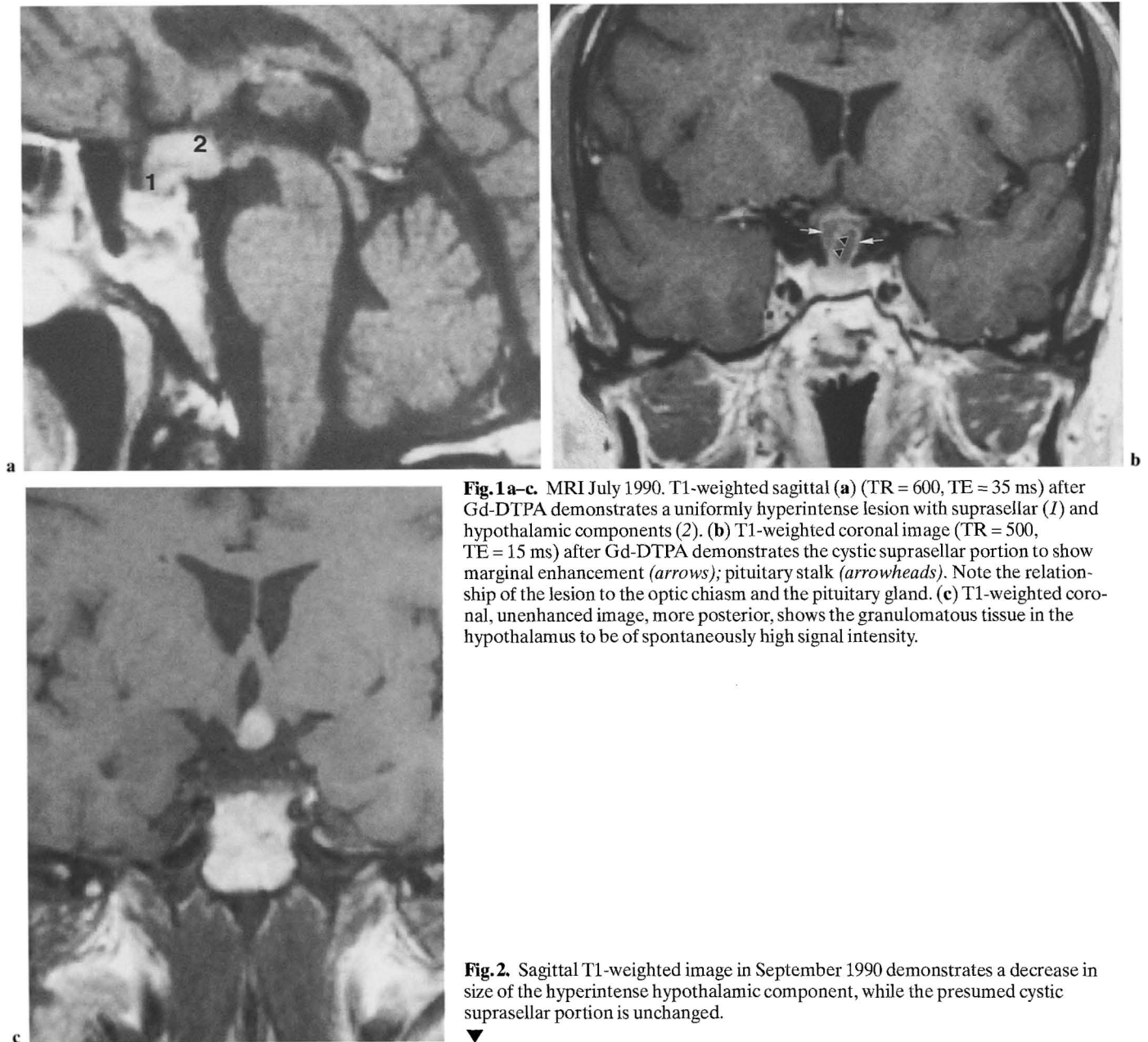


Fig. 2. Sagittal T1-weighted image in September 1990 demonstrates a decrease in size of the hyperintense hypothalamic component, while the presumed cystic suprasellar portion is unchanged.



defects at this time. He went back to work on replacement therapy with thyroxine, testosterone and DDAVP. Prednisolone, started with 80 mg/d, was gradually reduced to 10 mg/d (December 1990).

Due to the increasing visual defect the patient agreed to surgical therapy in May 1991. The intraoperative findings showed a cystic suprasellar portion and a homogeneous component in the hypothalamus. The microscopical appearance of the surgical specimen tends to verify the diagnosis of sarcoidosis.

