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CASE REPORT

Sneaking Its Way Up: External Auditory Canal Involvement by an Otherwise Inconspicuous Nasopharyngeal Carcinoma via Spread Through the Eustachian Tube

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ABSTRACT

External auditory canal and middle ear involvement is an uncommon initial presentation for early primary nasopharyngeal carcinoma. Previously reported cases were only associated with advanced skull base erosion or recurrence after radical treatment. We present a patient with nasopharyngeal carcinoma with a small inconspicuous primary tumour in the nasopharynx without skull base erosion, but who first presented with auditory symptoms and was found to have biopsy-proven external auditory canal involvement. Contrast-enhanced magnetic resonance imaging and ^{18}F-fluorodeoxyglucose–positron emission tomography clearly demonstrated the route of extension via the Eustachian tube into the middle ear and mastoid air cells. This report highlights the importance of heightened vigilance for nasopharyngeal carcinoma and careful nasal endoscopic examination when encountering an undifferentiated carcinoma of the external auditory canal, especially in areas endemic for nasopharyngeal carcinoma. Similarly, otoscopic examination and modern imaging techniques such as magnetic resonance imaging or positron emission tomography are invaluable for investigating suspected nasopharyngeal carcinoma that presents as auditory symptoms.

Key Words: *Ear canal; Eustachian tube; Magnetic resonance imaging; Nasopharyngeal carcinoma; Positron-emission tomography*

中文摘要

不顯眼的鼻咽癌經咽鼓管進入外耳道：靜悄悄地向上游

黎詠宇、鄭志堅、唐美思、邱振中

外耳道和中耳受累是早期原發性鼻咽癌罕見的首發癥狀，過往的病例報告也僅見於廣泛顱底侵蝕或根治性放療後復發的案例。本文報告一名鼻咽癌患者最初出現聽覺功能障礙，後發現鼻咽位置有一粒不起眼的小原發腫瘤，雖無顱底侵蝕，活檢證實累及外耳道。對比增強磁共振成像（MRI）和18F-氟—正電子發射斷層掃描（PET）清楚顯示鼻咽癌擴散途徑經由咽鼓管進入中耳和乳突氣房。該病例提示當外耳道發現未分化癌時，尤其是在鼻咽癌流行的地區，應仔細進行鼻腔內鏡檢查及考慮鼻咽癌的可能性。同樣，對於有聽覺症狀並疑似鼻咽癌的病人，耳鏡檢查和現代的成像技術如MRI或PET是有效的診斷方法。

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INTRODUCTION

We report a patient with rare external auditory canal (EAC) and middle ear involvement by an inconspicuous primary nasopharyngeal carcinoma (NPC) via spread through the Eustachian tube (ET) at first presentation.

CASE REPORT

In November 2013, a 42-year-old man of good past health presented to an otolaryngologist with hearing impairment and a 'blocked sensation' in the left ear for 2 months. There was also intermittent blood-stained discharge from the left ear. There was no associated nasal symptom, nor were there any cranial nerve deficits or palpable cervical lymph nodes.

The initial otoscopic finding was granulation tissue with contact bleeding in the left EAC. Magnetic resonance imaging (MRI) of the left ear showed a soft tissue lesion within the left middle ear cavity abutting the cochlear promontory. The lesion demonstrated contrast enhancement in post-gadolinium images. The lesion extended anteriorly into the left ET and posteriorly into the aditus, and was associated with congestion of the mastoid air cells. A small extension of contrast enhancement was noted in the mastoid air cells, just lateral to the semi-circular canal. The MRI diagnosis was a glomus tympanicum paraganglioma. The nasopharynx was uniformly thickened with slight contrast enhancement over the left side, but no bulky mass was shown (Figures 1 to 3). Computed tomography (CT) of the left temporal bone also showed a soft tissue lesion occupying the left middle ear cavity, which had blocked the mastoid air cells as well as the ET. Bony erosion was not evident on CT. The CT finding was non-specific and the differential diagnoses included inflammatory tissues, a cholesteatoma, or an extensive glomus tumour (Figures 1 and 2).

