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Nasopharyngeal Carcinoma with Peri-Eustachian Tube Extension: A Rare Pathway of Spread

Vikash Sharma¹; Rajaram Sharma²*; Sunil Kast²; Saurabh Goyal²; Tapendra Tiwari²

¹Resident doctor, Pacific institute of medical sciences, Umarda, Udaipur, Rajasthan, India-313001. ²Assistant Professor, Pacific institute of medical sciences, Umarda, Udaipur, Rajasthan, India-313001.

*Corresponding Author(s): Rajaram Sharma Assistant Professor, Pacific institute of medical sciences,

Umarda, Udaipur, Rajasthan, India-313001. Email: hemantgalaria13@gmail.com

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Introduction

Nasopharyngeal carcinoma (NPC) is the most common primary malignancy arising from the nasopharynx's epithelium. It is commonly diagnosed in between 40 and 60 years. Females are less commonly affected than males.NPC is a classic disease with clinical behaviour, epidemiology, and histopathology different from squamous cell carcinomas in the head and neck [1].

In advanced disease, palpable neck mass and ear complaints are common in nasopharyngeal carcinoma (NPC). The vast majority of data propose Epstein–Barr virus (EBV) as the predominant pathogenic factor in NPC [2].

This case report discusses a patient with NPC presented with extensive skull base disease, lymph nodes involvement, and ear extension.

Abstract

Nasopharyngeal carcinoma (NPC) is a widespread malignant disease in endemic regions such as South Asia. Palpable nodal metastasis in the neck or involvement of adjacent structures and ear complaints are common presentations.

A 53 years old male presented with six months history of progressive unilateral face pain and a history of ear discharge for one month. Ultrasound examination of the neck showed multiple bilaterally enlarged lymph nodes. Magnetic resonance imaging (MRI) scan depicted heterogeneous mass in the right side of the nasopharynx. Nasopharyngeal histopathology revealed it to be a non-keratinizing squamous cell carcinoma. The patient underwent radiotherapy for disease management. MRI helps detect disease in the early stage as well as for precise extension of the disease.

Case presentation

A previously healthy 53-year-old man presented to our hospital with ear discharge and pain on the right side of the face that started six months before this visit. There were associated hearing impairment and fullness in the bilateral ear, but no balance disorders. Nasal blockage and postnasal drip were experienced sometimes, but there were no nasal discharge and epistaxis. There was no history of injury or any neurological deficit. Physical examination of the patient revealed palpable lymph nodes in bilateral neck levels. He had a 24 pack/year smoking history but was otherwise absent known cancer risk factors.

Investigations

The patient underwent an ultrasound of the neck, which showed "numerous round to oval hypoechoic nodes". The larg-



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est had a15 x 16mm diameter on the left side, while on the right side, the largest had a 21 x 25 mm diameter. These nodes had increased internal vascularity with decreased Resistance Index (RI) value without any necrosis.

Un enhanced, and Gadolinium-enhanced Magnetic Resonance Imaging (MRI) of skull base that included cavernous sinus, orbital apex and neck, performed which revealed an ill-defined heterogeneous lesion involving the right lateral wall of the nasopharynx, torus tubarius, the fossa of Rosenmuller, carotid sheath, paravertebral space, right medial and lateral pterygoid muscles. The lesion medially protrudes into the nasopharyngeal lumen, and superiorly causes erosion of the floor of the middle cranial fossa, clivus, involves the right cavernous sinus with heterogenous parotid lymph node on the right side (Figure 1). The lesion described above shows post-contrast heterogeneous enhancement (Figure 2). Anteriorly, the lesion was obstructing the bilateral posterior chaonae. Fluid signal intensities were seen in bilateral middle ear cavities and mastoid air cells due to eustachian tube blockage. The right eustachian tube also showed postcontrast enhancement in its entire length s/o tumour extension along the tube (Figure 3). Mucosal thickening was present in the bilateral maxillary, frontal and ethmoid sinus, representing changes of sinusitis. Nasal turbinates and meati were normal. There were also multiple heterogeneously enhancing enlarged lymph nodes seen at various levels on both sides of the neck.



Figure 1: Nasopharyngeal carcinoma **(A)** T1 weighted coronal plane image shows an ill-defined hypointense lesion in the right side of the nasopharynx (white arrows) with appears heterointense on T2 weighted image **(B)**. T2 weighted STIR coronal plane image reveals the extension of the mass into the middle cranial fossa through foramens and causing expansion of the right cavernous sinus **(C)**. **(D)** T1 weighted post-contrast fat-suppressed (T1 FS) axial image shows heterogeneously enhancing right parotid lymph node (black arrow).



Figure 1: T1 weighted fat-suppressed (T1 FS) post-contrast images: **(A)** axial plane image shows heterogeneous mass at right nasopharynx, invading the pterygoid muscles. Posteriorly, the lesion is seen partially encasing the right internal carotid artery, **(B)** sagittal plane image shows an extension of the disease (white arrow) to the clivus posteriorly (black arrow), **(C)** axial plane image shows the disease is extended into the right parapharyngeal space (white arrow) and **(D)** coronal plane image shows an expansion of the right cavernous sinus due to tumour invasion (white arrow).



Figure 3: Three-dimensional (3D) constructive interference in steady state (CISS) image shows tumour extension along the tube (white arrow).

Following oncological consultation, a biopsy of a lymph node was performed. Histological examination revealed the presence of "lymphatic metastasis of undifferentiated cell carcinoma with Regaud growth pattern, likely nasopharyngeal neoplasm". In addition, Rhinoscopy confirmed an"undifferentiated nasopharyngeal carcinoma", suggesting a stage IV (T4 N2cM0; AJCC 8th Edition) nasopharyngeal carcinoma.

