Case Report

Fibrosing pseudotumor associated with a Rathke’s cleft cyst causing cavernous sinus syndrome

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Abstract: Inflammatory fibrosing pseudotumors of the head most commonly occur in the orbit, but rarely, this condition may be encountered in the parasellar area. A 56-year-old female presented with left 3rd nerve palsy and a headache, whose magnetic resonance images displayed two associated sellar lesions with distinct characteristics, namely, a Rathke’s cleft cyst in the right sellar area, and a homogenously enhancing mass spreading from the left sellar area to the cavernous sinus and clivus. The mass was later diagnosed based on pathology findings to be an inflammatory fibrosing pseudotumor. The patient was managed using transsphenoidal surgery for the Rathke’s cleft cyst, and this was followed by prolonged steroid medication to treat the inflammatory fibrosing pseudotumor. The authors advise that inflammatory fibrosing pseudotumors be included in the differential diagnosis of lesions that coexist with Rathke’s cleft cyst and spread over the cranial base.

Keywords: Fibrosing pseudotumor, Rathke’s cleft cyst, cavernous sinus syndrome

Introduction

A fibrosing pseudotumor of the head is an inflammatory, fibrotic lesion which most commonly involves the orbit [1]. Its pathological features in different organs have been described as retroperitoneal fibrosis [2], mediastinal fibrosis [3], sclerosing cholangitis, and Riedel’s thyroiditis [4]. An inflammatory fibrosing pseudotumor of the sellar area is a very rare non-neoplastic lesion; furthermore, the coexistence of Rathke’s cleft cyst and inflammatory fibrosing pseudotumor has not been reported previously.

Here, we describe a case of Rathke’s cleft cyst and a concomitant fibrosing pseudotumor extending into the cavernous sinus and manifesting as cavernous sinus syndrome.

Case presentation

History and examination

A 56-year-old post-menopausal female presented with diplopia, left ptosis, a headache, and vomiting of 10 days duration. She had experienced no previous medical problems except for a severe headache occurrence several months earlier. A Brain CT scan with contrast enhancement at admission demonstrated a left cavernous sinus mass extending to the clivus. Routine laboratory test results were within normal limits, except for a moderate C-reactive protein elevation and leukocytosis. Immunological tests (rheumatoid factor, antinuclear antibodies, C3 and C4 levels, antineutrophil antibodies, angiotensin-converting enzyme) were also normal. An endocrinological examination showed that the basal plasma values of her pituitary hormones were within normal ranges, except for a slightly elevated prolactin level. An analysis of her cerebrospinal fluid showed average cell numbers and protein levels, and a cytological examination and culture findings were negative, as were the fungal and acid-fast bacilli studies. The ophthalmologic evaluation revealed left oculomotor nerve and abducens nerve palsy, but normal visual acuity and visual fields.

Brain magnetic resonance imaging (MRI) (Figure 1) displayed a sellar lesion with two dis-
Fibrosing pseudotumor with Rathke’s cleft cyst

tinct components. The right side of the lesion on contrast-enhanced T1-weighted images showed a peripheral contrast enhancement sparing the central cystic portions and was hyperintense on T2-weighted images. On the other hand, the left side of the lesion infiltrated into the left cavernous sinus and extended to the clivus and was homogenously enhanced on contrast-enhanced T1-weighted images. Furthermore, the left tentorium connected to the left cavernous sinus was thickened and well enhanced on contrast-enhanced T1-weighted images.

**Operation**

Endoscopic endonasal transsphenoidal surgery was addressed to obtain a pathologic specimen and to relieve the intrasellar pressure, thus helping in recovery from the cavernous sinus syndrome. Intravenous corticosteroid treatment was started 3 days before the surgery and continued 7 days postoperatively. After removing the anterior wall of the sphenoid sinus, there was exposed granulated tissue, and a biopsy specimen was acquired. Soon after incision of the basal dura, a well-encapsu-

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**Figure 1.** Coronal (A and B) and sagittal (C and D) gadolinium enhanced MR images showing a cystic mass (arrow) with rim enhancement in the right pituitary fossa and a relatively well-enhanced mass infiltrating the left cavernous sinus (arrowhead) and spreading to the clivus (double arrows).
A mass was identified at the left pituitary fossa. During dissection of the tumor margin, a yellowish, milk-like material containing necrotic tissue extruded from the gap between the encased mass and the dura. Because the encased mass was attached to the clivus along the dura, complete removal was impossible. Once achieving the acquisition of a small specimen for the histopathologic analysis, no further removal of the lesion was attempted.

**Histological examination**

A microscopic examination of the right intrasellar lesion revealed amorphous eosinophilic materials and several scattered macrophages (Figure 2A). They were lined with epithelial cells with a tall columnar shape and apical cilia (Figure 2B). The histopathological diagnosis of the right intrasellar lesion was Rathke’s cleft cyst. However, the left sellar lesion demonstrated much plasma cell infiltration admixed with fibroblasts and vascular proliferation which can be interpreted as an inflammatory fibrosing pseudotumor (Figure 2C and 2D).