Repeated otoscopy revealed hyperaemic non-pulsatile nodules over the posterior-inferior and posterior walls

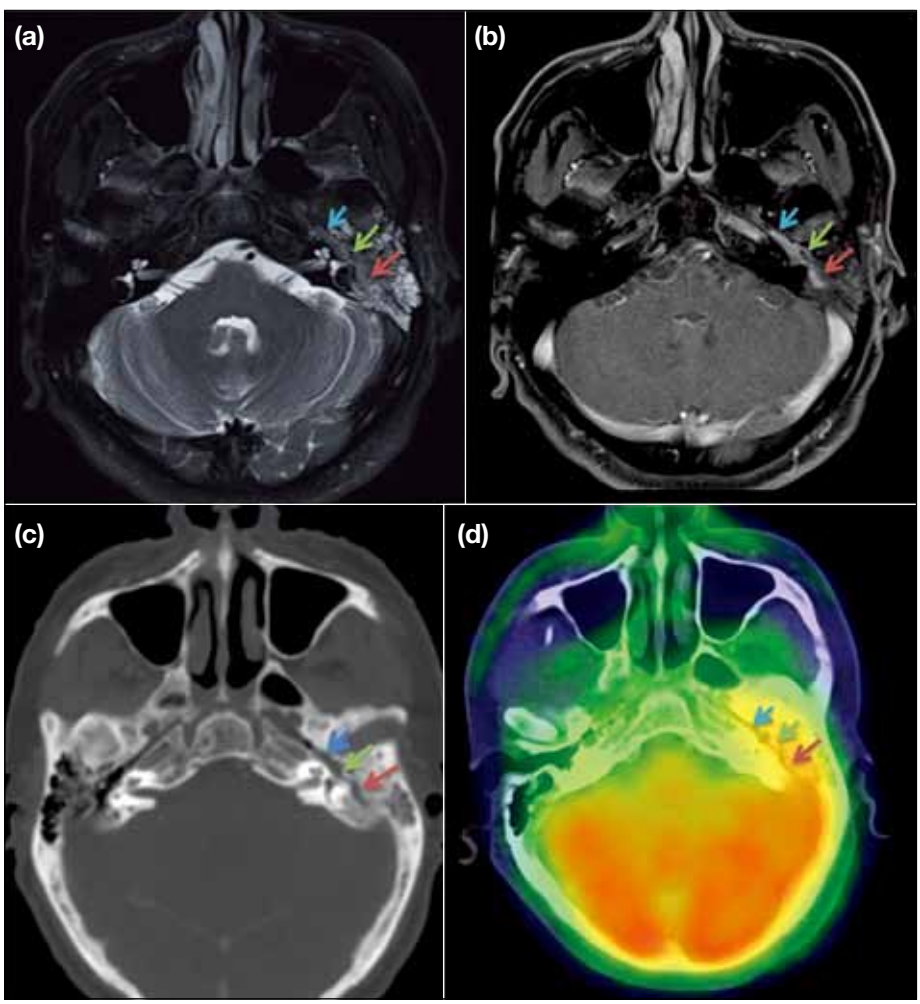

Figure 1. Axial images of the auditory apparatus in the superior aspect of the temporal bone. (a) T2-weighted and (b) T1-weighted magnetic resonance images, post-gadolinium. (c) Computed tomography image, bone window. (d) ^{18}F-fluorodeoxyglucose positron emission tomography–computed tomography fusion image. These depict the nasopharyngeal carcinoma extending from the bony Eustachian tube (blue arrows) through the middle ear (green arrows), and into the congested mastoid air cells (red arrows). Contrast enhancement on magnetic resonance imaging and standardised uptake value on positron emission tomography highlight the route of spread, and distinguish the tumour from the uninvolved surrounding mastoid air cells. There is no obvious bony destruction.

