

Recurrent nasopharyngeal carcinoma masquerading as acoustic neuroma

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ABSTRACT This is the first report of recurrent nasopharyngeal carcinoma (NPC) presenting as a cerebellopontine angle mass. The clinical presentation, investigation and management of this case, as well as the confusion and dilemma caused, are discussed. We hope to increase awareness on the multifaceted ways in which recurrent NPC could present and to share the lessons learnt from our management of this unusual and unfortunate case.

Keywords: cerebellopontine angle tumour, nasopharyngeal carcinoma, sudden sensorineural hearing loss
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INTRODUCTION

Nasopharyngeal carcinoma (NPC) is a common cancer among the Chinese population in Southeast Asia. In Singapore, its incidence is estimated to be 10.8 per 100,000 and it is the sixth most common cancer affecting males.⁽¹⁾ The first-line treatment for early stage NPC is radiotherapy. Combined modality treatment with chemoradiotherapy is advocated for locoregionally advanced disease. As patients with recurrent disease are often asymptomatic, tumour recurrence is detected by lifelong clinical follow-up, with surveillance of the nasopharynx by flexible nasoendoscopy. Biopsies are taken and imaging arranged as deemed necessary. Rarely, patients with recurrent NPC present with headaches and cranial neuropathies from intracranial involvement.⁽²⁾ However, headaches and cranial neuropathies are also common complications of irradiation.⁽²⁾ Radiation-induced neuropathies commonly involve the abducens nerve (causing diplopia), vestibulocochlear nerve (causing gradual sensorineural hearing loss [SSNHL]), and glossopharyngeal, vagus and hypoglossal nerves (causing dysphonia, dysphagia and aspiration). Nevertheless, in patients with cranial neuropathies, imaging is often performed to exclude tumour recurrence before a diagnosis of radiation-induced cranial nerve damage can be made. To the best of our knowledge, this is the first ever published report on recurrent NPC presenting clinically with sudden-onset SSNHL and radiologically as a cerebellopontine (CP) angle mass masquerading as acoustic neuroma.

CASE REPORT

A 49-year-old Chinese man presented to our department with a two-week history of left tinnitus and hearing loss. He was a chronic smoker but was otherwise fit and well with no significant past medical history. There was no family history of NPC. Ear examination was unremarkable and hearing was normal on pure-tone audiometry. However, right-sided cervical lymphadenopathy was incidentally found and a postnasal

space mass was revealed on flexible nasoendoscopy. Biopsy of the postnasal mass confirmed undifferentiated non-keratinising carcinoma (WHO type IIb). Following staging computed tomography (CT) and bone scintiscan, the tumour was staged as T1N1M0 Stage II NPC. The patient was treated with radical radiotherapy to the nasopharynx at a dose of 70 Gy divided into 33 fractions. He was reviewed regularly thereafter, and at ten months post-treatment, he complained of sudden-onset hearing loss in the left ear. There was no associated giddiness. He also noticed double vision when he looked to the left. On examination, there was no palpable cervical lymphadenopathy. The ear canals and tympanic membranes were normal bilaterally. Endoscopic examination of the postnasal space did not reveal any obvious tumour recurrence. However, cranial nerve examination revealed left abducens nerve palsy and left-sided hearing loss. Pure-tone audiogram confirmed a dead left ear.

Urgent magnetic resonance (MR) imaging was arranged to exclude tumour recurrence. This revealed a 17 mm × 16 mm enhancing mass in the left CP angle, with extension into the left internal auditory meatus (Figs. 1a–c). No enhancing mass lesion was seen in the nasopharynx to suggest recurrence (Fig. 2). Biopsy of the nasopharynx was also negative for malignancy. A provisional diagnosis of left-sided acoustic neuroma was made. The case was discussed at the multidisciplinary Head and Neck Cancer meeting, and it was felt that tumour recurrence in the left CP angle could not be excluded. Nonspecific diffuse thickening of the dura with enhancement posterior to the clivus and anterior to the pons, which was initially attributed to previous radiotherapy, was now thought to be possible for tumour recurrence. The decision was made to confirm diagnosis with a biopsy by the neurosurgical team.

The patient, however, deteriorated rapidly and developed cerebellar signs with left oculomotor, trochlear, trigeminal and facial neuropathies. Repeat MR imaging revealed progression of

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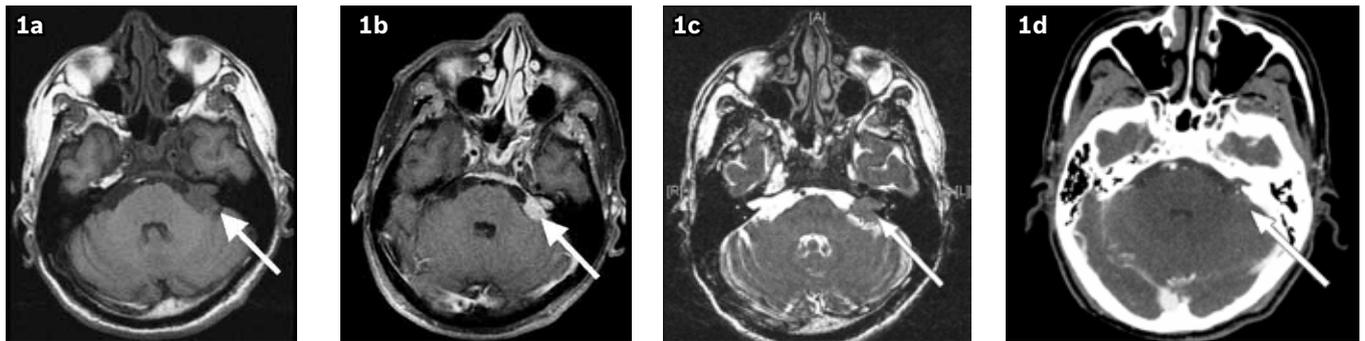


