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Inflammatory Pseudotumor of Nasopharynx: The Great Mimicry

Abstract – Inflammatory pseudotumor (IPT) of the nasopharynx is a rare differential diagnosis for nasopharyngeal masses, often mimicking nasopharyngeal carcinoma both clinically and radiologically. We report a case of a 40-year-old male who presented with a one-year history of chronic, throbbing headache followed by a six-month history of spontaneous bilateral epistaxis, which had increased in frequency. Nasal endoscopy showed a midline mass over nasopharynx extending to the right fossa of Rosenmuller. Contrast-enhanced CT paranasal sinuses show an ill-defined, heterogeneous enhancing mass within nasopharyngeal mucosal space with evidence of base of skull erosion. Multiple biopsies of the nasopharyngeal mass revealed inflamed granulation tissue with no evidence of malignancy, and tissue cultures were negative for microbial growth. The patient was treated as IPT as a diagnosis of exclusion, receiving high-dose corticosteroids for a total of six weeks, resulting in a significant clinical improvement. Given its aggressive behavior, which mimics malignancy both clinically and radiologically, IPT should be considered as one of the differentials, despite its rare occurrence.

Keywords – *Inflammatory pseudotumor, nasopharynx, head and neck, benign*

1 INTRODUCTION

Inflammatory pseudotumor (IPT) is a benign, non-neoplastic inflammatory condition [1,2] that often mimics malignant neoplasms [2] due to its locally aggressive behavior which may present as a rapidly enlarging mass with contact bleeding and expansile features capable of eroding adjacent bony structures. It has been reported across all age groups with no gender predilection and can occur in almost any anatomical location [2], with liver and lungs being the most commonly affected sites [3].

IPT occurring over the head and neck region is relatively rare, accounting for less than 5% of all reported cases, with the orbit being the most commonly affected site [4]. Extraorbital involvement, particularly in the nasopharynx and skull base, is uncommon, with less than 50 cases reported to date. Due to its clinical and radiological resemblance to malignancy, IPT poses a diagnostic dilemma which often leads to delays or errors in management [1,5].

2 CASE PRESENTATION

A 40-year-old male who is a chronic smoker with underlying dyslipidemia presented with a one-year history of chronic, throbbing headache, followed by multiple episodes of spontaneous bilateral epistaxis over the past six months, increasing in frequency up to four episodes of epistaxis per week. He denied any constitutional symptoms such as weight loss, loss of appetite and denied any neck swelling. He also denied any otological complaints such as ear fullness, recurrent otitis media, or any neurological symptoms including diplopia, blurry vision, or eye pain, numbness over the facial region or facial asymmetry.

Nasal endoscopy shows a midline mass over nasopharynx extending into the right fossa of Rossemuller (FOR) (Figure 1a), therefore a biopsy was taken for tissue diagnosis. A contrast-enhanced CT paranasal sinuses (CECT PNS) showed an ill-defined heterogeneous enhancing mass within nasopharyngeal mucosal space, measuring 2.9 x 4.0 x 3.1 cm, obliterating the right FOR.

Superiorly, mass eroded the floor of bilateral sphenoid sinuses, extending into both sinuses and erosion of clivus. Anteriorly, the erosion involves the vomer and extension of mass to the choanae and into left posterior nasal cavity and widening of left pterygopalatine fossa suspicious of extension. No intracranial, intraspinal extension, no enlargement of cervical lymph nodes were visualized in this imaging (Figure 2a).

Biopsy results revealed a severe acute-on-chronic inflammatory process predominantly neutrophils infiltration, some forming micro and macro abscesses, no malignant cells see (Figure 3). Hence, a repeat biopsy was done and similar findings were yielded. Special stains revealed no acid-fast bacilli and were negative for fungal elements on Periodic acid-Schiff (PAS) and Grocott's Methenamine Silver (GMS) staining.

