Inflammatory Pseudotumor of the Skull Base Involving Fissura Petrooccipitalis: A Rare Case with Challenging Diagnosis

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ABSTRACT

Inflammatory pseudotumor (IPT) is a benign entity that may present as a solid mass mimicking a malignant neoplasm. Histologically, they are composed of varying proportions of myofibroblastic spindle cells, lymphocytes, and plasma cells. Skull base IPT is rare and usually occurs in adults with no sex predilection. The skull base IPT typically presents with headache, and/or cranial nerve palsy. There is no consensus regarding treatment of skull base IPT due to its rarity. Surgical resection and corticosteroid therapy have generally been used. The preoperative diagnosis of skull base IPT is usually difficult due to its nonspecific clinical and radiologic features. We report a case of a 42-year-old woman with IPT originating from the skull base. To our knowledge, this is the first case of IPT invading fissura petrooccipitalis. When a mass in the skull base appears hypointense on T2-weighted imaging with aggressive growth and bony destruction, IPT should be considered in the differential diagnosis.

KEYWORDS: Inflammatory pseudotumor, skull base, magnetic resonance imaging, diagnosis

Inflammatory pseudotumor (IPT) is a benign entity that can mimic a malignant neoplasm. It has been described in both genders, at all ages, and in nearly every anatomic site.1 In the head and neck region, it is uncommon but most often affects the orbit. Skull base IPT is rare and usually pursues an aggressive course indistinguishable from malignant tumor.2 The lesion may involve adjacent tracts or cranial nerves. Most reports of skull base IPT have included only a single case or a small case series. Therefore, little is known concerning the natural history of this lesion. We present a rare case of IPT originating from the skull base, extending intracranially via the fissura petrooccipitalis. To our knowledge, this is the first case of IPT invading the fissura petrooccipitalis. The radiographic presentations and clinical features are discussed. The literatures regarding pathologic mechanism, preoperative diagnosis, and treatment of skull base IPT are reviewed.

CASE REPORT
A 42-year-old woman with a history of left-sided headaches presented with new-onset left facial palsy and diplopia. For nearly 2 years before admission, the patient had complained of an idiopathic left occipital headache...
which could be relieved with Carbamazepine. She de-
nied any history of associated fever, nausea, vomiting, or
weight loss. A physical examination demonstrated left
binocular horizontal diplopia as well as loss of sensation
to her left face to light touch. Her cognitive function and
tendon reflexes were normal. Epstein-Barr virus (EBV)
capsid antigen IgG titers were 1:160, however, other
laboratory investigations were within normal limits.

A CT scan revealed a soft-tissue density mass in
the left skull base with enlargement of left fissura
petrooccipitalis (Fig. 1). The lesion had also invaded
the left clivus and left petrous apex. Magnetic resonance
imaging (MRI) of her brain and nasopharynx was
performed using a 1.5 Tesla scanner. MRI disclosed an
irregular nodular mass in the same region. The mass
measured 59 mm × 26 mm × 31 mm in size. It was
hypointense compared with cerebral gray matter on
T2-weighted images (Fig. 2A) and isointense on T1-
weighted images (Fig. 2B). Marrow replacement of the
left clivus was noted on the T1-weighted images. The
tumor exhibited moderately uniform enhancement
(Fig. 2C). On postcontrast T1-weighted imaging, the
tumor extended intracranially superiorly via an enlarged
left fissura petrooccipitalis. The adjacent dura were
thickened and enhanced (Fig. 2D). The bilateral cav-
ernous sinuses were not involved by the lesion. The
nasopharyngeal mucosa was not thickened.

The patient underwent partial resection of the
lesion via the nasopharyngeal cavity. The lesion was firm
with limited vascularity. On routine HE-stained sec-
tions, the lesion was composed of spindle cells in a
collagenous background of abundant lymphocytes,
plasma cells with varying admixtures of collagen bundles
(Fig. 3). Mitoses were very rare. The final pathologic
diagnosis was inflammatory pseudotumor. The patient
had no complications and was discharged on oral corti-
costeroids. Her symptoms of facial palsy and diplopia
resolved completely 2 weeks after operation, and the
mild residual pain disappeared after 1 month on cortico-
steroid treatment. The low-dose steroid maintenance
therapy with prednisolone 10 mg daily was continued for
10 weeks. Over 4 months after the operation, the patient
was symptom-free and the follow-up MRI showed no
sign of recurrence. Dural thickening and contrast en-
hancement was decreased.