Fig. 1 T1-w axial MR image (a) without contrast; (b) with contrast; and (c) T2-w axial MR image show a left-sided cerebellopontine angle mass (17 mm x 16 mm) extending into the left internal auditory meatus (arrows). Diffuse non-specific dural enhancement is also noted posterior to the clivus. (d) CT image at the time of initial diagnosis of nasopharyngeal carcinoma shows no obvious mass at CPA (arrow).



Fig. 2 T1-w axial MR image of the nasopharynx with contrast shows a normal nasopharynx without any abnormal enhancing lesion to suggest tumour recurrence.

the disease within one month (Fig. 3). It showed soft tissue infiltration of the sella, suprasellar spaces and cavernous sinus, with involvement of the pituitary gland and cavernous portions of the internal carotid artery. There was also retroclival intracranial extension and infiltration of the premedullary and cerebellomedullary cisterns. The left CP angle mass had increased in size and now measured 27 mm x 23 mm. It extended into the internal acoustic meatus and caused widening of the porus acusticus. There was also focal mass effect on the brainstem and the left cerebellar peduncle. The nasopharynx remained unremarkable with no evidence of any abnormal enhancing mass. CT of the thorax revealed multiple pulmonary nodules suspicious for metastatic disease. Bone scintiscan also revealed increased osteoblastic activity at the skull base in the midline, compatible with tumour invasion, as well as multifocal osseous metastases along the cervical and thoracic spine. In view of the clinical and radiological findings, the diagnosis of recurrent NPC was favoured over that of an acoustic neuroma. The patient underwent palliative chemotherapy and was discharged from the hospital with home hospice care.

DISCUSSION

This case demonstrates an intriguing and unusual presentation of recurrent NPC. To date, a literature search on PubMed and Medline did not reveal any publication on recurrent NPC presenting as a CP angle mass. Its presentation with SSNHL and

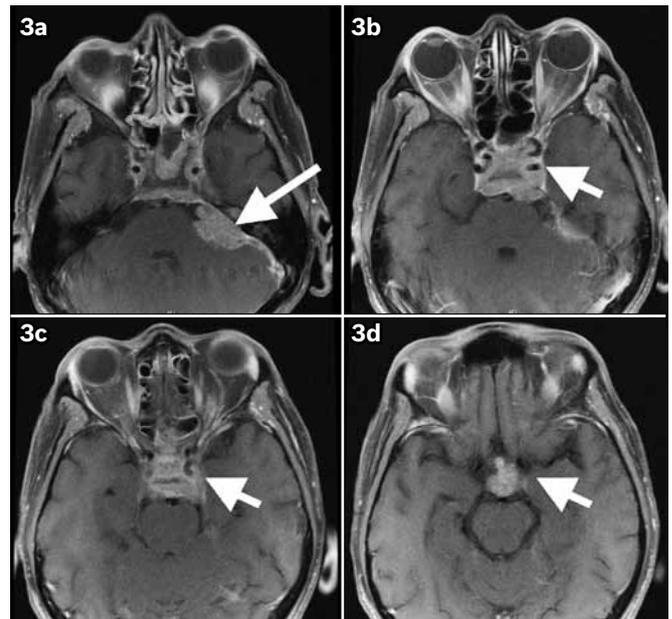


Fig. 3 T1-w axial MR image with contrast, one month later, shows (a) rapid progression of the disease. The cerebellopontine angle mass now measures 27 mm x 23 mm (arrow), and is causing mass effect on the brainstem and cerebellum; (b-d) There is soft tissue infiltration of the sella, suprasellar region and cavernous sinus (arrows).

the MR imaging appearance of an enhancing mass with extension into the internal acoustic meatus could easily mislead clinicians and experienced radiologists into diagnosing an acoustic neuroma. The associated finding of left abducens nerve palsy triggered warning bells that led to urgent MR imaging being performed in order to exclude tumour recurrence. The lack of an enhancing mass in the nasopharynx as well as subtle nonspecific dural enhancement posterior to the clivus, similar in appearance to post-radiotherapy changes, further misled the team.

To the best of our knowledge, this is the first ever reported case of recurrent NPC masquerading as an acoustic neuroma. This case report highlights the importance of considering tumour recurrence in a previously treated NPC patient irrespective of the unusual presentation, a multidisciplinary Head and Neck cancer team that includes experienced Head and Neck radiologists, and diagnostic confirmation, as far as possible, prior to initiation of treatment. This patient could have undergone surgical resection of the 'acoustic neuroma' or gamma-knife therapy if we had

accepted the initial diagnosis of an acoustic neuroma. He would have been subjected to unnecessary procedures with the attendant risks and morbidities. A misdiagnosis would also have resulted in significant emotional and psychological trauma, in addition to heavy financial costs for the patient and his family.

In conclusion, this report hopes to increase awareness of the multifaceted ways in which recurrent NPC can present and

shares lessons learnt from the management of this unusual and unfortunate case.

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