

Case Report

Vision Impairment: An Unusual Initial Presentation of Nasopharyngeal Carcinoma

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Abstract— Nasopharyngeal carcinoma (NPC) is a rare malignancy worldwide. It is the fifth most common cancer among the Malaysian male. NPC is complex and unique in terms of its initial presentation. The patient usually presents late when the NPC is already in its advanced stage due to unusual and confusing nature of its manifestation which often misleads the clinician. Initial presentation of vision loss or impairment is rare in NPC. Up to date, there are only 9 reports of NPC cases presenting initially with impaired vision. We report a case of NPC with an uncommon initial presentation of bilateral visual impairment.

Keywords— Cranial nerves, Orbital apex, Nasopharyngeal carcinoma, Vision loss

I. INTRODUCTION

Nasopharyngeal carcinoma (NPC) is one of the less commonly occurring cancers worldwide constituting only 0.7%. The global incidence rate of NPC is 1 in 100000 people. The prevalence of NPC is high in South East Asia, Southern China and North Africa. NPC is the fifth most common cancer among Malaysian males, accounting for 4.0%, with life time risk of 1 in 175 [1].

NPC usually presents with non-specific symptoms which may not be related to the nose. Thus it is difficult to diagnose NPC and patients usually present late in the advance stage of the disease. In Malaysia, many other factors contribute to late presentation of NPC. These include unusual and confusing nature of its manifestation, poor

awareness of the disease, its relatively painless nature in the early stage, and the patient's tendency to seek traditional medicine first which further delay the diagnosis [2][3]. Late presentation with advanced disease definitely affects the patient's survival rate resulting in poor prognosis.

Cranial nerve (CN) involvement in NPC indicates advanced disease. The most common cranial nerves affected are CN V and CN VI. Vision loss or impairment as an initial presentation is rare. To date, there are only 9 reports of vision impairment as the initial presentation of NPC [4][5][6]. We report an uncommon presentation of NPC manifesting as bilateral visual impairment as its initial presenting feature.

II. CASE REPORT

A 47 years-old Chinese gentleman, an active chronic smoker with no medical illness, was referred to Otorhinolaryngology (ORL) Department, Hospital Ampang with a complaint of progressive painless bilateral eye blurring of vision and diplopia of 6 months duration. He denied any eye redness or discharge. There was no history of trauma or radiation prior to onset of symptoms. He developed nose block, epistaxis and neck swelling 4 months after the initial symptoms. He also complained of pain in the right cheek for the past one month prior presentation to ORL clinic. He had significant loss of weight but there was no loss of appetite.

On examination, there was fullness over the right cheek but there was no obvious swelling or mass. There was proptosis of the right eye, presence of esotropia and there was a convergent squint. However, no proptosis was noted on the left eye. He was unable to perceive light with the right eye. For the left eye, he was able to count fingers at a distance of 2 feet away. The right eye had restricted

extraocular muscle movements to all gaze. While on the left eye, there was restriction to the lateral gaze only. The intraocular pressure was 16 bilaterally with marked relative afferent pupillary defect (RAPD) in the right eye. There was bilateral cervical lymphadenopathy at level II measuring 4 x 6 cm. Intraoral examination showed a fungating mass on the hard palate and bulging of the soft palate with medialization of left pharyngeal wall beyond the midline. Rigid naso-endoscopy revealed a friable mass with a smooth surface occupying the right nasal cavity and left posterior choana which bled easily on contact. The scope was unable to pass beyond the mass. The patient also had multiple bilateral palsy of the cranial nerves (CN II, III, IV, VI, IX, XII) including unilateral left upper motor neuron seventh cranial nerve palsy.