During the follow-up, nasal endoscopy shows a midline nasopharyngeal mass increasing in size, extending to the left sphenoid sinus and bleeding on contact. A malignant process remained highly suspected based on clinical and radiological findings, including progressive increase in mass size, contact bleeding, and evidence of bony erosion on imaging. He was subjected to examination under anaesthesia, biopsy of nasopharynx mass and left sphenoid mass. The biopsied mass was sent for histopathological examination (HPE) and culture to rule out infectious causes which yielded no organism. All tissue HPE again showed inflamed granulation tissue with no evidence of malignancy (Figure 3).

A diagnosis of IPT of nasopharynx and base of skull base was made as a diagnosis of exclusion. He was started on high-dose corticosteroid therapy (HDST) for a total of 6 weeks. The initial daily dosage of prednisolone 60 mg was maintained for 2 weeks before tapering it over the next 4 weeks. A repeat nasal endoscopy post HDST showed marked improvement with total resolution of symptoms (Figure 1b). Low-dose corticosteroid maintenance therapy (LDSMT) was then continued for over 8 weeks. His symptoms completely resolved, post 3 months of steroid treatment, nasal endoscopy showed no residual mass in the nasopharynx. Repeat MRI paranasal sinuses post-treatment showed previous mass significantly reduced in size with minimal residual lesion at roof of nasopharynx, at the site of clival erosion (Figure 2b).

The mass appeared T2 hyperintense with no restricted diffusion and homogeneously contrast enhanced, measuring 2.1 x 2.1 x 1.2 cm. Subsequent MRI paranasal sinuses as surveillance tools have shown a stable size for a total duration of 2 years since first presentation.

3 DISCUSSION

All anatomical sites have reported cases of inflammatory pseudotumor (IPT), however the head and neck accounted for less than 5% of all cases{4}. The first case of IPT over head and neck is reported over orbit, which is the most common site for IPT of head and neck. IPT of the skull base and nasopharynx is rare and has been reported as a case series of a total of 4 cases which were compared [1,6]. It is a benign, idiopathic disease with unknown etiology [6]. Various case reports proposed that autoimmune [1] and infectious diseases [2] are causes of disease [1,5,7]. However, there is a report suggesting that anaplastic lymphoma and cytogenetic clonal abnormality may raise the possibility of malignancy [2,8].

For this case, the patient was screened and negative for HIV, Hepatitis B and C. The patient denies any chronic illness such as diabetes mellitus, not in an immunocompromised state, does not have chronic infection such as chronic sinusitis or chronic ear infection. Hence, it does not support the theory of long-standing infection that might contribute to the development of IPT of the skull base [9]. Some cases proposed autoimmune as one of the risk factors for IPT, but ANA/ANCA, C3, C4 and Rheumatoid Factor were negative for this case.

IPT most commonly presented with headache [1,2], and cranial nerve palsies depending on the site of pseudotumor with eye symptoms such as painful ophthalmoplegia or abducens nerve palsy predominating [1,4,5]. This is likely due to higher probability of occurrence of IPT in orbit. For this current case, he presented with a chronic headache for the past 1 year with increasing frequency of unprovoked epistaxis.

