A Rare Isolated Bilateral Abducens Nerve Palsy In Nasopharyngeal Carcinoma (NPC)

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Citation

Abstract
Nasopharyngeal carcinoma is an intriguing tumor of the upper aerodigestive tract in its subtle presentation, aggressive clinical behaviour, multifactorial cause and difficult management. The common clinical presentations are neck masses, ear symptoms, bloody nasal discharge and cranial nerve palsy. Among patients with cranial nerve palsy, trigeminal and abducens are the most observable neurologic manifestations. Isolated bilateral abducens nerve involvement is indeed rare. In this report we present a case of NPC associated with bilateral abducens nerve palsies.

INTRODUCTION
The nasopharynx is situated just below the base of the skull. Because of this proximity to the base of the skull, the infiltrating ability of the tumor and the non-specific nature of the symptoms, NPC may present with base of skull and cranial nerve involvement. The involvement of cranial nerves and the base of the skull erosion in association with NPC were recognized as poor prognostic factors. Cranial nerve involvement and base of skull erosion are related and result from superior extension of the tumor.

Review of the literature showed NPC with isolated bilateral abducens nerve involvement is indeed rare. Only 2 cases of isolated bilateral abducens nerve palsy with NPC were reported. Other reported cause is due to head injury, diffuse pontine neoplasm and aneurysm of the posterior circulation of the circle of Willis.

CASE REPORT
A 28 years old Malay man presented with blurring of vision associated with bilateral nasal blockage and intermittent bilateral epistaxis for one and a half year. He didn't have reduced hearing or tinnitus. There was no hyposmia or anosmia. Subsequently, he developed generalized throbbing headache for two weeks before he was admitted to Hospital Kota Bharu. The headache was associated with vomiting. It was worse in the morning, aggravated by straining and not relieved by paracetamol. It became worse each day and unbearable. He could not do his daily activity due to the headache.

On examination he was conscious and alert. His Glasgow Coma Scale was full. Blood pressure was 120/80 mmHg. There was bilateral 6th nerve palsies (Figure 1). All other cranial nerves were intact. There was no paranasal sinus tenderness or hypoesthesia of the cheek. Rigid nasoendoscopy showed an irregular mass at the roof of the nasopharynx predominantly on the right side with involvement of the right fossa of Rosenmuller. Mucoid and blood stained discharge was also noted on the surface of the mass. Indirect laryngoscopy showed normal findings.

Examination of the neck revealed enlarged cervical nodes on the right side at level 5 while on left side at level 2 and 3. Fundoscopy only showed congested vessels and no papilloedema seen. Biopsy of the nasopharyngeal mass showed an undifferentiated carcinoma (WHO Type 3).

CT scan of the paranasal sinus demonstrated an asymmetry of the fossa of Rosenmuller with obliteration of the parapharyngeal space on the left side (Figure 2). Irregularity of the skull base was seen. The sphenoid and ethmoid sinuses were filled with soft tissue density lesions. CT scan of the brain showed an enhancing parasellar lesion extending posteriorly in the prepontine cistern associated with bony erosions of dorsum sella (Figure 3). Erosion of the clivus was also seen. The mass was not separable from the cavernous sinus. A diagnosis of nasopharyngeal carcinoma (NPC) with intracranial extension and bilateral 6th cranial nerves involvement was made and he was subjected to palliative radiotherapy.
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Figure 1
Figure 1: Cranial nerves examination showed bilateral abducens nerve palsy.

Figure 2
Figure 2: CT scan of the paranasal sinus demonstrated an asymmetry of the fossa of Rosenmuller with obliteration of the left parapharyngeal space.

Figure 3
Figure 3: CT scan of the brain showed an enhancing parasellar lesion extending posteriorly in the prepontine cistern associated with bony erosions of dorsum sella.

DISCUSSION

In most parts of the world NPC is relatively rare, with an age-adjusted incidence rate of one per 100,000 population. However, it is one of the most common cancers affecting the people in South China, Taiwan, Hong Kong, Singapore and Malaysia. It was interesting to note that the different ethnic groups (Malay, Chinese and Indian) who have been living together in the same country for generations had marked differences in the incidence of NPC. The age-adjusted incidence rate per 100,000 population for Chinese was 11.3 for males and 4.1 for females compared with 1.3 for Malay males and 0.3 for Malay females and only 0.3 for Indian males and 0.2 for Indian females.

A study done in the Northeast of Malaysia by Indudharan et al showed the Chinese were found to be affected at a younger age compared to Malays. The affected Malays were mainly in the fifth to seventh decades of life. Their study also showed that Chinese were six times more at risk than Malays for NPC (odds ratio 5.73). Their patients with NPC, both Malay and Chinese, were found to have a male to female ratio of 2.9 to 1.

Indudharan et al reported 30.3% of patients with NPC having cranial nerve palsy while another study done in the Northeast of Malaysia by Suzina and Hamzah showed 33.9%. The incidence of cranial nerve palsy in the two
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Malaysian studies was higher than that found by Baker and Wolfe (15%), Neel (18%) and Skinner et al (13.3%). One of the reasons in this difference might be due to the fact that most of our patients presented late and already in advanced stage.

Extension and invasion by the NPC into the cavernous sinus was found to produce lesions of nerves V, VI and III. The optic nerve was involved in the presence of parasellar invasion and nerves X, XI and XII were affected at the jugular foramen. However the lower cranial nerves IX-XII may also be involved when there is a posterolateral extension of NPC directly into the carotid space.

It is generally accepted that the most commonly involved cranial nerves are nerve V and VI. Paralysis of the lateral rectus produces diplopia while numbness of the face result from the trigeminal nerve palsy. Prasad indicated that the most commonly affected nerves are VI followed by the maxillary division of the trigeminal nerve. Both Indudharan et al and Suzina and Hamzah showed similar findings. In contrast, Neel reported that the most commonly affected nerve in North American patients are V followed by VI. The direction of spread of NPC and the different type of NPC in different races could explain these differences.

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