CASE REPORT

Inflammatory pseudotumors of the head and neck in pathology-proven cases

Pseudotumeurs inflammatoires de la région tête et cou dans des cas histologiquement prouvés

D.T. Ginat a,*, A. Bokhari b, S. Bhatt a, V. Dogra a

a Department of imaging sciences, university of Rochester medical center, 601, Elmwood avenue, Rochester, NY 14642, USA
b Department of pathology, university of Rochester medical center, Rochester NY 14642, USA

KEYWORDS
Inflammatory pseudotumors;
Head;
Neck;
MRI;
CT

Summary The purpose of this case series is to characterize the CT and MRI features of pathology-proven inflammatory pseudotumors in the head and neck. Our search identified three orbital, one maxillary sinus, and one skull base inflammatory pseudotumor. All of the lesions demonstrated some degree of infiltrative features on imaging. On CT, all of the orbital inflammatory pseudotumors were of homogeneous soft tissue density. One of the orbital inflammatory pseudotumors demonstrated bone erosion and two others demonstrated stranding of the orbital fat. The maxillary sinus lesion initially appeared aggressive with bone erosion and orbital invasion. Calcifications were identified in the dural inflammatory pseudotumor. Among the lesions that were given contrast during CT or MRI. All exhibited some degree of enhancement. The two pseudotumors that underwent MRI were isointense on T1 and T2, with scattered areas of low signal. The orbital inflammatory pseudotumors underwent orbitotomy. However, the maxillary sinus and skull base lesions regressed with steroid therapy. Inflammatory pseudotumors of the head and neck regions typically manifest as enhancing soft tissue masses associated with infiltrative changes. Despite their sometimes-aggressive appearance, these lesions may respond well to steroid treatment. Imaging plays an important role in diagnosing and following inflammatory pseudotumors.

© 2011 Elsevier Masson SAS. All rights reserved.

Introduction

Inflammatory pseudotumors (IPT) are benign lesions that consist of unencapsulated mass-like aggregates of myofibroblastic spindle cells and inflammatory cells, including both B and T-cell lymphocytes [1]. These lesions have been reported to occur in virtually any tissue or organ in the body, although the orbit and lungs are the most common sites [1,2]. Involvement of the head and neck is uncommon.

The clinical presentation and imaging appearances of IPT can mimic other benign conditions including meningiomas and granulomatous diseases, such as sarcoidosis [3,4]. Alternatively, IPT can appear aggressive and mimic malignancies.
such as lymphomas, fibrosarcomas, and metastases [4–6]. This poses a diagnostic and therapeutic dilemma. In this case series, the imaging features of a series of cases of IPT found in head and neck. In addition, the course of treatment and follow-up imaging is described and illustrated.

**Case series**

**Skull base**

On the initial brain lab MRI T1-weighted sequence with contrast, the intracranial IPT appeared mildly heterogeneous and infiltrative, extending along the dura from the region of the skull base to the posterior fossa along the tentorium cerebelli (Fig. 1A). The lesion encased, but did not invade the internal carotid artery. The differential diagnosis was atypical meningioma versus lymphoma and is reminiscent of Tolosa-Hunt syndrome. A diagnostic craniotomy was subsequently performed, during which a biopsy sample was obtained. Histology demonstrated chronic inflammation with fibrosis, angiomatosis, and meningeal hyperplasia.

During the operation, it was noted that the lesion was firmly adherent to the skull base and resection was not attempted. The patient was started on steroid therapy once the diagnosis of IPT was established. On follow up CT 11 months later, a residual soft-tissue mass associated with thickening and diffuse calcification of the dura was identified (Fig. 1B). On MRI the mass was of low to intermediate signal intensity on T2-weighted images (Fig. 1C) and isointense to brain on T1-weighted sequences (Fig. 1D). The mass enhanced strongly and homogeneously with Gadolinium-based contrast (Fig. 1E).

**Orbit**

Among the orbital IPT, there were two cases of myositis and one case of dacryoadenitis. The myositis type IPT were limited to a single intraocular muscle: either the lateral rectus or the medial rectus muscles. The lesion involving the medial rectus muscle was associated with mild stranding of the orbit fat (Fig. 2A) and showed homogeneous enhancement (Fig. 2B).

The lateral rectus IPT demonstrated fusiform enlargement of the muscle and the tendinous insertion at the globe (Fig. 3A). In addition, a small portion of the lateral rectus muscle protruded into the maxillary sinus via a defect in the orbital floor, which was otherwise thickened and sclerotic (Fig. 3B). There was a mild degree of orbital fat stranding and infiltrative changes apparent in CT.

The case of dacryoadenitis demonstrated homogeneous enlargement of the lacrimal gland (Fig. 4A), and was associated with mild inflammatory changes and enhancement (Fig. 4B). There was no bony destruction observed in this case. All three case of orbital IPT were treated via surgical resection for immediate relief of visual symptoms.

