Intracranial inflammatory pseudotumor mimicking malignant neoplasm

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Inflammatory pseudotumor is a non-neoplastic process with unknown etiology, and it most frequently involves the lungs and orbits. It is also characterized by proliferation of the connective tissues with inflammatory infiltrates.[1] Primary intracranial inflammatory pseudotumor is exceptionally uncommon and typically arises from the meninges with a predilection for skull base and cavernous sinus. In this report, we describe a rare lesion in the brain parenchyma of the left temporal lobe in a 41-year-old man, which was initially suspected to be a glioma and considered to be a non-Hodgkin's lymphoma after intraoperative frozen-section examination. We also describe its rare progression developing along the meninges after surgery.

A 41-year-man presented with headache for 7 years and aggravation for one month. Fundus examination revealed optic disc pallor in the left eye, which also had a visual acuity of 6/24 on Snellen's chart. The vision of the contralateral eye appeared normal. The CT results showed a low-density lesion in the left temporal lobe. The contrast-enhanced MRI results showed a lesion hypointense to gray matter on T1-weighted images and hyperintense on T2-weighted images with homogenous and avid contrast enhancement partially encasing the cerebral dura mater in the polus temporalis (**Figure 1**). A diagnosis of glioma was considered, and he was planned for right orbitozygomatic craniotomy and tumor excision. The temporal lesion was found grayish white and mucinous intraoperatively, and increased thickness of the cerebral dura mater in the polus temporalis was also observed. Frozen-section examination showed the signs of non-Hodgkin's lymphoma. Therefore, total excision of the temporal lesion and decompressive craniectomy was performed. The final diagnosis of inflammatory pseudotumor was confirmed through histopathology. His headaches were diminished after the surgical recovery, but recurred with complaints of vision diminution of the left eye 2 months later. The MRI results showed a total resolution of the temporal lesion. There was extensive meningeal thickening, enhancement of the bilateral cavernous sinuses, high posterior clinoid area, and distal petrous internal carotid artery canals. A lesion developing along the cerebral dura mater was also observed. The patient was treated with dexamethasone at a dosage of 20 mg daily for 7 days, and the treatment then continued with steroids at 70 mg of prednisone daily. The headaches diminished with no further progression of vision diminution. An MRI also demonstrated diminution of the lesion 3 months after surgery. He was doing well with no signs of developing new symptoms 10 months post surgery.

Inflammatory pseudotumor is also referred to by several names such as plasma cell granuloma, inflammatory fibrosarcoma, inflammatory myofibroblastic tumor, inflammatory myofibrohistiocytic proliferation, and xanthomatous pseudotumor.[2] The etiology of inflammatory pseudotumor is still unclear, but it has been reported that the disease is commonly associated with exaggerated immunological processes after trauma or infection. It may be also associated with several autoimmune diseases. Although inflammatory pseudotumors can occur in many organs in the body such as the stomach, pancreas, heart, kidney, and skin, the most frequently involved ones are lungs and orbits. Primary intracranial inflammatory pseudotumor is exceptionally uncommon and typically arises from the meninges with a predilection for the skull base and cavernous sinus. The involvement of the brain parenchyma is extremely rare. Inflammatory pseudotumor typically involves the skull base and often causes painful or painless ophthalmoplegia (Tolosa-Hunt

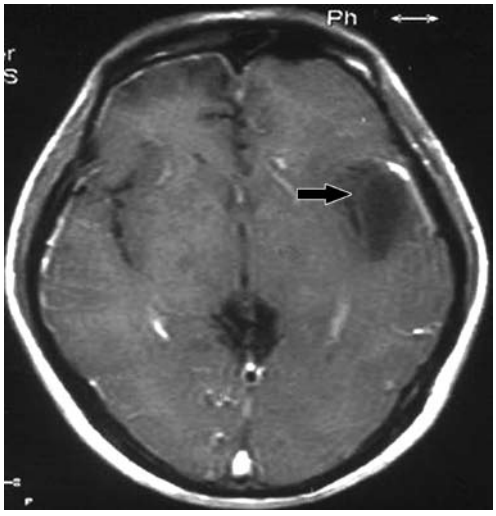

Figure 1 - Contrast-enhanced MRI showed a lesion (arrow) hypointense to gray matter on T1-weighted images and hyperintense on T2-weighted images with homogenous and avid contrast enhancement partially encasing the cerebral dura mater in the polus temporalis.

