

Received: 14.12.2017

Accepted: 08.01.2018

Published: 29.06.2018

Adam Mohamad^{1,2}, Bathma Devi Susibalan², Irfan Mohamad¹

Paraplegia as a rare presentation of nasopharyngeal carcinoma

Porażenie kończyn dolnych jako nietypowa manifestacja raka jamy nosowo-gardłowej

¹ Department of Otorhinolaryngology, Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus, 16150 Kota Bharu, Kelantan, Malaysia

² Department of Otorhinolaryngology, Tengku Ampuan Afzan Hospital, 25100 Kuantan, Pahang, Malaysia

Correspondence: Adam Mohamad, Department of Otorhinolaryngology, Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia Health Campus, 16150 Kota Bharu, Kelantan, Malaysia, e-mail: persona522115@gmail.com

Abstract

Nasopharyngeal carcinoma is a common encounter in otorhinolaryngology. It is a non-lymphomatous squamous cell carcinoma that occurs in the epithelial lining of the nasopharynx and shows varying degrees of differentiation. The aetiology is multifactorial. The disease is more common in the Chinese, those exposed to Epstein–Barr virus and in individuals with environmental factors, such as cigarette smoking, consumption of salted fish or preserved vegetables containing volatile nitrosamine, and exposure to industrial fumes or household smoke. Signs and symptoms include neck swelling, hearing loss, nasal blockage, epistaxis, cranial nerve palsy, headache, neck pain, earache or discomfort, weight loss and central nervous system manifestation when distant metastasis is present. Amongst these manifestations, 60% of patients will present with cervical lymphadenopathy followed by epistaxis which occurs in 40% of cases. However, distant site involvement uncommonly becomes the sole presenting symptom. We report a 58-year-old Chinese patient with nasopharyngeal carcinoma who presented with bilateral lower limb weakness. The patient was initially seen by an orthopaedic surgeon, with various radiological investigations conducted, including magnetic resonance imaging. Metastatic lesions were detected at the lumbar vertebra, sacrum and iliac bone. After positron emission tomography, a suspicious primary malignant lesion was detected in the nasopharynx and its biopsy confirmed the diagnosis of nasopharyngeal carcinoma. The patient was referred to the oncology team for definitive treatment, but refused further therapy.

Keywords: nasopharyngeal carcinoma, metastasis, PET scan

Streszczenie

Rak jamy nosowo-gardłowej to często spotykany nowotwór w praktyce lekarza otorynolaryngologa. Jest to nielimfoidalny rak płaskonabłonkowy rozwijający się w warstwie nabłonkowej jamy nosowo-gardłowej i wykazujący różne stopnie zróżnicowania. Etiologia jest wieloczynnikowa. Częściej chorują Chińczycy, a inne czynniki, np. zakażenie wirusem Epsteina–Barr oraz czynniki środowiskowe, takie jak palenie tytoniu, spożywanie peklowanych ryb lub warzyw konserwowych zawierających lotne nitrozoaminy, a także ekspozycja na opary przemysłowe lub pochodzące z gospodarstw domowych, mogą dodatkowo predysponować do rozwoju choroby. Rak nosogardła objawia się obrzękiem szyi, utratą słuchu, niedrożnością nosa, krwawieniem z nosa, porażeniem nerwów czaszkowych, bólem głowy i szyi, bólem lub uczuciem dyskomfortu w uchu i utratą masy ciała, a także dolegliwościami ze strony ośrodkowego układu nerwowego w przypadku przerzutów odległych. Występują powiększone węzły chłonne szyjne (u 60% chorych), a następnie krwawienie z nosa (w 40% przypadków). Niemniej przerzuty odległe rzadko powodują jedyne pierwsze objawy choroby. Przedstawiamy przypadek 58-letniego pacjenta narodowości chińskiej z rakiem jamy nosowo-gardłowej manifestującym się obustronnym osłabieniem kończyn dolnych. Początkowo pacjent zgłosił się do ortopedy, który zlecił różne badania radiologiczne, w tym badanie rezonansem magnetycznym. Zmiany przerzutowe wykryto w obrębie kręgów lędźwiowych, kości krzyżowej i kości biodrowej. Pozytonowa tomografia emisyjna wykazała obecność podejrzaną pierwotnej zmiany złośliwej w jamie nosowo-gardłowej, a biopsja potwierdziła rozpoznanie raka nosogardła. Pacjent został skierowany na oddział onkologiczny celem leczenia radykalnego, lecz odmówił dalszej terapii.