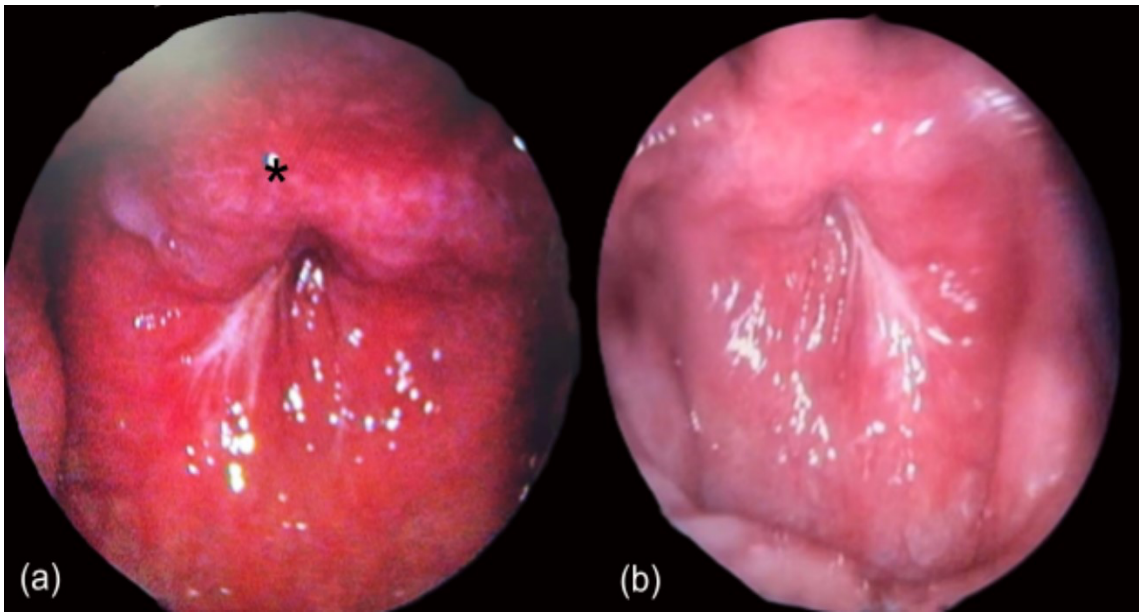

Figure 1. Nasal Endoscopy of nasopharynx. (a) Nasal endoscopy showing midline mass (asterisk) over nasopharynx with erythematous mucosa. (b) Nasal endoscopy post steroid treatment showing resolution of mass

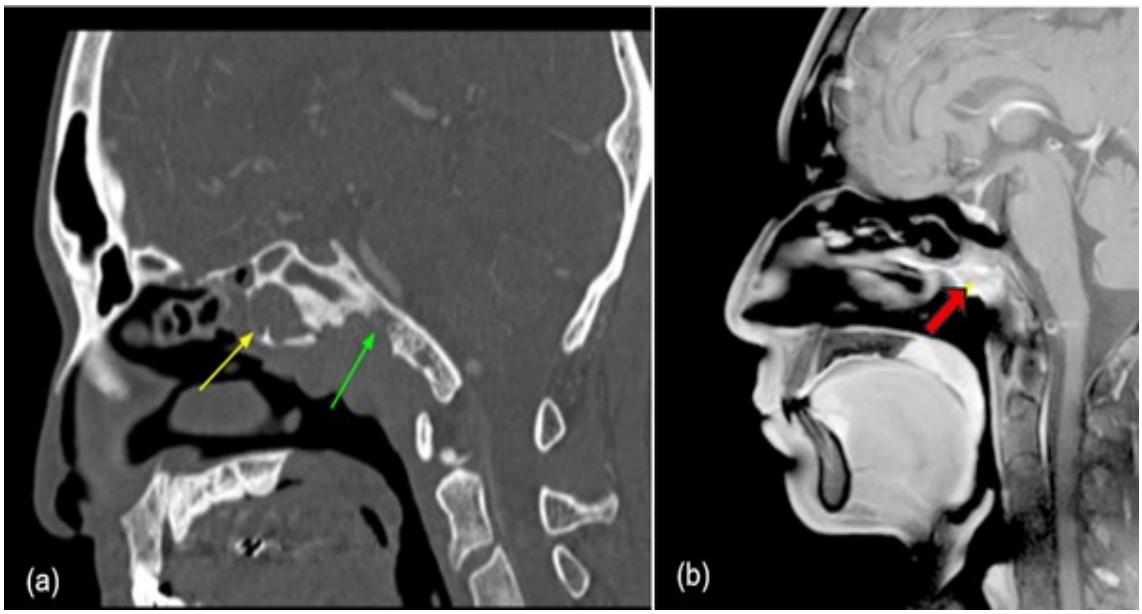

Figure 2. The imagings done pre and post treatment. (a) CECT PNS (sagittal view) shows erosion of sphenoid sinus (yellow arrow) and clivus (green arrow). (b) MRI paranasal sinuses 3 months post steroid treatment shows reducing size of nasopharyngeal mass (red arrow)