Computed Tomography (CT) scan showed a large heterogeneous enhanced mass with the epicentre at the nasopharynx measuring 9.0 x 7.1 x 6.3 cm with multiple necrotic components. It occupied the entire nasal cavity with destruction of the nasal septum and bilateral

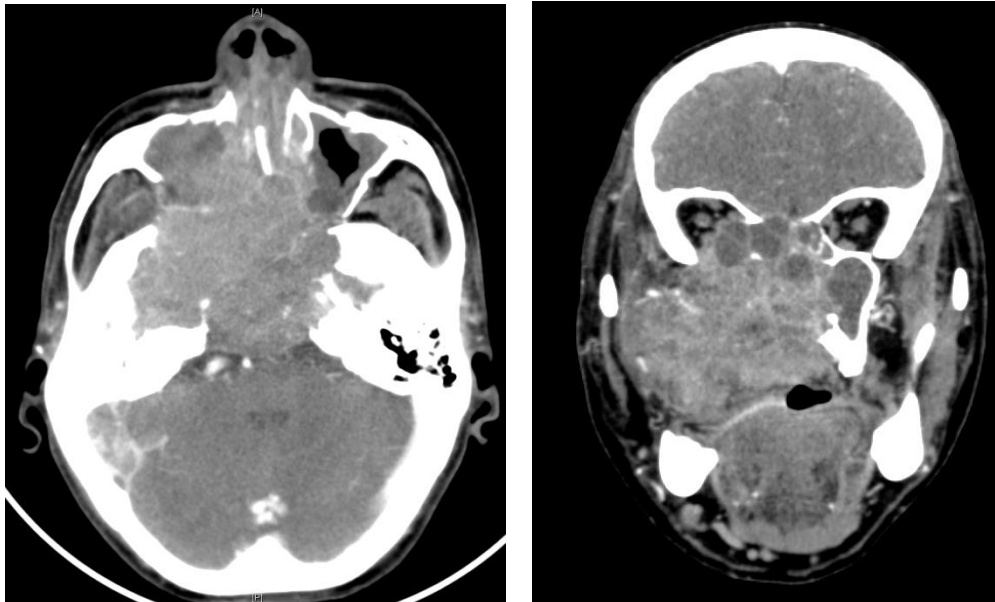


Fig. 1 & 2 CT scan on axial and coronal view showed Large heterogenous enhancing mass occupying nasal cavity with epicenter in nasopharynx involving right orbit, intracranial, sinuses and palate.

turbinates. There was extension into the maxillary, ethmoids, frontal sinuses and right orbit. The mass had no clear demarcation with the right optic nerve and right extraocular muscle at the orbital apex. Laterally it extended to the bilateral pterygopalatine fossa, right infratemporal and masticator spaces. Superiorly it extended to both anterior and middle cranial fossa with no clear demarcation with the frontal and temporal lobes. Sphenoid bone and clivus destruction was also present. Inferiorly, it involved the prevertebral muscle and parapharyngeal space up to the epiglottic level (Fig. 1 & 2). CT scan of thorax showed reticulo-nodular densities in the left lung which was indicative of metastases.

Multiple biopsies were taken from the right nasal cavity, bilateral posterior choana and lateral wall of the nose. Fine needle aspiration cytology of right cervical lymph node was done mainly to rule out the possibility of lymphoproliferative malignancy. Histopathological

examination revealed poorly differentiated squamous cell carcinoma. The EBV staining and tumour marker for lymphoma were negative. Patient underwent palliative chemoradiation therapy.

III. DISCUSSION

NPC is a rare malignancy worldwide but relatively common among Malaysians. It is complex and unique in terms of the disease presentation. Most common presenting complaint is neck swelling (57.2%), followed by nasal symptoms (19.5%), headache (14.3%) and aural symptoms (7.2%). Ophthalmic manifestation was about 5.4% in which blindness was only 1.8% [3]. There were uncommon presentations reported in the literatures which include periauricular mass, parotid mass, retropharyngeal abscess and neuro-ophthalmic symptoms. The neuro-ophthalmic manifestation, specifically, vision loss as only or the first symptom is very rare in NPC [4][6][7][8][9]. The pathway

of NPC spread causing visual disturbance is due to optic nerve compression, direct tumour invasion, orbital apex invasion or orbital invasion through pterygopalatine fossa and inferior orbital fissure, followed by ethmoid or sphenoid sinuses [7][8] which was present in this case based on CT scan findings.

