CASE REPORT

# Inflammatory Pseudotumor of the Nasopharynx: An Unusual Diagnosis of a Destructive Skull Base Lesion

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## Abstract

**Aim:** To describe a rare case of nasopharyngeal inflammatory pseudotumor (IPT) with skull base invasion.

**Background:** Inflammatory pseudotumor is a nonneoplastic lesion characterized by fibrosis and inflammation. It can be locally aggressive and resemble malignancy and extranodal lymphoma. It has been described in nearly every anatomic site throughout the body. Although rare, the involvement of the skull base is associated with more aggressive behavior.

**Case description:** The authors present the case of a 63-year-old male with a 3-month history of right otorrhea, headache, and facial pain in the sensitive territory of the right trigeminal nerve (V3). Magnetic resonance imaging (MRI) showed an infiltrative lesion of the posterior wall of the nasopharynx with poorly defined limits, extended deep into the retropharyngeal and prevertebral space, with bone erosion of the occipital clivus and petrous apex. The patient performed three biopsies of the nasopharynx suggestive of a fibroinflammatory process. Inflammatory pseudotumor was diagnosed and the patient did a long course of high-dose systemic corticotherapy with good clinical and radiological response.

**Conclusion:** Biopsy is essential for the diagnosis of IPT, although it usually just shows inflammatory changes. Magnetic resonance imaging features of this entity are ill-defined lesions with local infiltration, hypointensity on T2-weighted sequences, and weak contrast enhancement, usually with an absence of cervical lymphadenopathies. The treatment of IPT involving the skull base has been controversial but can include corticotherapy and radiotherapy.

**Clinical significance:** This case report highlights the relevance of including IPT in the differential diagnosis of infiltrative nasopharyngeal and skull base lesions, especially when biopsies and cultures are negative.

**Keywords:** Case report, Inflammatory pseudotumor, Nasopharynx, Skull base.

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## Background

Inflammatory pseudotumor (IPT) is a nonneoplastic fibroinflammatory lesion with typical findings of acute and chronic inflammatory cells with different types of fibrous responses.[1] As its pathogenesis is not yet well established, different theories include an autoimmune response to a bacterial or viral infection, an abnormal production of fibrogenic cytokines, or a primary infectious response. It has been described in nearly every anatomic site throughout the body.[2] This fibroinflammatory response can be locally aggressive and simulate malignancy or extranodal lymphoma.[3]

## Case Description

The authors present the case of a 63-year-old male, with a personal history of non-Hodgkin lymphoma in remission, treated 12 years before with surgery and chemotherapy [cyclophosphamide (cytoxan), doxorubicin (adriamycin), vincristine (oncovin) and prednisone (CHOP) regimen]; diabetes mellitus type 2; chronic renal disease stage 5 under hemodialysis after the failure of renal transplant; paroxysmal atrial fibrillation; hypertension and ischemic cardiopathy with coronary stenting.

He had a 3-month history of right otorrhea, headache, and facial pain in the sensitive territory of the right trigeminal nerve (V3). The headache and facial pain increased progressively despite oral medication, and he was admitted for pain control and investigation.

Otomicroscopy showed otorrhea and signs of suppurated right otitis media and the nasofibrolaryngoscopy revealed hypertrophy of the tissue of the posterior wall of the nasopharynx, obstructing partially both of the torus tubarius (Fig. 1).

A computerized tomography (CT) scan revealed bilateral swelling of the nasopharyngeal mucosa, with ill-defined limits and erosions from the adjacent region of the skull base, on the right, to the petrous apex. Bilateral signs of mastoiditis probably related to tubal involvement due to nasopharyngeal injury and no pathological cervical adenopathies were identified (Fig. 2). Magnetic resonance imaging (MRI) showed an infiltrative lesion with poorly defined limits, in the submucosal space of the posterior wall of the median and bilateral paramedian nasopharynx, with water

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**Conflict of interest:** None