Słowa kluczowe: rak jamy nosowo-gardłowej, przerzuty, PET

INTRODUCTION

Nasopharyngeal carcinoma (NPC) is the commonest malignancy of the head and neck region. The causes are multifactorial and include smoking, diet, genetic factors and exposure to Epstein-Barr virus⁽¹⁾. In early stage of NPC, patients usually report epistaxis and nasal blockage, while in later stages patients present with neck swelling. However, the latter is commoner⁽²⁾. A small number of patients may present with neurological symptoms after they have sought traditional treatment, but it is rarely the sole presenting symptom.

CASE REPORT

A 58-year-old Chinese man presented with bilateral lower limb weakness and numbness that had persisted for 3 months. It was preceded by neck swelling 6 months earlier, associated with intermittent nasal blockage and epistaxis. Apart from that, he also had reduced hearing 2 months prior to the diagnosis. He had been a chronic smoker for 40 years. There was no family history of malignancy. Due to worsening limb weakness, he reported to an orthopaedic surgeon. Lumbosacral magnetic resonance imaging (MRI) showed multiple metastases in the lumbar vertebrae, sacrum and iliac bone (Fig. 1). The subsequent positron emission tomography (PET) scan revealed several metastases in the brain and a suspicious primary malignant lesion in left nasopharynx (Fig. 2), which had already spread to the central and left lateral skull base. There were several metastatic nodes in the left cervical and left supraclavicular region. The patient was referred to the otolaryngologist for further assessment.

On examination, the patient was alert but in a cachexic state. The abducens nerve and other cranial nerves were intact. He had mild-to-moderate mixed hearing loss over the left ear with type B tympanogram. On peripheral nerve examination, the power of the lower limbs was 3/5 bilaterally, otherwise sensation was intact. Nasoendoscopy identified a friable mass occupying the left fossa of Rosenmüller and impinging on the left Eustachian tube opening. There were



Fig. 1. Lumbosacral MRI showing multiple lesions of varying sizes and involvement of almost all the visible bones (arrow showing lesions in L4)

the left level II nodes measuring 7×7 cm and the left supraclavicular lymph node measuring 7×7 cm. Otoloscopic examination revealed dull tympanic membrane. Biopsy confirmed a non-keratinizing undifferentiated subtype of NPC (World Health Organization, WHO type III). Computed tomography (CT) confirmed obliteration of the nasopharynx by the enhancing mass measuring $3.4 \times 2.2 \times 3.1$ cm which arose from the fossa of Rosenmüller with effacement of the lateral pterygoid fossa and early extension into the posterior part of the left nasal cavity and the sphenoid sinus (Fig. 3). There were multiple left cervical

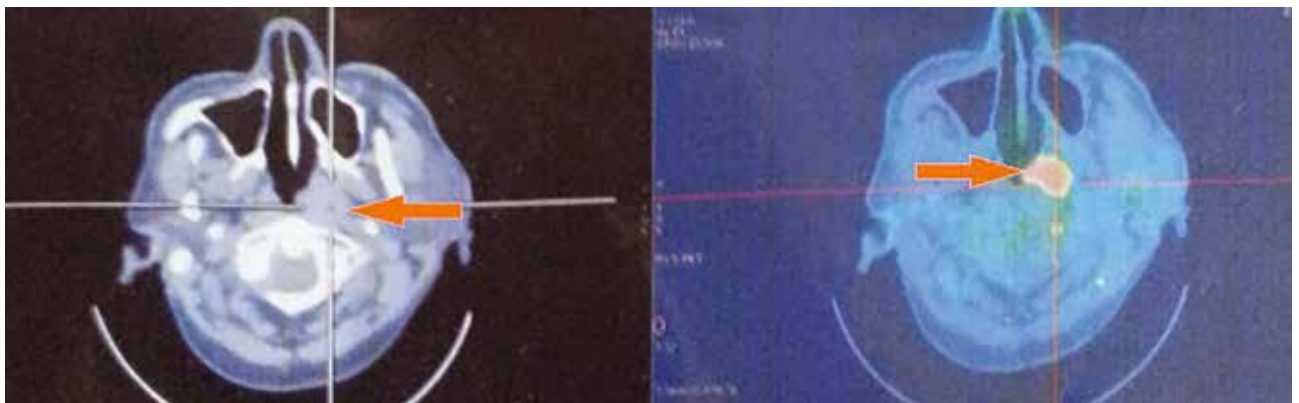


Fig. 2. Incidental PET findings suggestive of a primary malignant lesion in the left nasopharynx (arrow)