It is well known that NPC may have cranial nerve involvement which indicates advanced disease. The incidence of cranial nerve involvement in NPC is about 12-35% [10][11]. It results from extension of the disease to the base of skull or infiltration through skull base foramina particularly the foramen ovale and lacerum. This route of spread may continue into the cavernous sinus which may present as cavernous sinus syndrome. To the best of our knowledge, this is the second reported case with bilateral cavernous sinus involvement [11]. In a study by Lee et al and Mo et al, majority of the patients presented with unilateral cranial nerve palsy, whereby the fifth and sixth cranial nerve were the most commonly affected; 12.5% and 10.5% respectively [12][13]. Our patient had multiple bilateral cranial nerves palsy with rare involvement of unilateral upper motor neuron lesion (UMNL) seventh nerve palsy. The involvement of UMNL facial nerve palsy was probably due to damage to the motor neuron which occurred above the facial motor nuclei located at the precentral gyrus. In this case, the tumour extended intracranially to the anterior and middle fossa. Further radiological investigation such as Magnetic Resonance Imaging (MRI) scan is needed for further detailed evaluation regarding the nerve involvement. However, it was not done in this case in view of the advanced stage of the disease at presentation which would not change the management plan.

NPC presenting with cranial nerve palsy carries a lesser survival rate and poorer prognosis [10][12][13]. Mo et al observed poorer prognosis in patients with multiple cranial nerves involvement in comparison to single cranial nerve. However, there is no significant difference in terms of involvement between upper or lower motor neuron. National comprehensive cancer network (NCCN) has recommended a standard treatment regime of concurrent chemoradiotherapy plus adjuvant or induction chemotherapy for NPC with cranial nerve involvement [13].

Sing et al in 2006 reported that 181 out of 213 (85%) of NPC patients in Sarawak, Malaysia had late presentation [2]. In our case, the patient presented 6 months after the onset of initial symptoms with features of a very aggressive tumour. The clinical features were not typical presentation of NPC. It is difficult to differentiate between NPC and sino-nasal undifferentiated carcinoma (SNUC) since the patient presented at an advanced stage of the disease. In view of the fact that NPC has higher prevalence and it is more common in our community, therefore the diagnosis of NPC was most likely in this case. SNUC usually rapidly progress over a period of few weeks [14]. However, our patient's onset of symptoms occurred over a period of 6 months duration. Cervical lymphadenopathy is the most common presenting symptom among NPC cases compared to SNUC as seen in our patient. It was supported by the CT scan findings which showed that the epi-center was in the nasopharynx. In addition, the biopsy was positive in the

post nasal space but negative in other sites including the lateral nasal wall, which made the diagnosis of SNUC unlikely. NPC has been traditionally associated with Epstein-Barr virus (EBV). However, there were reported cases of NPC which were EBV negative [15][16]. EBV positive NPCs were associated with a non-keratinizing histological phenotype. T1/2 stage at diagnosis occurred more frequently in EBV positive NPCs, while EBV negative tumours are associated with a higher T stage (T3/4) [16]. NPC has a better prognosis compared to SNUC. The primary mode of treatment for NPC is chemoradiation while SNUC is primarily treated with surgery followed by chemoradiation.

IV. CONCLUSION

Nasopharyngeal carcinoma presents with a variety of symptoms which may not be related to the nose. Visual impairment is one of the unusual initial presentation. A patient who presents with visual impairment of unknown cause warrants nasopharyngeal examination to rule out NPC. Early detection of NPC will improve the patient's quality of life with good survival rate and better prognosis.

CONSENT TO PARTICIPATE

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

CONFLICT OF INTERESTS

The authors declare that there is no conflict of interest.

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