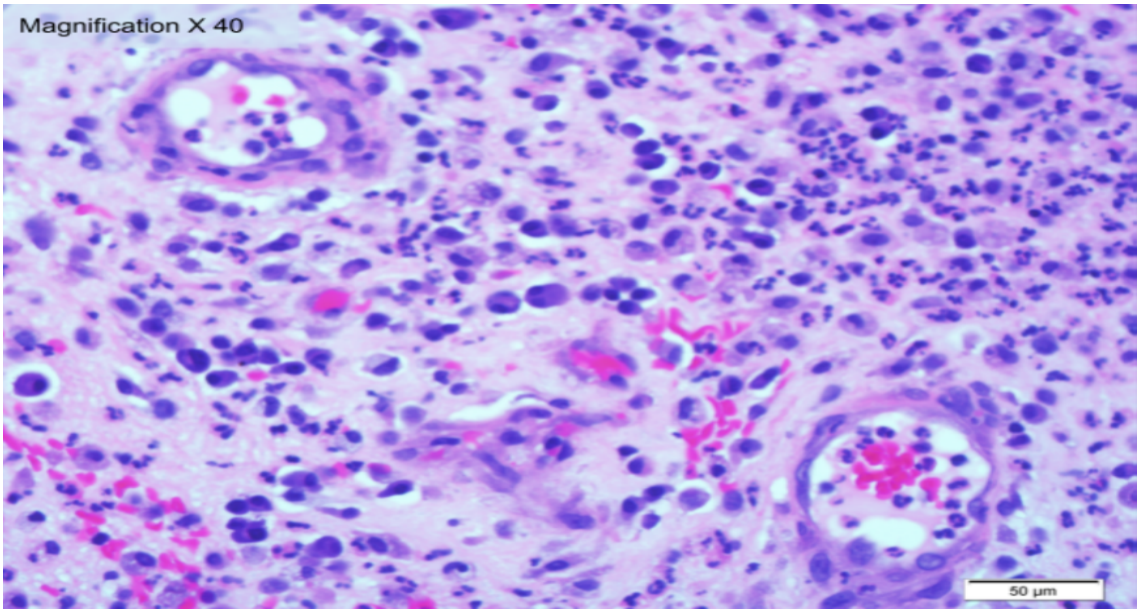

Figure 3. Hematoxylin and Eosin staining of nasal biopsies (Magnification x40) showing granulation tissue with predominance of neutrophilic infiltrates

However, he did not have any cranial nerve palsy or ophthalmoplegia which makes intracranial extension unlikely. This speculation is supported with imaging. He did not have any constitutional symptoms, no cervical lymph node involvement which is common symptoms in the case of nasopharyngeal carcinoma.

IPT involving skull base and the nasopharynx is rare, but it should be considered as one of the differential diagnoses for a nasopharyngeal mass. The diagnosis of IPT caused a significant diagnostic dilemma due to its clinical presentation of locally aggressive features [2] such as rapid growth and bleeding on contact and in some cases neurological deficit. Images show expansile masses with erosion of nearby bony structures. Accurate diagnosis therefore requires correlation of clinical, radiological, histopathological, and microbiological findings, along with repeated biopsies when initial results are inconclusive.

Nasal endoscopy is an essential modality in the evaluation and management of nasopharyngeal masses, particularly mass over the FOR. It is minimally invasive, allowing for the inspection of the nasopharyngeal mucosa during clinic visits and providing valuable information regarding the location, extent, and surface characteristics of the mass. A bedside biopsy can also be taken under endoscopic guidance. It also plays a key role in the ongoing assessment of disease progression and treatment response.