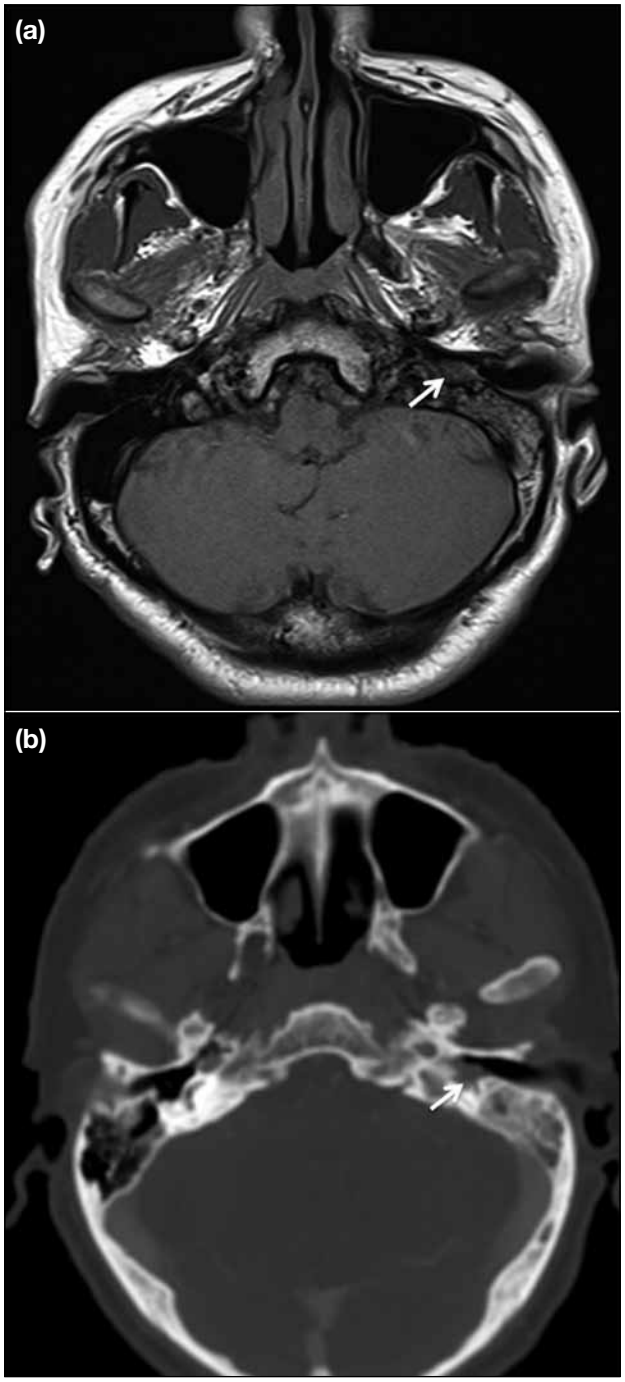

Figure 2. Axial images taken at the inferior aspect of the temporal bone: (a) T1-weighted magnetic resonance image and (b) computed tomography image, bone window. These images show the soft tissue mass filling the external auditory canal (white arrows). The nasopharynx is uniformly thickened without a bulky mass.

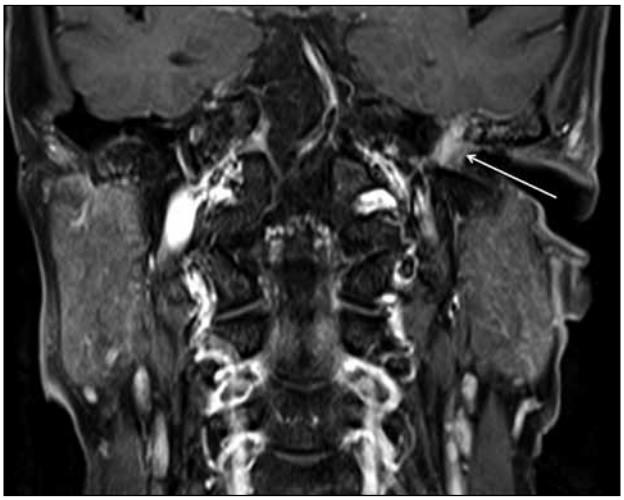

Figure 3. A T1-weighted coronal magnetic resonance image with gadolinium contrast. The tumour extends posteriorly to involve the medial mastoid air cells, showing contrast enhancement (white arrow).

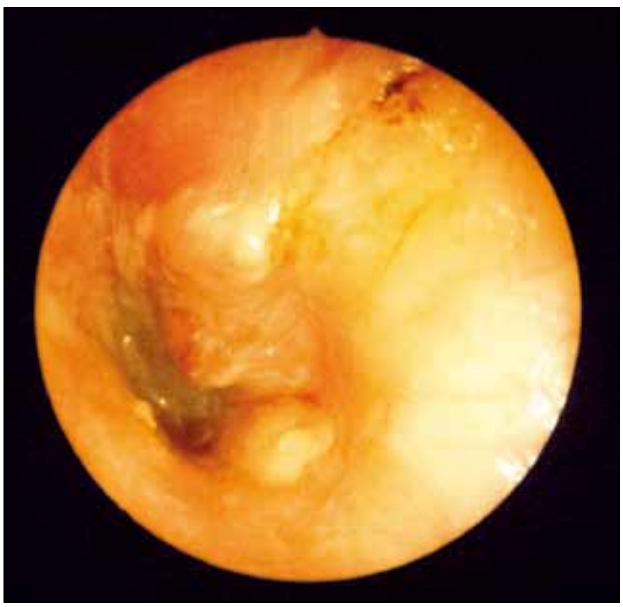

Figure 4. Otoscopic findings of hyperaemic non-pulsatile nodules deep inside the left external auditory canal.

deep inside the left EAC (Figure 4). Nasal endoscopy showed a congested nasopharynx. The appearance resembled a florid adenoidal growth atypical of NPC.