**Maxillary sinus**

The maxillary sinus IPT initially manifested as extensive thickening of the sinus mucosa associated with aggressive features, including bone erosions and orbital, ethmoid sinus, and frontal sinus invasion (Fig. 5A). The mass was heterogeneous with scattered areas of low signal intensity interspersed throughout intermediate to slightly hyperintense signal on T2-weighted sequences (Fig. 5B) and intermediate signal on T1-weighted images (Fig. 5C). In addition, the lesion demonstrated avid, but somewhat heterogeneous enhancement on MRI (Fig. 5D). Following steroid treatment, the mass almost completely regressed at follow-up eight months later, leaving behind areas of bony destruction. A follow-up MRI demonstrated persistent enhancement of the residual lesion.

**Discussion**

In the head and neck, IPT most commonly occur in the orbit [7]. In contrast, extraorbital inflammatory pseudotumors of the head and neck are rare, but have been reported in several locations, such as the nasal cavity, nasopharynx, paranasal sinuses [8], parapharyngeal space [9], larynx [10], salivary glands [11,12], skull base [4,13], choroid plexus [14], facial nerves [15], brain [16], dura [17], and temporal bone [4]. Imaging with CT and MRI plays an important role in diagnosing these lesions, particularly for asymptomatic cases. In general, the imaging appearances of IPT vary with location. For example, orbital IPT are most often well defined without associated lysis, which mimic benign tumors. On the other hand, IPT of the sinus, nasopharynx and deep spaces, are often infiltrative masses with erosions or lysis, mimicking malignant tumors that require histological proof to exclude malignancy. IPT of the dura, including Tolosa-Hunt syndrome, are also infiltrative lesions, but typically lack the adjacent destructive bone changes.

Several types of orbital IPT have been described, including dacryoadenitis, myositis, and sclerouveitis [2,7]. The lesions typically present with acute pain and exophthalmos, which can help differentiate them from true neoplasms [2,7]. As noted in this series of cases, orbital IPT are usually solitary. Orbital IPT generally consist of focal soft-tissue masses of variable sizes and shapes. These lesions are commonly associated with inflammation and edema of the surrounding tissues [16], although this was noted in only one of the three orbital IPT in our series. In the myositis type, both tendons and muscle can enlarge, creating a tubular appearance. This is a distinguishing feature from thyroid opthalmopathy, in which the tendons are typically spared [7].

Although orbital IPT can extend through fissures and cause bony erosion, the presence of sclerosis favors an indolent, benign process [16]. Nevertheless, there are occasional reports of more aggressive behavior, such as intracranial extension as well as associated cranial nerve and meningeal enhancement [17]. Despite such features, response to steroid treatment is favorable [6]. Orbital IPT classically demonstrate low to intermediate signal on T1 and T2-weighted sequences. These lesions also exhibit variable enhancement on CT and strong homogeneous enhancement on MRI [7,16]. The differential diagnosis for IPT of the orbit is broad and is ultimately a diagnosis of exclusion [2,16].

Unlike the orbital counterpart, maxillary sinus IPT commonly displays aggressive features, such as infiltrative mar-
A 45-year-old male who presented with long-standing headache and was diagnosed with a dural IPT. A. Axial contrast-enhanced T1-weighted image shows extensive dural thickening extending from the cavernous sinus region to the tentorium cerebelli (arrows). The mass encases the internal carotid artery (circle). B. Axial CT obtained 11 months later demonstrates a residual partially calcified, ill-defined mass (arrow). Note the interval right temporal craniotomy. C. Axial T2-weighted MRI obtained at about the same time as the CT shows that the residual lesion is heterogeneously low signal intensity (arrow). D. The corresponding T1-weighted image shows that the lesion is nearly isointense to the brain with some areas of slightly lower signal intensity (arrow). E. Despite steroid treatment and considerable decrease in size, the residual IPT still demonstrates avid contrast enhancement.

gins, osseous destruction, and invasion into the orbit [6,8]. Other features include a soft-tissue mass with mild enhancement on CT and intermediate signal intensity on both T1- and T2- weighted MR images [8]. Prognosis is favorable following surgical excision [16]. Intracranial IPT can involve the dura, brain, and even less frequently both [16,17]. The case of IPT involving the dura and cavernous sinus is compatible with Tolosa-Hunt syndrome and the differential
Inflammatory pseudotumors of the head and neck in pathology-proven cases

Figure 2  An 11-year-old male with myositis type orbital IPT. Pre- (A) and post-contrast (B) axial T1-weighted MR images show marked enlargement of the medial rectus muscle (*), which enhances mildly and homogeneously. There is mild stranding of the orbital fat adjacent to the lesion.

