Nasopharyngeal Carcinoma: Recognizing it Early in Children with Otitis Media with Effusion

Y Noorizan, MBBCh, Y K Chew, MBBS, A Khir, MS, S Brito-Mutunayagam, MS

Department of Otorhinolaryngology, Head and Neck Surgery, Hospital Pakar Sultanah Fatimah, 84000 Muar, Johor, Malaysia

SUMMARY

Nasopharyngeal carcinoma (NPC) is a rare disease in children. Children with NPC almost always have the undifferentiated variant of the disease, which is associated with advanced locoregional and distant metastasis. We report two cases to illustrate that high index of suspicion is necessary to diagnose NPC in children especially those with atypical presentation of otitis media with effusion (OME).

KEY WORDS:
Nasopharynx, Carcinoma, Children

INTRODUCTION

Nasopharyngeal carcinoma constitutes only 1-5% of all cancers. The incidence of NPC in children is furthermore rare. NPC has a bimodal age distribution. A small peak is observed in late childhood, and a second peak occurs in people aged 55-65. The youngest case of NPC reported was a two year old and the eldest was 91 years old[1]. In our setting, the cancer is generally found in patients over 40 years of age. We reviewed all cancer cases presented to our department and from there selected NPC cases diagnosed in 2001 to 2005. The distribution of our NPC cases is also showing this bimodal age distribution, with small peak in children aged 10 – 20 (including our two cases) and in adult aged 41 – 50. In children, NPC can be difficult to diagnose as they may present differently from adults.

CASE REPORT

Case 1

An 11 years old Malay boy presented to ENT clinic with bilateral neck swelling for four months. The swelling started on his right neck and later involved the left side. He had episodes of epistaxis and haemetemesis, and intermittent left-sided headache, which was followed by reduced hearing of his left ear. On examination, bilateral cervical lymphadenopathy was noted, the left side matted and right side mobile. Throat examination was normal. Otoscopy revealed intact, dull left tympanic membrane. Tuning fork test indicated conductive hearing loss of his left ear. Tympanogram of the right ear was normal, and left ear type B. Rigid nasoendoscopy revealed adenoid enlargement. A provisional diagnosis made was bilateral cervical lymphadenopathy to rule out lymphoma, with left otitis media with effusion (OME). He was then admitted for excision biopsy of the right cervical lymph node and left myringotomy and grommet insertion under general anesthesia. Post operatively patient was well, and grommet was in-situ. The histopathology examination showed metastatic carcinoma. A CT scan revealed mass from nasopharynx. Patient was diagnosed to have nasopharyngeal carcinoma and underwent chemotherapy and radiotherapy which he completed in November 2005 at Hospital Kuala Lumpur. He is being regularly followed-up at our clinic and until now there is no evidence of disease recurrence.

Case 2

An 8 years old Malay boy presented in December 2004 with headache, left otalgia with recurrent ear discharge for six weeks. No reduced hearing, tinnitus or vertigo was present. Examination revealed wax in right ear canal, pus in left ear. Tympanic membrane was intact bilaterally. Tympanogram of his right ear was normal but the left ear was type B. He was treated medically for left otitis externa and OME. On follow up, he had recurrent symptoms and persistent type B tympanogram on his left ear. In April 2005, he developed severe otalgia and was admitted for emergency left myringotomy and grommet insertion under general anaesthesia, the finding was glue ear. Rigid nasoendoscopy at the same setting revealed an enlarged adenoid with mucus, the left eustachian tube opening was swollen and oedematous. He had a temporary pain relief but later developed trismus associated with poor oral intake. His left cheek was painfully swollen and increasing in size. Bilateral cervical lymphadenopathy was also noted. The symptoms rapidly progressed to ptosis with lateral rectus palsy of the left eye. CT scan revealed a large enhancing mass at left upper parapharyngeal/retropharyngeal space with extension into left parasellar region causing erosion and mixed sclerotic lesion of the skull base. Biopsy was taken from his left post nasal space and confirmed the diagnosis of NPC. He completed neoadjuvant chemotherapy followed by radiotherapy in December 2005. Unfortunately in January 2006, this boy was found to have metastasis to his left 7th and 8th ribs. Repeat CT scan showed residual left NPC. Currently his condition is poor with reduced oral input. We are managing him palliatively.