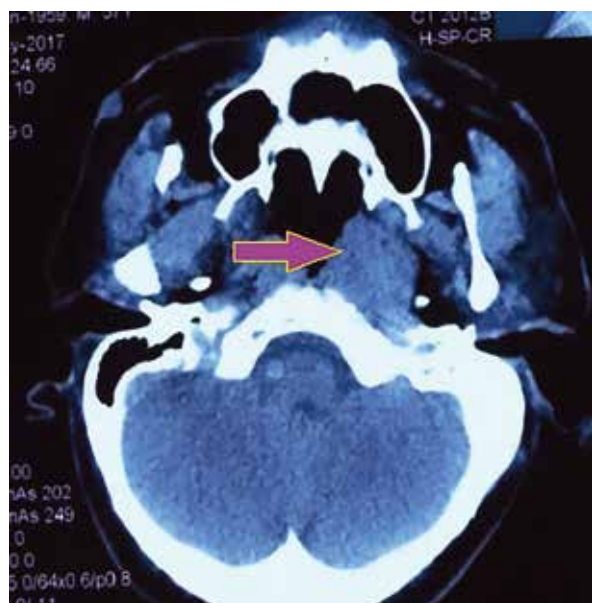


Fig. 3. CT of the neck showing obliteration of the nasopharynx (arrow) by a mass arising from the fossa of Rosenmüller

lymphadenopathies of varying sizes. The thorax and abdomen were clear except compression fracture of L4 with spinal stenosis (Fig. 4). The patient was referred to the oncology team with the diagnosis of NPC T3N3M1. However, he refused the planned chemotherapy.

DISCUSSION

Non-keratinizing NPC is an epithelial neoplasm which is endemic in Southeast Asia, Southern China, North Africa and Alaska. It can be either differentiated (WHO type II) or undifferentiated (WHO type III)⁽³⁾. These types of endemic NPC respond well to radiotherapy and chemotherapy, but they are characterised by greater tendency to locoregional spread⁽⁴⁾. In most patients, the initial presentations of NPC are unilateral conductive hearing loss and a painless slow-enlarging neck mass⁽²⁾. When the lesion becomes larger, nasal obstruction, epistaxis and cranial nerve involvement, such as ophthalmoplegia, will develop.

A total of 93 from 256 patients (36%) diagnosed with NPC had distant metastasis to the lungs, liver and skeleton owing to the haematogenous spread⁽⁵⁾. The skeleton was the commonest site for metastasis in NPC patients; by comparison, the lung is the commonest site for metastasis in other head and neck cancers⁽⁶⁾. In 1991, Sham et al. evaluated the role of bone scans in detecting subclinical bone metastasis in 132 NPC patients⁽⁷⁾. They found that 5 patients had positive bone scans consistent with metastasis at initial follow-up while 13 out of 132 of them developed symptomatic bone metastasis after 41-month follow-up. In view of low sensitivity and specificity of bone scanning, they concluded that routine bone scanning is not recommended. However, in research settings, a bone scan is helpful to make the subsequent bone scan valuable.



Fig. 4. CT of the abdomen, pelvis and thorax showing compression fracture of L4 (arrow) with spinal stenosis as well as metastasis in the pelvic bone and both femurs

In our patient, PET initially detected the primary lesion at the nasopharynx. PET using 18 fluorodeoxyglucose is very effective as it has a unique capability in detecting metabolically active lesions, and can identify both a locoregional lesion and an unsuspected distant metastasis⁽⁴⁾. In 2006, Liu et al. evaluated PET using fluorine-18-labeled fluorodeoxyglucose and conventional skeletal scintigraphy (SS) for detecting bone metastasis in NPC at initial staging. They concluded that PET is more sensitive as compared to SS in this respect. However, SS can be done as a supplementary investigation⁽⁴⁾. In our patient, MRI was done earlier because he was seen by the orthopaedic team at initial presentation to rule out a soft tissue tumour. Then, a CT scan was performed as it is the best for bony involvement evaluation. The primary lesion was detected at the first presentation, though the presentation was quite late, and the patient refused further palliative radiotherapy.

CONCLUSION

In late stage of NPC, patients might present with CNS symptoms. Advancement in imaging may help physicians detect a possible primary lesion. Thorough assessment based on the history and clinical examination should be able to suggest the possible primary site of head and neck malignancy. The presence of conductive hearing loss and neck lymphadenopathies on clinical examination should help the physician to narrow down the differential diagnosis.

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