Discussion

Clinical involvement of the central nervous system (CNS) is seen in approximately 5% of cases of systemic sarcoidosis and may be the initial presentation in 2.5% [3]. In-

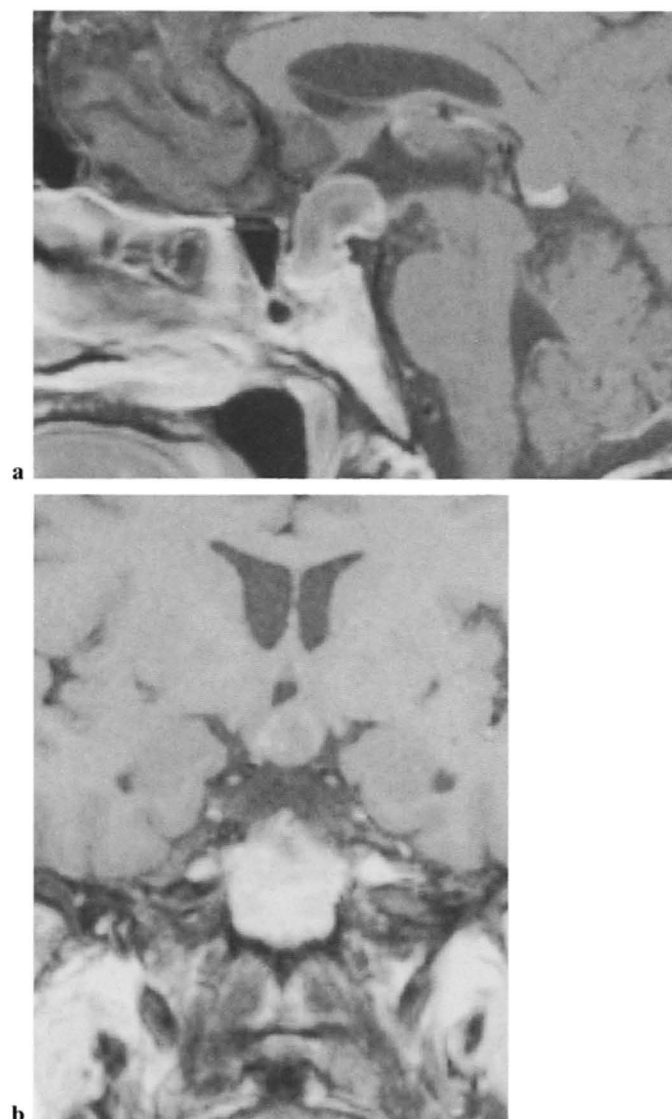


Fig. 3a and b. Sagittal (a) and coronal (b) image after Gd-DTPA in November show alterations in signal intensity and size in both parts of the lesion; the presumed cystic component being more extensive.

involvement of the hypothalamic-pituitary region with resulting anterior pituitary insufficiency is even rarer, with 51 cases reported up to 1989 [4]. CNS manifestations of sarcoidosis include cranial neuropathies due to compression or infiltration in 50–70% of affected cases. The most frequent manifestation (up to 50%) is a peripheral seventh nerve palsy. Meningeal infiltration, leading to aseptic meningitis, is seen in approximately 20%, hydrocephalus due to ventricular outlet obstruction in 15% and local parenchyma lesions with resulting hypophyseal-hypothalamic dysfunction in 15–25% [3, 5].

Contrast-enhanced CT is helpful in the investigation of suspected intracranial sarcoid granulomas [3, 5, 6], although the findings are not specific and may resemble those at meningiomas, lymphoma and primary or metastatic tumours [8, 9]. CT is of limited value for detection of lesions in the hypothalamus and the hypophyseal region. In our case, enhanced CT did not show the hypothalamic

lesion and gave insufficient detail to assess the size of the suprasellar portion. If involvement of this region is suspected, MR is indicated.

MR provided greater sensitivity than CT in the assessment of hypothalamic-hypophyseal involvement, although there is a great variability in the appearances of intracranial sarcoidosis on MR [7]. The signal may be iso- or hypointense compared to grey matter on T1-weighted images [7, 10].

These signal characteristics do not help to differentiate these lesions from en plaque meningiomas, lymphoma and unusual metastases [8, 9]. Especially the craniopharyngioma with cystic and solid components is an important differential diagnosis in the suprasellar region. The MR appearance of the craniopharyngioma is variable. Solid portions of this lesion tend to be isointense to grey matter, whereas cystic components tend to be hyperintense on T1-weighted images [11]. The first MR examination in our case showed the signal intensities of the two components of the lesion to be different and they changed in the follow-up examinations; both showed lower signal intensity before and after Gd-DTPA administration on the last examination.

There was good correlation of clinical and MR findings. The patient developed diabetes insipidus which could be explained by the hypothalamic lesion. The suprasellar portion of the granuloma extended above the optic chiasm and infiltrated the pituitary stalk, explaining anterior pituitary insufficiency, with low cortisol and TSH levels, and the visual defect.

MRI is most helpful for detection of sarcoid involvement of the hypothalamus and pituitary region and is the technique of choice for the follow-up and assessing the response to steroid therapy.

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