**Postoperative course and follow-up**

At the immediate post-operative exam, there was no improvement of her 3rd nerve and 6th nerve palsy, but the headache and vomiting disappeared. The patient was discharged after a short hospital stay. At her 1-month follow-up exam, the diplopia seemed to be improved.
slightly. The oral steroid medication was continued for 6 months. After 4 months of steroid treatment, her 3rd and 6th nerve palsy were completely recovered, and a follow-up MRI (Figure 3) demonstrated that the left solid mass had reduced significantly. An ophthalmologic evaluation performed at the time showed no evidence of either 3rd or 6th nerve palsy, and an endocrinological re-examination revealed no evidence of hormone abnormality. She has been well during the 23 months after that.

Discussion

Inflammatory fibrosing pseudotumor of the head is a localized form of idiopathic hypertrophic pachymeningitis [4] and an idiopathic, inflammatory lesion characterized by inflammatory cell infiltration and fibrotic responses [5]. Its common involvement includes the skull base, orbit, ear, parotid gland, paranasal sinuses, pterygomaxillary space and multiple systemic organs, such as upper respiratory tract, omentum, mesentery, retroperitoneum, and genitourinary tract [1, 6].

The diagnosis of fibrosing pseudotumor, especially when it involves the pituitary and skull base, is not straightforward based on radiologic findings, because its differential diagnosis requires the exclusions of infectious, neoplastic, and immunological diseases. Accordingly, sophisticated diagnostic procedures, such as histological, histochemical, and immunohistochemical analysis are often necessary to provide an accurate diagnosis and exclude lesions of known etiology [7].

Histologic features vary from extensive fibrosis to the predominance of mixed inflammatory cells, but it seems that inflammatory reactions caused by lymphocytes, plasma cells, and macrophages initiate the disease process followed by fibrotic responses and the formation of granulation tissue, which finally becomes fibrotic tissue.

Regarding the treatment, corticosteroids have been reported to be most effective in preventing inflammatory progression [8-10]. Immunosuppressive agents like methotrexate and azathioprine may help to prevent the pathologic process when attempting to taper corticosteroids [8]. Although radiotherapy does not appear to be an effective treatment modality for inflammatory pseudotumors [9], there was a report that adjuvant radiotherapy after steroid treatment helped the resolution of the disease [11].

Although the etiology of inflammatory fibrosing pseudotumor has not been elucidated, in this described case, the authors presume that its etiology was associated with the presence of...
Fibrosing pseudotumor with Rathke’s cleft cyst

Rathke’s cleft cyst rupture. Various reports on Rathke’s cleft cyst rupture causing inflammatory reactions have been issued [12, 13]. Schittenhelm [13] reported hypophysitis caused by Rathke’s cleft cyst rupture and postulated that the inflammatory process developed due to a persisting foreign body reaction caused by the leakage of the cystic contents.

Given that, a differential diagnosis of lymphocytic hypophysitis should be considered. Based on previous imaging and clinical findings, lymphocytic hypophysitis appears as an expansive contrast-enhancing lesion, accompanying endocrinological abnormalities usually involving thyroid, adrenocortical, or gonadal insufficiency [7]. And it is generally found to be a form of localization and is almost always limited to the sellar area. Therefore, the authors could differentiate it from the lymphocytic hypophysitis in that the lesion had such an extensiveness that which reached not only the cavernous sinus, but the clivus, and the patient had not had any hormonal imbalance at the presentation.

To the best of our knowledge, this is the first report on the occurrence of an inflammatory fibrosing pseudotumor caused by rupture of a Rathke’s cleft cyst resulting in cavernous sinus syndrome. It is possible in our case that the cystic contents provoked a lymphocytic inflammatory process in the parasellar area, which extended to the clivus and tentorium. Presumably, this resulted in a localized form of lymphocytic meningitis and the formation of granulation tissue, which consequently converted into fibrotic tissue.

Generally, Rathke’s cleft cyst rupture causes a headache due to inflammation or cavernous sinus syndrome due to the tumor mass per se. We describe a case of Rathke’s cleft cyst rupture that caused the formation of an inflammatory fibrosing pseudotumor, which manifested as cavernous sinus syndrome.

Conclusion

In conclusion, we suggest that Rathke’s cleft cyst rupture was the first pathogenic event that induced the development of an inflammatory fibrosing pseudotumor causing cavernous sinus syndrome. Inflammatory fibrosing pseudotumor should be included in the differential diagnosis of lesions that spread over the cranial base, especially in the sellar area when the lesion coexists with Rathke’s cleft cyst.

Disclosure of conflict of interest

None.

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Fibrosing pseudotumor with Rathke's cleft cyst