DISCUSSION

The most common presenting symptom of NPC is a painless mass in the upper neck, observed in 80% of patients. Nasal symptoms are commonly obstruction, bleeding and discharge. Auditory symptoms are otalgia, tinnitus, conductive hearing loss and serous otitis caused by obstruction of eustachian tubes. When the tumour invades

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Corresponding Author: Noorizan bte Yahya, Department of ENT, Hospital Pakar Sultanah Fatimah, 84000 Muar, Johor, Malaysia
Email: noorizan_yahya@yahoo.com

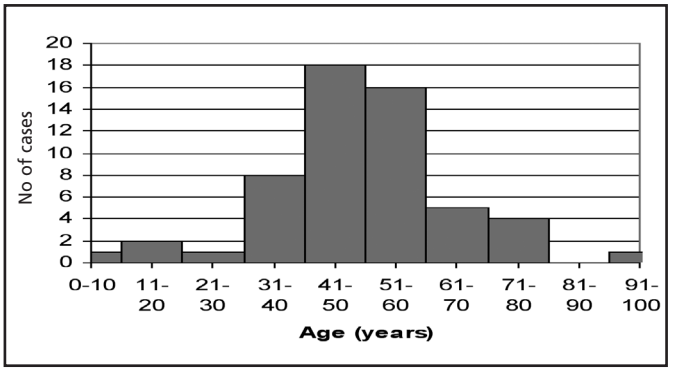

Chart 1. Distribution of patients with NPC according to their age (Hospital Pakar Sultanah Fatimah Database, 2001 to 2005).

the base of skull, cranial nerves palsy may result commonly affecting third, fifth, sixth and 12th nerves. Other symptoms include trismus, headache, facial pain and neck pain [2].

Children are prone to have otitis media with effusion (OME) mainly due to poor function of the eustachian tubes. Some of them need to undergo myringotomy and grommet insertion if the disease persists. In these two cases, it was demonstrated that we had difficulties in deciding to biopsy the post nasal space. This is mainly due to the rarity of NPC in children leading to low index of suspicion in both cases. Both of them had symptoms of eustachian tube dysfunction leading to OME. It is worthwhile to note that OME is an early presentation of NPC. It is probably an excellent idea to screen all OME cases for NPC. But do we need to take biopsy in all OME cases? The primary tumour can usually be located by endoscopic examination of the nasopharynx. Slight fullness in the fossa of Rusenmuller or a small submucosal bulge in the roof may be the only sign of a primary tumour. Nevertheless this could also be adenoids in children. In the first case, the OME was associated with bilateral cervical lymphadenopathy. The second case was associated with severe pain and cranial nerves involvement. Therefore we are suggesting that to diagnose NPC, children with OME having atypical presentation should be biopsied post nasally. Atypical presentation should include cervical lymphadenopathy, severe pain and cranial nerves palsy. CT scan or MRI scan should be used to assess the extent of local tumour growth and base of skull involvement once HPE has confirmed the diagnosis.

The mainstay of treatment is radiotherapy to the nasal and pharyngeal cavities as well as the entire neck. Doses used are adjusted according to age of children. Chemotherapy is used in advanced cases. To improve survival in childhood NPC, early administration of effective chemotherapeutic agent is needed. Complications need to be observed in children following radiotherapy and chemotherapy. Late toxicity of radiotherapy can be significant in younger children e.g. xerostomia, hypothyroidism, fibrosis of the neck and dental abnormalities. Growth retardation or panhypopituitarism may occur secondary to radiotherapy to pituitary gland. Sensorineural hearing loss is associated with the use of cisplatin and radiotherapy. Renal toxicity of cisplatin, poor dental hygiene and caries are also associated complications [3].

In conclusion, NPC in children is rare. High index of suspicion is required to detect early disease in children. OME in children should not be taken lightly and those with atypical presentation, post nasal space should be examined and biopsied.

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