Tissue biopsies taken from the left EAC nodules and the nasopharynx were both infiltrated by undifferentiated carcinoma cells with oval nuclei and distinct nucleoli with background chronic inflammatory cells. In the EAC specimen, the carcinoma cells were positive for cytokeratin. Staining for synaptophysin and S100 protein were negative. In-situ hybridisation for Epstein-Barr virus encoded–RNA was positive, compatible with NPC of a non-keratinising undifferentiated type (Figures 5 and 6).

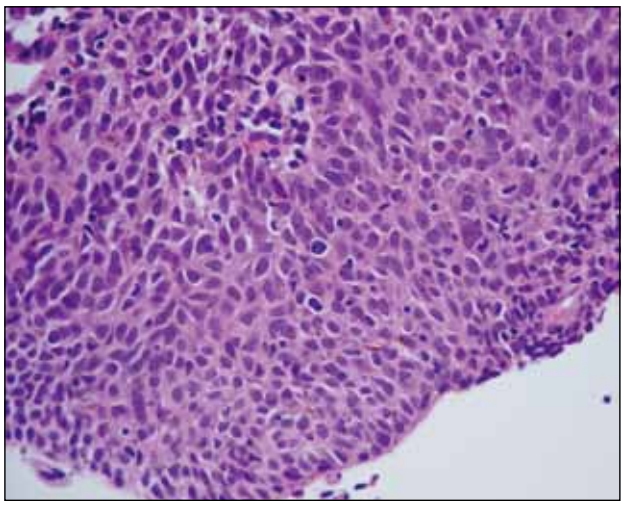

Figure 5. Microscopic view of undifferentiated carcinoma within the left external auditory canal (H&E; original magnification, x 400).

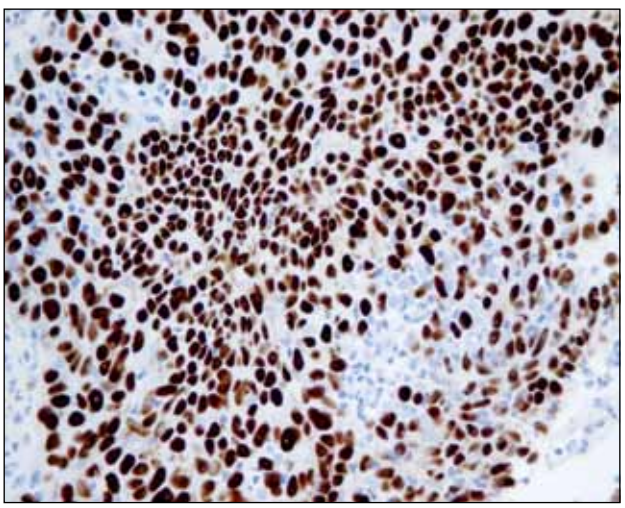

Figure 6. The external auditory canal specimen demonstrates diffuse nuclear reactivity with Epstein-Barr virus–encoded RNA in-situ hybridisation (DAB, 3,3' diaminobenzidine; original magnification, x 400).

To complete the tumour staging, ^{18}F-fluorodeoxyglucose (FDG) positron emission tomography (PET)–CT was performed. Soft tissue thickening with mild FDG hypermetabolism of maximum standardised uptake value (SUV_{max}) of 2.9 was noted in the left side of the nasopharynx, obliterating the left fossa of Rosenmuller. There was extension of FDG hypermetabolism across the midline and posterolaterally to the bilateral foramina ovale. No gross bony erosion over the skull base was depicted. There was increased FDG uptake along the left ET with SUV_{max} of 4.7. FDG hypermetabolism was noted at the left inner and middle ear, and possibly part of the congested left mastoid air cells, which was suspicious of malignant involvement (Figure 1). No regional lymph nodes or distant metastasis were noted.