Figure 3  A 89-year-old female with myositis type orbital IPT. A. Postcontrast axial CT image demonstrates marked enlargement of the lateral rectus muscle belly (*). There is also thickening of the tendon (arrow). B. Coronal CT shows extension of the IPT through a defect in the orbital floor (*). There is also thickening of the orbital floor elsewhere (arrow). Mild infiltrative changes are also apparent.

diagnosis includes meningioma, sarcoidosis, Wegener’s granulomatosis, tuberculoma, and lymphoma. The presence of calcifications is often seen in meningiomas, but is unusual for IPT and may be related to the specific location and longstanding nature of this particular lesion.

The MRI signal characteristics of the skull base IPT were similar to those found in IPT elsewhere, with low T2-weighted signal intensity, intermediate T1-weighted signal intensity and strong enhancement. The low signal intensity on T2-weighted MRI appears to be related to the lack of mobile protons in the underlying fibrotic tissue component [13]. Furthermore, the degree of fibrosis is inversely related to the degree of inflammatory reaction, such that the more fibrotic lesions demonstrate less inflammation [13]. There are no calcifications on CT in the areas corresponding to low T2 signal intensity on MRI. The presence of T2 hypointensity without visible calcifications can also be observed in postsurgical scar tissue as well as malignant neoplasms with

Figure 4  A 31-year-old female with left orbital dacryoadenitis IPT. A. Non-contrast axial CT image shows diffuse enlargement of the left lacrimal gland (*). B. Following contrast administration, there is heterogeneous enhancement. There are also inflammatory changes in the surrounding subcutaneous tissues (arrows).
high nuclear-to-cytoplasmic ratios and absence of serous and mucinous contents.

The MR perfusion, MR diffusion-tensor imaging, and MR spectroscopy features of IPT have been described in very few cases [17,18]. Intracranial IPT demonstrate decreased N-acetylaspartate (NAA) and increased choline (Cho) peaks. The decreased NAA levels are related to the substitution of normal neural elements with plasma cell proliferation, while increased Cho levels represent increased cellularity and cell membrane synthesis due to plasma cell hyperplasia. Thus, there is overlap with typical MR spectroscopy findings in true neoplasms. However, elevated lipid lactate peaks can also found in IPT, which is attributed to the lack of vascularity [17,18]. Hypoperfusion is also apparent on perfusion MRI, which manifests as decreased relative cerebral blood volume (rCBV). Similarly, contrast kinetic curves in a case of atypical plasma cell granuloma showed moderate to rapid enhancement and lack of washout, which differs from criteria for malignancy [17]. Diffusion tensor imaging can depict infiltration and destruction of white matter tracts, resulting in reduction or total loss in fractional anisotropy [17]. Significant differences in the diffusion weighted MRI properties have been reported among orbital IPT, cellulitis, and lymphoid lesions [19]. Lymphoid lesions are more hyperintense than IPT and IPT are brighter than cellulitis.

While advanced MRI features can to certain extent help delineate the non-neoplastic nature of IPT, ultimately conventional MRI accurately depicted the degree of invasiveness and helped guide the surgeon appropriately. A difficult and potentially complicated resection was averted and the procedure was limited to obtaining a tissue diagnosis. Instead, steroid therapy was successfully initiated.

Orbital IPT generally respond well to steroid therapy, with prompt involution of the lesion on follow-up imaging. However, there are limited data regarding the role of MRI in evaluating treatment response in extra-orbital IPT of the head and neck. Recurrence of symptoms is common after cessation of steroid therapy for skull base IPT, which parallels reactivation of disease [20]. Similarly, most skull base IPT treated with radiation therapy develop symptomatic recurrence, which manifests as residual disease on MRI [20]. New lesions may also arise in remote locations, such as the facial region, after radiation therapy. In contrast, the maxillary sinus IPT in this series showed considerable decrease in size with steroid therapy. However, there was persistent enhancement after the treatment. Thus, the degree of enhancement is not a reliable indicator of treatment response. Investigation of the role of advanced imaging modalities for evaluating treatment response is warranted.

In summary, head and neck IPT are rare lesions that can pose diagnostic and therapeutic challenges. Nevertheless, our case series illustrates that CT and MRI serve important roles in identifying these lesions and monitoring their response to treatment.

**Figure 5** A 41-year-old male with right maxillary sinus IPT. A. The initial coronal CT shows an ill-defined soft-tissue mass arising from the right maxillary sinus mucosa that has eroded the right conchae and inferior and medial orbital walls with extension into the orbit (arrows). B. Coronal T2-weighted MRI obtained several days later shows heterogeneous thickening of the right maxillary sinus mucosa and extension into the orbit. C. The lesion is of intermediate signal intensity on the corresponding T1-weight coronal sequence. D. Following contrast administration, there is strong heterogeneous enhancement.
Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References