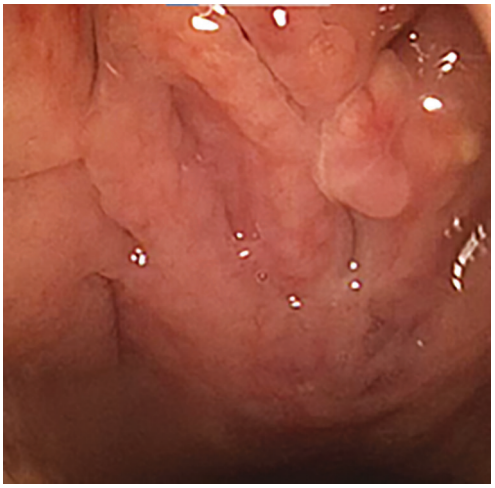

**Fig. 1:** Hypertrophy of the lymphoid tissue of the posterior wall of the nasopharynx

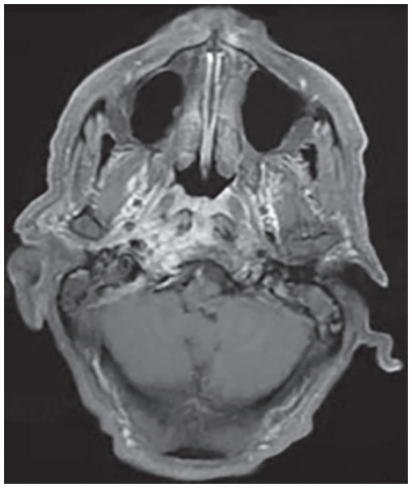

**Fig. 3:** Magnetic resonance imaging (MRI) T1 with gadolinium showing an ill-defined lesion with contrast enhancement of the nasopharynx

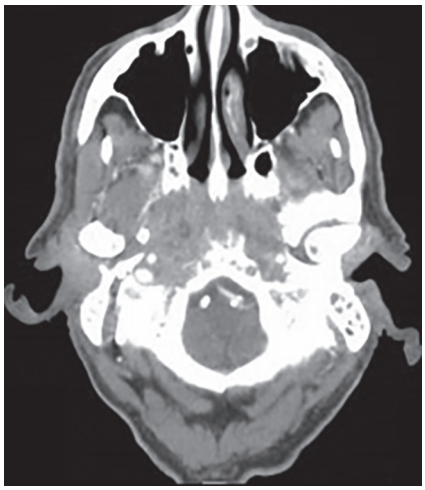

**Fig. 2:** Computerized tomography (CT) scan showing infiltrative lesion of the nasopharynx with ill-defined limits and erosions from the adjacent region of the skull base

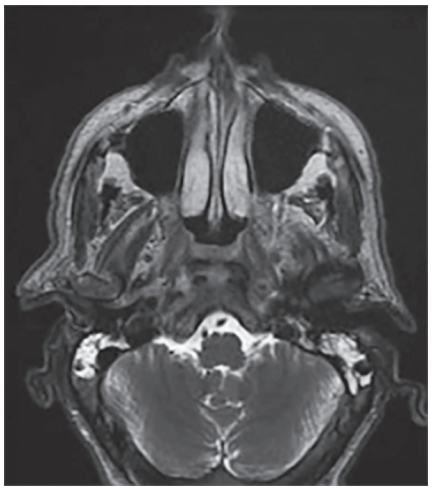

**Fig. 4:** Magnetic resonance imaging (MRI) 2 sequence showing a hypointense sign of the nasopharyngeal lesion (characteristic of IPT)

restriction and contrast impregnation. There was obliteration of the tubal and Rosenmüller recesses, with signs of bilateral otitis media. The lesion extended deep into the retropharyngeal and prevertebral space, with hypersignal bone and erosions from the right clivus reaching the petrous apex to the region of the petrous apex (Figs 3 and 4).

The laboratory workup including tumor markers [carcino-embryonic antigen (CEA); CA 19.9; CA 15.3; CA 125; neuron-specific enolase; α-fetoprotein; prostate specific antigen (PSA) and β-human-derived chorionic gonadotropin hormone (βHCG)] and autoimmune panel was negative.

A first nasopharynx endoscopic biopsy was done under local anesthesia showing only lymphoid hyperplasia. The patient was submitted to a second biopsy under general anesthesia also negative for neoplasia. The microbiologic examination was negative on the two samples.

An 18F-fluorodeoxyglucose positron emission tomography (PET) CT was performed with evidence of a hypermetabolic focus on the upper region of the nasopharynx [standardized uptake value ($SUV_{max}$ = 10.8)], suggestive of malignancy and a second focus inferior to the temporal bone, along the styloid apophysis ($SUV_{max}$ = 12.8), also suggestive of a malignant etiology, with probable adjacent bone invasion.

Due to his history of non-Hodgkin lymphoma, a medullary biopsy and a myelogram were done, both showing no signs of recurrence.