Overall, the radiological and pathological findings were compatible with locally advanced NPC with left ET, middle ear, and EAC involvement.

Primary radical chemo-irradiation was offered to the patient. However, he refused treatment because of fear of treatment-related toxicities and after thorough discussion of advantages and disadvantages. His wish was respected.

DISCUSSION

NPC is rare in most parts of the world, with onset rates of less than 1 per 10,000 of all cancers.[1] Nonetheless, it is endemic in certain areas, such as Southeast China. The highest incidence of NPC has been observed in Hong Kong, where it ranked the seventh commonest cancer in 2011, with an incidence of 12.2% per 100,000 population.[2]

The commonest presentation of NPC is cervical lymphadenopathy, although nasal symptoms (discharge, bleeding, obstruction) and otologic symptoms (tinnitus, deafness, pain, discharge) should also evoke clinical suspicion. Auditory symptoms are the main presenting symptoms in 18% of patients[3] and a middle ear effusion is found in 38% of patients first diagnosed with NPC.[4,5] Many theories have been postulated for the mechanism of secretory otitis media resulting from ET tube dysfunction in NPC. One of the most popularly discussed theories is paralysis of the tensor veli palatini muscle. The tensor veli palatini muscle actively dilates the ET during swallowing. The nerve for the tensor veli palatini muscle passes through the superior part of the parapharyngeal space. In a cross-sectional imaging and electromyography study, the tensor veli palatini muscle was paralysed in 67% of symptomatic ears, suggesting invasion or compression of the nerve by a T2 NPC.[6] Middle ear effusion has also been found to be associated with major displacement of the cartilaginous ET by a tumour.[7]

Direct NPC invasion into the middle ear is rare. In an MRI series of 102 'sides' of nasopharynx involving NPC, only 13 had invasion of the cartilaginous ET,

and none had invasion of the lumen of the bony ET or middle ear.[7] Only six NPC patients with middle ear extension have been reported in the English-language literature, and most were recurrences after primary treatment with radical (chemo)radiotherapy.[8-12] Two more patients have been reported in Turkish,[13] and a case series has been written in Chinese, which included nine NPC patients with tumour growth in the EAC, of which three were found at first diagnosis and six occurred during recurrence.[14]

Similar to this patient, the reported patients commonly presented with ear discharge, hearing deficit, and otalgia; and careful otoscopic examination often found EAC polyps. A lower motor neuron type of facial nerve palsy was another interesting manifestation.[11-13] CT always showed soft tissue mass in the middle ear cavity, frequently with associated skull base bone destruction and mastoid air cell involvement.[9,11] Most of the earlier patients did not undergo MRI or PET. In patients with recurrence, most did not recur in the nasopharynx, and many subsequently developed distant metastases despite aggressive local salvage treatment, including mastoidectomy with or without adjuvant radiotherapy,[9,11] and upfront re-irradiation concurrent with chemotherapy.[14]

In this patient, the route of invasion to the middle ear was through the ET as evidenced by the contrast enhancement on MRI and the SUV uptake on PET along its course. Direct tumour invasion of the ET has been documented by MRI studies, although for reasons that are unclear, this has rarely been reported.[7,15] Interestingly, in this patient, there was little erosion of the surrounding temporal bone. This could point to true mucosal or submucosal spread along the ET, rather than a locally advanced NPC invading the ET and middle ear externally through extensive skull bone involvement. The tumour was suspected to further reach the mastoid air cells, which communicate with the middle ear posteriorly via the tympanic antrum. True malignant involvement was difficult to differentiate from congestion on CT or MRI, but seemed likely as only part of the congested mastoid air cells demonstrated contrast enhancement on MRI and FDG hypermetabolism on PET. In contrast, in some of the previously reported patients, more extensive and destructive, hence radiologically more obvious, tumour involvement of the mastoid air cells was noted.[16] If this patient had agreed to radical radiotherapy, we would have co-registered the PET images to the planning CT images and included the FDG-avid mastoid air cells as part of the gross tumour volume.

The radiological differential diagnosis for this patient was glomus tympanicum paraganglioma. Paraganglioma are slow-growing vascular tumours that arise from neuroectodermal paraganglion cells located near nerves and vasculature. Although glomus tympanicum, arising along the inferior tympanic nerve, is the most common primary tumour of the middle ear, glomus tumours are generally rare. In contrast, NPC is endemic in Hong Kong.[2] In this patient, the otolaryngologist has demonstrated high clinical vigilance in performing a nasal endoscopy to look for spread from a nasopharyngeal primary tumour, even in the absence of a bulky NPC on radiological imaging.

