

Intracranial Inflammatory Pseudotumor: A Case Report and Review of the Literature

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Abstract

Inflammatory pseudotumors are lesions characterized by inflammatory cell infiltration and fibrosis. Central nervous system (CNS) inflammatory pseudotumors are much rarer, and generally manifest in the sellar and parasellar regions. We report a patient with an inflammatory pseudotumor in the temporal and occipital lobes treated in our department in 2017, and present a literature review. We discuss the etiology, imaging, pathology, and treatment of intracranial inflammatory pseudotumors.

Keywords: Central Nervous System; Inflammatory Pseudotumor; Treatment

Introduction

Inflammatory pseudotumors are lesions characterized by inflammatory cell infiltration and fibrosis. Inflammatory pseudotumors can occur in almost any organ, including the lungs, mesentery, omentum, retroperitoneum, genitourinary system, and upper respiratory tract [1-4]. Central nervous system (CNS) inflammatory pseudotumors are much rarer [5], and generally manifest in the sellar and parasellar regions. These pseudotumors lack specific clinical manifestations or imaging characteristics. Histopathological diagnosis is the gold standard for inflammatory pseudotumors. The outcome of the treatment of CNS inflammatory pseudotumors remains poor; they are difficult to cure completely and often recur. Therefore, the etiology of CNS inflammatory pseudotumors must be determined to improve treatment outcomes.

We report a patient with an inflammatory pseudotumor in the temporal and occipital lobes treated in our department in 2017, and present a literature review. We discuss the etiology, imaging, pathology, and treatment of intracranial inflammatory pseudotumors to facilitate recognition of the disease by clinicians, and optimize treatment efficacy.

Case report

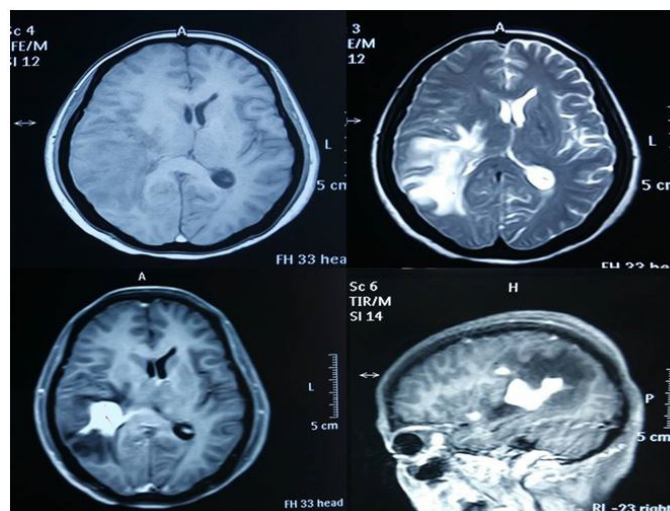


Figure 1: Magnetic resonance imaging (MRI) showing an irregular lesion in the temporal and occipital lobes accompanied by peripheral edema with irregular, nodular enhancement

A 51-year-old woman presented with a headache for 6 days that had worsened in the previous 16 hours. She had no recent history of fever or drug use, and no history of diabetes or hypertension. Neurological examination was normal. There was no abnormality in routine laboratory examination, including total blood account, biochemical parameters. Intracranial magnetic resonance imaging (MRI) showed abnormal signals in the right temporal and occipital lobes, with irregular and nodular enhancement accompanied by peripheral edema (Figure 1), which was thought to be a lymphoma.

The patient presented with clinical symptom of headache, and magnetic resonance imaging showed space-occupying lesions in the right temporal and occipital lobe, so surgical treatment was planned. Neuronavigation-guided microsurgery was performed. Corticectomy was conducted over the temporal and occipital lobes and the lesion was resected fully. Pathologically, there was no clear evidence of malignant transformation. Hematoxylin and eosin (H&E) staining showed diffuse infiltration of inflammatory cells in the brain tissue and perivascular lymphocytic sheath, and occasional foam cells, mitotic signs, and glial hyperplasia (Figure 2). The lesion was considered to be inflammatory. Pathological sections sent to Huashan Hospital for immunohistochemistry examination showed: Syn(+), NeuN(+), Ki67(1%), β -amyloid (-), IDH1(-), P53(+), Olig2(+), GFAP(+), LCA(+), CD3(+), KP1(+). The results were conformed to inflammatory lesion. After methylprednisolone treatment for 2 weeks, the patient resumed normal activities and did not feel any discomfort.