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syndrome). In general, inflammatory pseudotumors are isointense to gray matter on T1-weighted images, and hypointense on T2-weighted images with homogenous and avid contrast enhancement, and tend to radiologically mimic expansible invasive malignant tumors.[3,4] We report a rare case involving brain parenchyma in the left temporal lobe, showing a lesion hypointense to gray matter on T1-weighted images and hyperintense on T2-weighted images with homogenous and avid contrast enhancement under MRI. It also partially encased the cerebral dura mater in the polus temporalis with a slight space-occupying effect. A preoperative diagnosis of glioma was made based on these findings at first. Two months after the surgery, the patient started to feel headaches again with complaints of vision diminution and painless ophthalmoplegia (Tolosa-Hunt syndrome) of the left eye. An MRI revealed an extensive meningeal thickening of the skull base and cavernous sinus. The lesion developed along the cerebral dura mater and reached the optic nerve sheaths and tentorium cerebelli, which was clinically and radiologically in accordance with typical previous reports. We, thus, radiologically present a process in which the inflammatory pseudotumor metastasized along the cerebral dura mater.

The characteristics of inflammatory pseudotumor for diagnosis by histopathology and immunohistochemistry include hyperplasia of abundant fibrous stroma and infiltration of inflammatory cells such as lymphocytes and plasma cells. Plasma cells are frequently predominant, but they may be outnumbered by lymphocytes in some cases, which may histologically mimic lymphomas and lead to excessive treatment. In our case, the patient had a decompressive craniectomy due to the signs of non-Hodgkin's lymphoma, which was revealed by intraoperative frozen-section examination. Steroids are the mainstay in management of inflammatory pseudotumors. Most reported cases responded quickly to steroid therapy, but when the lesion has a large fibrosis component, indicating a more chronic process, steroids are less effective.[2] Because intracranial inflammatory pseudotumor commonly involves the skull base and cavernous sinus, surgeries are often planned for biopsy. A complete surgical resection, if possible, is still the treatment of choice for inflammatory pseudotumors occurring in non-functional areas. We completed a total excision of the temporal lesion, but steroids were not given within 2 months after surgery. The patient's headaches recurred together with a new symptom of vision diminution of the left eye. A subsequent MRI revealed a lesion involving the skull base and cavernous sinus. The patient was then treated with steroids, and the MRI showed diminution of the lesion 3 months after surgery. He was doing well by the end of the tenth month post surgery, which suggests that steroids are still necessary after completely excising the inflammatory pseudotumor. Previous case reports have also shown that radiation treatment and chemotherapeutic medication are effective for inflammatory pseudotumor. The recurrence rate after treatment is difficult to assess for intracranial inflammatory pseudotumors, because most publications consist of a single case. We found only one case reporting intracranial inflammatory pseudotumor located in the skull base that recurred after steroid treatment.[5] The lesion in our case recurred 2 months after surgery and developed along the cerebral dura mater.

In conclusion, intracranial inflammatory pseudotumor is exceptional and behaves in a multiplex fashion. It can radiologically and histologically mimic a malignant neoplasm, such as glioma and lymphoma. Distinguishing an inflammatory pseudotumor from a brain tumor is very important in avoiding misdiagnosis and overzealous treatment. Lesions could recur after surgery and develop along the cerebral dura mater. Steroid treatment is still necessary even after total resection.

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