The patient was submitted to a third nasopharynx biopsy with endoscopic control under general anesthesia and the pathologic examination showed an abundant chronic inflammatory infiltrate, granulation tissue, areas of fibrosis, and hemorrhage suggestive of a fibroinflammatory process. The immunohistochemical examination was negative for neoplasia.

The diagnosis of IPT was presumed and the patient began a long course of oral systemic corticotherapy, initially with 60 mg/day of prednisolone for 1 month, reduced to 40 mg/day for another 2 months. The patient improved the symptoms of the neuropathic trigeminal pain and had no more episodes of otitis. Magnetic resonance imaging was repeated after 3 months of corticotherapy and revealed a volume reduction greater than 80% of the infiltrative lesion of the nasopharynx and a complete resolution of the signs of bilateral mastoiditis.

## Discussion

Inflammatory pseudotumor of the head and neck can occur more frequently in the orbit but also in a minority of cases in other anatomical sites such as the nasopharynx, nasal cavity, maxillary sinus, larynx, and trachea. Perineural spreading through maxillary (V2), mandibular (V3), and hypoglossal nerves has also been reported.[4]

Due to this aggressive behavior, biopsies are essential during the diagnosis. Multiple biopsies (open or endoscopic, under local or general anesthesia) and cultures will be negative for malignant disease and infection, typically revealing inflammatory reactions.[5]

Magnetic resonance imaging features usually show ill-defined lesions with local infiltration, hypointensity on T2-weighted sequences, weak contrast enhancement, and a favorable response to systemic steroid therapy, as was the case of our patient.[6,7]

Another important negative imaging finding is an absence of cervical lymphadenopathies, regardless of the presence of a local destructive lesion, which was also in favor of IPT.

The treatment of IPT involving the skull base has been debatable. Lee DK, et al studied the effect of corticotherapy and radiotherapy in patients with skull base IPT and found that because of his aggressive clinical features with high recurrence rates, high-dose radiation therapy could be an initial approach. Systemic steroids could be added, if the patient's general condition allows, to pain control and to decrease cranial nerve manifestations.[1] In the present case, the remission of the symptoms and the significant reduction of the lesion were achieved with systemic corticotherapy.

### Clinical Significance

This case report highlights the relevance of entailing IPT in the differential diagnosis of infiltrative nasopharyngeal and skull base lesions, especially when biopsies and cultures are negative and there is evidence of an aggressive lesion. Another important sign that should be aware of this entity is the absence of pathologic cervical adenopathies despite aggressive lesions of the nasopharynx and skull base.

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## References

1. Lee DK, Cho YS, Hong SH, et al. Inflammatory pseudotumor involving the skull base: Response to steroid and radiation therapy. Otolaryngol Head Neck Surg 2006;135(1):144–148. DOI: 10.1016/j.otohns.2006.01.016.
2. Gadde J, Franck B, Liu X, et al. Inflammatory pseudotumor of the nasopharynx with spread along the trigeminal nerve. Am J Otolaryngol 2013;34(3):252–254. DOI: 10.1016/j.amjoto.2012.11.013.
3. Cho KS, Kim HJ, Lee CH, et al. Nasopharyngeal inflammatory pseudotumor showing abducens nerve palsy. Auris Nasus Larynx 2011;38(4):543–546. DOI: 10.1016/j.anl.2010.12.005.
4. De Vuysere S, Hermans R, Sciot R, et al. Extraorbital inflammatory pseudotumor of the head and neck: CT and MR findings in three patients. AJNR Am J Neuroradiol 1999;20(6):1133–1139. PMID: 10445457.
5. Chwang WB, Jain R, Narayan A, et al. Inflammatory pseudotumor of the nasopharynx and skull base: Mimicking an aggressive neoplasm or infection. Arch Otolaryngol Head Neck Surg 2012;138(8):765–769. DOI: 10.1001/archoto.2012.1540.
6. Choi SY, Yu IK, Han MH, et al. Fibrosing inflammatory pseudotumor of the nasopharynx: MR features and histopathologic correlation. Eur J Radiol 2009;72(2):274–277. DOI: 10.1016/j.ejrad.2008.07.026.
7. Lu CH, Yang CY, Wang CP, et al. Imaging of nasopharyngeal inflammatory pseudotumours: Differential from nasopharyngeal carcinoma. Br J Radiol 2010;83(985):8–16. DOI: 10.1259/bjr/98400347.