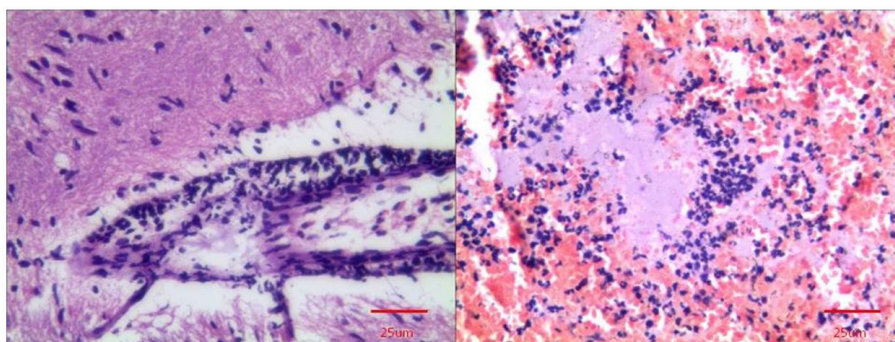


Figure 2: Microphotographs showing diffuse infiltration of inflammatory cells in the brain tissue and perivascular lymphocytic sheath, with occasional foam cells, mitotic signs, and glial hyperplasia (hematoxylin and eosin [H&E] $\times 400$)

Two months later, the patient deteriorated and showed headache and memory decline. MRI revealed mixed signals in the right suprasellar area and medulla oblongata, mainly hyperintense T1 and T2 signals. An enhanced scan showed abnormal enhancement (Figure 3). The cortisol treatment was repeated and the patient underwent radiotherapy for 2 months. At the 2-year follow-up, the patient had recovered.

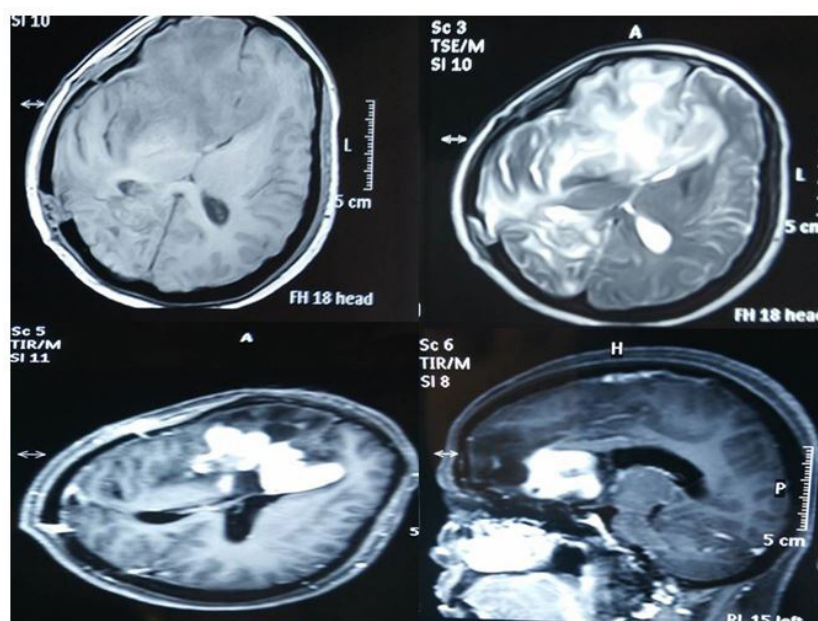


Figure 3: Follow-up MRI showing strong mixed signals in the right suprasellar area and medulla oblongata. These were mainly hyperintense T1- and T2-weighted signals, with abnormal enhancement

Discussion

Inflammatory pseudotumors are thought to be associated with viral, bacterial, and fungal infections, as well as stem cell

transplantation [6]. The evidence for these associations includes the isolation of viral DNA, such as Epstein–Barr virus, from damaged tissues [7,8]. Some patients show laboratory findings indicating infection with human herpes virus, and exhibit autoimmune dysfunction. However, the pathogenesis of herpes virus is not clear and may involve interferon (IFN)- α , interleukin (IL)-4, or immunoglobulin G4 (IG-4), which can cause fibroblast proliferation [9,10]. Inflammatory pseudotumors of the CNS are characterized by fibroblast proliferation and B cell differentiation. Although inflammatory pseudotumors are considered benign, they can occasionally undergo malignant transformation [11].