Treatment

Once the diagnosis was obtained, the patient was referred to an oncology department. There, he was treated for six weeks with radiotherapy, cisplatinum and corticosteroids.

Outcome and follow-up

After six weeks patient was came to department for follow-up with relief of ear complains.

Discussion include a very brief review of similar published cases

NPCs are one of the head and neck malignancies common in the endemic geographical regions such as the southern region of Asia, including Bangladesh, Bhutan, India, Pakistan, Nepal, and Sri Lanka [1].

Consumption of salted fish has been related to the carcinogenic compound. Nitrosamine is one of the most frequently mentioned etiologic factors. The EBV genome is commonly detected in the biopsy specimens of NPC. Hence it has been considered to play an oncogenic role in this tumour[2]. However, in this particular patient, there was no history of exposure to carcinogens.

As for EBV infection, while it is widespread and affect over 90% of the Asian adult population, it is usually asymptomatic. The NPCs associated with EBV are of undifferentiated form [3].

Symptoms of NPC presentation can be placed in four groups. They are 1. Nasal symptoms such as discharge, nasal obstruction and epistaxis (these symptoms are related to the presence of tumour mass in the nasopharynx); 2. Otologic symptoms include tinnitus and deafness (related to the Eustachian tube dysfunction caused by the anteroposterior involvement of the tumour to the paranasopharyngeal space) ; 3. Cranial nerve palsies, commonly fifth and sixth cranial nerves (associated with the superior extension of the tumour leading to skull base erosion), might experience facial pain, headache, diplopia, and numbness; and 4. Neck masses usually appears in the upper 1/3rdneck [4].

Patients with an early stage of the NPC may present with complaints related to the fullness of the ear.

Eustachian tube dysfunction may caused by a small tumour in the fossa of Rosenmüller of the nasopharynx. The tumour usually expanded outward through the mucosa and sub-mucosa, along with the muscle bundles within the neuro-vascular planes or the fibro-fatty tissue planes. The Eustachian tube and middle ear or auditory nerve involvement with the resulting correlated aural manifestations also happen[5].

Early-stage NPC is challenging to diagnose clinically because of its hidden location in the nasopharynx [6]. Moreover, the presentations of NPC are variable. Still, they may also remain symptomless for a long time, making it harder to disclose the occult site of presentation. Moreover, sometimes patients consult doctors of different specialities who have little experience in managing nasopharyngeal carcinoma [7].

In this NPC case, the patient had a month of progressive ear discharge and six -months of the right side face pain. There was an associated feeling of fullness and hearing impairment in the bilateral ear. A thorough nasoendoscopic examination can make it possible to detect an early-stage NPC.

WHO classified NPCs in 2005 into three main types. They are keratinizing squamous cell carcinoma, non-keratinizing squamous cell carcinoma, and basaloid squamous cell carcinoma. Histopathology demonstrates, in this case, revealed that the tumour was a non-keratinizing squamous cell carcinoma. These classification of tumours are commonly more radiosensitive, which differentiate these from squamous cell carcinoma and have a stronger association with EBV [8].

MRI is advised to be an accurate examination to detect NPC. MRI can diagnose subclinical NPCs that are hidden at endoscopy. NPCs in MRI typically present low signal intensity on T1-weighted images and intermediate signal intensity on T2weighted images. Most NPCs arise from the posterolateral recess of the pharyngeal wall (Rosenmüller fossa) (82%), midline (12%) and 6-10% of patients showed normal nasopharyngeal mucosa appearance at endoscopy[9]. MRI scans reveal soft tissue thickening in the nasopharynx and involvement of adjacent structures. No further examinations are needed.

Significant prognostic factors for survival in NPC are the stage, WHO type, and age at diagnosis.[10] Therefore, this particular patient is expected to have a good prognosis.

Learning points/take home messages 3-5 bullet points

- In this case, effacement of the fat in torus tubarius and the fossa of Rosenmüller region with asymmetrical fullness is seen in early nasopharyngeal carcinomas.
- Subtle effacement with fullness along the right lateral nasopharyngeal wall, along with pathological right parotid lymph node, and bilateral mastoid effusion, due to Eustachian tube dysfunction in this patient with biopsy-proven nasopharyngeal carcinoma.
- The most common imaging appearance of NPC is a mass centered at the lateral nasopharyngeal recess with deeper extension(adjacent spaces) and associated cervical nodal disease.

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Name		Date signed	Signature	
1.	Dr. Vikash Sharma	06/06/2022	Vikash	
2.	Dr. Rajaram Sharma	06/06/2022	Réja Rom	
3.	Dr. Sunil Kast	06/06/2022	Hom	
4.	Dr. Saurabh Goyal	06/06/2022	aumantes	
5.	Dr. Tapendra Nath Tiwari	06/06/2022	Ju	

	Contributor 1 Dr. Vikash Sharma	Contributor 2 Dr. Rajaram Sharma	Contributor 3 Dr. Sunil Kast	Contributor 4 Dr. Saurabh Goyal	Contributor 5 Dr. Tapendra Nath Tiwari
Planning	+	+	-	-	-
Conduct report	+	+	-	-	-
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Authors' Names (in sequence) Email ID Contribution to Paper Signature.

Name	E-mail ID	Signature	
1. Dr. Vikash Sharma	Vikashjoin9me@gmail.com	Vikash	
2. Dr. Rajaram Sharma	hemantgalaria13@gmail.com	Reja Rom	
3. Dr. Sunil Kast	dr.kastsunil@gmail.com	Hom	
4. Dr. Saurabh Goyal	saurabhgoyal1234@gmail.com	Wirnentres	
5. Dr. Tapendra Nath Tiwari	tiwaritapendra@gmail.com	Ju	

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