In this reported case, nasal endoscopy on first visit revealed a midline nasopharyngeal mass extending into the left FOR. Tissue biopsy is essential and mandatory to exclude malignancy and other infectious or granulomatous diseases before establishing diagnosis of IPT of nasopharynx [6]. HPE of the biopsy of the nasopharyngeal mass and sphenoid sinus showed markedly inflamed tissue with extensive ulceration and underlying granulation tissue. There was a predominant neutrophilic infiltration, with formation of micro- and macroabscesses, and focal residual lymphoid tissue in some areas. Notably, there were no multinucleated giant cells, granulomas, or malignant features observed. It then differentiated from lymphoma and granulomatous diseases.

Tissue biopsies over nasopharyngeal mass, sphenoid sinus was sent for bacterial culture to rule out infectious causes which yielded no organisms. We proceeded with special stains, including Ziehl-Neelsen, PAS, and GMS, to

exclude acid-fast bacilli, fungal infections, and other granulomatous diseases, all of which returned negative results. Additionally, serological screening for systemic infections, including HIV, Hepatitis B, and Hepatitis C, was also negative. These findings supported the diagnosis of IPT, made by exclusion.

CECT PNS was selected as the initial imaging modality due to its wider availability, shorter waiting time, and superior assessment of bony structures, despite MRI paranasal sinus and brain offering better soft tissue resolution. In this particular case, CECT PNS is crucial to evaluate bony erosion and skull base involvement.

The CECT PNS showed an ill-defined, heterogeneous enhancing mass within the nasopharyngeal mucosal space obliterating the right FOR. The lesion caused erosion over the base of the skull [2] as mentioned above but without evidence of intracranial extension, bilateral cavernous sinus or internal carotid artery involvement. These imaging findings correlate with the absence of any neurological deficits on clinical examination. Imaging findings in this case revealed a mass with evidence of adjacent bony erosion, closely mimicking a malignant process [1], thereby reinforcing the necessity of obtaining a tissue biopsy to establish diagnosis and to exclude nasopharyngeal carcinoma. Hence, we highlight the importance of multiple diagnostic modalities to establish IPT as a diagnosis of exclusion.

Corticosteroid remains the cornerstone, most commonly discussed treatment approach in IPT of the nasopharynx and skull base due to its promising outcomes [5,9]. Although certain cases were resistant to corticosteroid and needed multimodal treatment. In this case, HDST was administered and considered as a diagnostic trial for IPT. This case showed good response to steroid treatment [9], as evidenced by the rapid relief of clinical symptoms which are headache and epistaxis, and the resolution of mass in nasal endoscopy.

LDSMT was then maintained for another 8 weeks for disease control [9]. The surveillance of the response to treatment is complemented by performing an imaging post-steroid treatment. An MRI was done and the study shows minimal residual lesion at the roof of nasopharynx. Radiation therapy, surgical resection [2] and chemotherapy have been recommended in cases refractory to steroids [3]. In this case, we did not proceed with low dose radiotherapy as a prophylaxis for recurrence due to good response and an MRI paranasal sinus over a period of 2 years shows stable lesion.

4 CONCLUSION

IPT of nasopharynx is often mistaken for malignancies or infection due to its similar clinical presentation and radiological findings. A combination of suggestive clinical findings, radiological characteristics, histological evidence and thorough blood investigations may distinguish it from aggressive malignancy.

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AUTHOR CONTRIBUTIONS

SWL: Conceptualisation, literature review, writing original draft.
NACAR: Validation, review and editing. SRPJR: Validation, review and editing.
DB: Histopathological analysis and validation of histological findings, data Interpretation.
IM: Supervision, conceptualization, review and editing, correspondence.

AUTHORSHIP CLARIFICATION

Author can confirm that all relevant data are included in the article.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

Ethical approval was not required for this case report. Informed consent for participation was obtained from the patient.

CONSENT FOR PUBLICATION

All authors have read and approved the final manuscript and consent to its publication in this journal.

CONFLICTS OF INTEREST

The author declares that there is no competing of interest.

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