CONCLUSION

This patient highlights the importance of careful endoscopic examination and biopsy of the nasopharynx to actively exclude primary NPC when encountering an undifferentiated carcinoma of the EAC, particularly in regions endemic for NPC. Similarly, otoscopic examination and modern imaging techniques such as MRI or PET are invaluable for investigating a suspected NPC that presents as auditory symptoms. Clinically inconspicuous NPC may involve the middle ear and EAC via the ET in the absence of a bulky primary tumour or significant skull base destruction. Similar vigilance should be applied when investigating auditory symptoms after radiotherapy, even though post-treatment toxicity to the auditory apparatus is common. Without bearing these possibilities in mind, we may consequently delay detection of disease in sites amenable to radical primary or salvage treatment.

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DECLARATION

There are no known conflicts of interest and there has been no financial support for this work.

REFERENCES

1. Yu MC, Yuan JM. Epidemiology of nasopharyngeal carcinoma. Semin Cancer Biol. 2002;12:421-9. crossref

2. Hong Kong Cancer Registry. Available from: http://www3.ha.org.hk/cancereg/default.asp. Accessed 9 Jun 2014.
3. Skinner DW, Hasselt CA. Nasopharyngeal carcinoma: methods of presentation. Ear Nose Throat J. 1990;69:237-40.
4. Low WK, Fong KW. Hearing disability before and after radiotherapy for nasopharyngeal carcinoma. J Laryngol Otol. 1996;110:121-3. cross ref
5. Tang NL, Choy AT, John DG, van Hasselt CA. The otological status of patients with nasopharyngeal carcinoma after megavoltage radiotherapy. J Laryngol Otol. 1992;106:1055-8. cross ref
6. Su CY, Hsu SP, Lui CC. Computed tomography, magnetic resonance imaging, and electromyographic studies of tensor veli palatini muscles in patients with nasopharyngeal carcinoma. Laryngoscope. 1993;103:673-8. cross ref
7. King AD, Kew J, Tong M, Leung SF, Lam WW, Metreweli C, et al. Magnetic resonance imaging of the eustachian tube in nasopharyngeal carcinoma: correlation of patterns of spread with middle ear effusion. Am J Otol. 1999;20:69-73.
8. Cundy RL, Sando I, Hemenway WG. Middle ear extension of nasopharyngeal carcinoma via eustachian tube. A temporal bone report. Arch Otolaryngol. 1973;98:131-3. cross ref
9. Kwong DL, Yuen AP, Nicholls J. Middle ear recurrence in two patients with nasopharyngeal carcinoma. Otolaryngol Head Neck Surg. 1998;118:280-2. cross ref
10. Low WK, Goh YH. Uncommon otological manifestations of nasopharyngeal carcinoma. J Laryngol Otol. 1999;113:558-60. cross ref
11. Yang MS, Chen CC, Cheng YY, Hung HC, Chen WH, Lee MC, et al. Nasopharyngeal carcinoma spreading along the eustachian tube: the imaging appearance. J Chin Med Assoc. 2004;67:200-3.
12. Korkut AY, Teker AM, Kahya V, Gedikli O, Somay A. Middle ear recurrence in nasopharyngeal carcinoma: a case report. Int Adv Otol. 2011;7:268-70.
13. Satar B, Tosun F, Ozkaptan Y. Nasopharyngeal carcinoma. A report of two cases with unusual extension. Tukiye Klinikleri J ENT. 2001;1:45-50.
14. Wang S, Yuan W, Cheng Q. Clinical analysis of 9 cases of nasopharyngeal carcinoma with external auditory canal neoplasma as first manifestation [in Chinese]. Journal of Oncology. 2003;9:340-1.
15. Naito Y, Hirono Y, Honjo I, Mori C, Hoshino K, Nishimura K, et al. Magnetic resonance imaging of the eustachian tube. A correlative anatomical study. Arch Otolaryngol Head Neck Surg. 1987;113:1281-4. cross ref
16. Zhang X, Luo J, Gao L, Xu G. Mastoid recurrence after radiotherapy for nasopharyngeal carcinoma: two case studies. Head Neck. 2011;33:1535-8. cross ref