Inflammatory pseudotumor is also known as inflammatory myoblastoma, nodal lymphoid hyperplasia, adipose fibroma, fibrous xanthoma, and cytoplasmic granuloma [12]. The histopathology shows tumor-like tissue consisting of lymphocytes, plasma cells, macrophages, and foam cells. Inflammatory pseudotumor usually occurs in the lungs, orbit, mesentery, omentum, retroperitoneum, genitourinary system, and upper respiratory tract. Orbital inflammatory pseudotumors are divided into three types: 1) lymphocytic infiltration type, with marked lymphocytic infiltration; 2) sclerosing type, with marked proliferation of fibrous connective tissue; and 3) mixed type, with many infiltrating lymphocytes and plasma cells surrounded by lymphatic follicles, germinal centers, and fibroblasts. Inflammatory pseudotumors in the brain are rare, and are generally limited to the sellar and parasellar regions, cervical epidural space, meninges, and peripheral nerves. Unlike inflammatory pseudotumors occurring elsewhere, intracranial inflammatory pseudotumors often destroy the skull base and cause neuralgia. Our patient's tumor was indistinguishable from the brain lobe, similar to invasion of glioma cells.

Images of intracranial inflammatory pseudotumors generally reveal diffuse soft-tissue lesions. On MRI, inflammatory pseudotumors are commonly hypointense in T1- and T2-weighted images, and are enhanced slightly by injection with contrast medium. There are reports of inflammatory pseudotumors that are hypointense on T1- and hyperintense on T2-weighted images [13], a pattern that characterizes pathological changes. Lymphocyte infiltration-type inflammatory pseudotumors are mainly characterized by lymphocyte proliferation, resulting in a hypointense T1 and hyperintense T2 signal on MRI with obvious enhancement, similar to a typical solid tumor. Pathological changes in a sclerosing inflammatory pseudotumor include collagen fibrous hyperplasia and small cell components. On MRI, sclerosing-type pseudotumors are hypointense or isointense on T1, and hypointense T2, and are not enhanced or only slightly enhanced, similar to fibrous tissue. In mixed-type pseudotumors in cases of chronic inflammatory disease, imaging features often include obvious contrast enhancement according to the degree of fibrosis, fatty infiltration, edema, and a space-occupying effect. Computed tomography may reveal erosion, remodeling, sclerosis, and thickening. Inflammatory pseudotumors of the CNS can be invasive, potentially destroying the skull base thus making it difficult to identify the malignant tumor on imaging examinations. Our patient's lesion was hypointense on T1-weighted MRI images and hyperintense on T2-weighted images, and showed inhomogeneous enhancement. Based on the pathological changes, the lymphocyte was classified as infiltrating type.

Intracranial inflammatory pseudotumors are often treated with steroid therapy, radiotherapy, or surgery. Because intracranial inflammatory pseudotumors are rare [5], there is no standardized treatment; clinicians often apply the same treatment used for lung inflammatory pseudotumors i.e., oral prednisone [14]. Radiotherapy is generally used for those who are insensitive to steroids, show recurrence after steroid therapy, or have contraindications to high-dose steroid therapy. Generally, low-dose (20–40 Gy) fractionation therapy is advocated. Low-dose radiotherapy showed efficacy in 66% of orbital inflammatory pseudotumors [15], but is not effective for intracranial lesions. Some doctors have proposed that high-dose radiation therapy be used to simultaneously control pain and intracranial neurological symptoms, where this might have a better curative effect [16]. The prognosis of inflammatory pseudotumors is poor because of incomplete resection and the postoperative recurrence rate is 25% [17]. No surgical treatment of suspected lesions has been proposed to prevent the spread of intracranial lesions. Surgical treatment is suitable for patients who can undergo partial or complete resection without sustaining damage to important nearby structures, but not for most intracranial inflammatory pseudotumors due to limitations associated with the anatomical site, i.e., the close proximity of important vessels and nerves. It is difficult to remove the lesion completely without damaging other important structures. Therefore, surgical treatment should not be a priority for intracranial inflammatory pseudotumors.

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