DISCUSSION

IPT has been reported to occur most often in the lung,
liver, orbit, mesentery, retroperitoneum, genitourinary
tract, and upper respiratory tract. In the head and neck,
IPT most commonly involves the orbit. Skull base
involvement is rare.2,3 Skull base IPT usually occurs in
adults with no sex predilection. Systemic symptoms such
as fever or weight loss are uncommon in skull base IPT,
but may occur in IPTs affecting other sites. The skull
base IPT typically presents with headache, and/or cranial
nerve palsy.4 Our patient presented with left-sided head-
aches and facial nerve palsies as the lesion was located in
the left middle cranial fossa and had invaded the petrous
bone and clivus. Cranial nerve palsies are most often due
to the aggressive, invasive nature of skull base IPT.

The pathologic mechanism of IPT is still con-
troversial. It is frequently associated with fever, sweats,
fatigue, and lymphadenopathy, which suggests an infec-
tious cause. Regardless of location, all IPTs share similar
histological features. They are composed of varying
proportions of myofibroblastic spindle cells, lympho-
cytes, and plasma cells.5,6 There is an absence of neo-
plastic cells. The isolation of EBV DNA from some
cases of IPT suggests that EBV may play a role in at least
a subset of IPT.7 The microorganisms are rarely detected
in the IPTs, however. Acute immune and vascular causes
of IPT have also been hypothesized. Our patient had an
elevated EBV antigen IgG titer supporting a postinfec-
tious cause. However, the term “inflammatory myofi-
brastic tumor” has recently been suggested as a more
descriptive name for inflammatory pseudotumor. Some
authors believe this tumor is a low-grade fibrosarcoma
with inflammatory cells. In 2002, the World Health
Organization classified inflammatory myofibroblastic
tumor as a soft-tissue tumor composed of differentiated
myofibroblastic spindle cells, usually accompanied by
numerous plasma cells and/or lymphocytes.8 Some stud-
ies have revealed cytogenetic clonal abnormalities and
anaplastic lymphoma kinase expression suggesting a
neoplastic etiology.9,10

Figure 1 Noncontrast axial CT scan with bone windows
reveals marked expansion of the left fissura petrooccipitalis
(asterisk) and destruction of the left clivus (arrow) and left
petrous apex.
The preoperative diagnosis of skull base IPT is usually difficult due to its nonspecific clinical and radiologic features. Although they are histologically benign, IPTs often show aggressive clinical behavior, with locally destructive features that mimic a neoplastic process. In our case, for example, there were no signs or symptoms of infection (such as fever) and the tumor was originally thought to be a nasopharyngeal carcinoma (NPC). On CT scan, IPT frequently reveals a soft-tissue density mass with moderate enhancement. In contrast to orbital disease, skull base IPT presents with more aggressive growth and bony destruction, often accompanied by bony sclerosis. The bone sclerosis is suggestive of slow growth such as that seen in inflammatory conditions. MRI
features of skull base IPT are variable and are usually iso- to hypointense relative to cerebral gray matter on both T1- and T2-weighted images. The presence of marked T2 hypointensity, as in our case, serves as an important clue in differentiating IPT from aggressive malignancies, such as infiltrative NPC, metastatic carcinoma, primary skull base lymphoma, or plasmacytoma, which are usually iso- to hyperintense on T2-weighted images. Lu et al. reported seven patients with nasopharyngeal IPT and compared them with seven patients with NPC. All IPT lesions were hypointense to brainstem on T2-weighted images, and six of the NPC lesions showed mild hyperintensity on T2-weighted images. The remaining NPC lesion was isointense on T2-weighted images. Lee et al. described a case of IPT originating from the clivus which also showed marked heterogeneous hypointensity on T2-weighted imaging. The hypointensity seen on T2-weighted images likely reflects a combination of fibrosis and attenuated cellularity. Intracranial extension with dural thickening and enhancement were observed in our case, which may be another feature of skull base IPT. Strasnick et al. reported on two patients with temporal case, which may be another feature of skull base IPT.

Radiographic abnormalities include mass effect, dural thickening and enhancement. In our case, dural thickening and enhancement were observed in our adjacent intracranial dural structures. Recurrence during the 2 years follow-up were as high as 15% in studies of extrapulmonary IPT. Long-term administration of low-dose prednisolone is recommended to prevent relapse. But the efficacy and safety of the low-dose steroid maintenance therapy in IPT still remains questionable. Radiation therapy has been used in patients with orbital IPT who cannot tolerate or fail corticosteroid treatment, but similar success has not been demonstrated in the limited number of reported cases of skull base IPT. In our case, the patient had significant clinical and radiologic improvement following surgical resection and responded well to subsequent corticosteroid therapy.

IPT rarely involves the skull base. When it does, it frequently presents with headache, and/or cranial nerve palsy due to its aggressive growth and bony destruction and can mimic a malignant tumor, radiologically. When a mass in the skull base appears hypointense on T2-weighted imaging with associated thickening and enhancement of adjacent intracranial dural structures, IPT should be considered in the differential diagnosis. Complete surgical removal appears the first choice for therapy and subsequent steroid therapy may help to prevent recurrence.

REFERENCES