Relapse of B-cell acute lymphoblastic leukaemia presenting as right aural polyp with facial and mandibular nerves palsy

Chew Shiun Chuen, MD1, Mohd Khairi Md Daud, MMed1, Nur Asyilla Che Jalil, MPath2, Hilma Hazmi, MB BCh2

¹Department of Otorhinolaryngology, School of Medical Sciences, Health Campus, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia, ²Department of Pathology, School of Medical Sciences, Health campus, Universiti Sains Malaysia, Kubang Kerian, Kelantan, Malaysia

SUMMARY

A patient presenting with an ear polyp is a common finding in otorhinolaryngology practice. The common causes include chronic otitis media and cholesteatoma. We report an adult female patient with a history of acute leukaemia presenting with chronic otitis media symptoms and right ear polyp. She was subsequently diagnosed as relapse of B-cell acute lymphoblastic leukaemia based on histopathological examination. The presentation may be similar to an inflammatory pathology of the middle ear, making it misleading.

KEY WORDS:

Polyps, auditory canal, external, facial nerve palsy, acute lymphoblastic leukaemia

INTRODUCTION

Polypoidal lesion occupying the external ear canal or aural polyp is a common finding in otorhinolaryngology (ORL) practice. The differential diagnosis ranges from simple foreign body granuloma to chronic inflammatory diseases such as chronic otitis media and cholesteatoma. As the result of a chronic inflammatory process, persistent otorrhea, otalgia and hearing loss are the most common presentations. Facial nerve palsy will occur in advanced disease. Persistent aural polyps which are not responsive to medical treatment required further investigations such as computed tomography (CT) scan or histopathological studies. We report an adult female patient presenting with chronic otitis media symptoms and right ear polyp who was later diagnosed as relapse of B-cell acute lymphoblastic leukaemia based on histopathological examination.

CASE REPORT

A 35-year-old female patient presented to the ORL department with a three month history of painless right ear mass associated with minimal bloody and non-foul smelling discharge, reduced hearing and tinnitus. She also had right facial numbness for one month. She had a past medical history of B-cell acute lymphoblastic leukaemia which was diagnosed two years ago, and she had completed chemotherapy. She was under regular haematology clinic

follow-up and was symptoms free, but defaulted appointments for the past six months.

On examination, there was an inflamed polypoidal mass occupying the right ear meatus; soft in consistency, nontender, reddish and without discharge or active bleed (Fig. 1A). The right tympanic membrane was not visualised. The right pinna and mastoid area appeared normal. There was presence of paraesthesia over the right mandibular nerve distribution; however, the motor function was intact.

There was right facial nerve palsy House Brackmann grade III. The left ear was normal. The nose and the throat were unremarkable. No neck nodes palpable. Pure tone audiometry showed right moderate mixed hearing loss and left normal hearing.

The right ear was examined under the microscope, and a tissue biopsy was taken from the right ear mass. Post procedure, there was no profuse bleeding and haemostasis was secured by packing. She was treated as chronic otitis media with underlying mastoiditis and a CT scan was planned. She was given topical antibiotic ear drops and oral antibiotics. However, the condition was not responsive.

During the subsequent visit, the high-resolution CT scan (Fig. 2B) was reviewed and it imaged a heterogeneous enhancing lobulated extra-axial lesion at the right temporal region, which extended inferiorly to involve the medial and lateral pterygoid muscles. The muscles appeared to be bulky and heterogeneously enhanced. The lesion had a poor fat plane with the muscles, which was suggestive of muscle involvement. The right parapharyngeal space was obliterated. The lesion occupied the right middle ear till the external auditory canal. The scutum and ossicles were intact with no adjacent bony erosion. The impression was a right temporal extra-axial lesion with extension to the right parapharyngeal space, right middle and external ear.

The histopathological study result (Fig. 2) showed the tissue to be consistent with leukemic infiltration. Subsequently, a bone marrow biopsy was taken and the patient was diagnosed with relapse of B-cell acute lymphoblastic leukaemia.

This article was accepted: 8 June 2017 Corresponding Author: Mohd Khairi Md Daud Email: khairikck@usm.my



Fig. 1a: Right ear canal polypoidal lesion (blue arrow) seen occupying the right ear meatus (right pic).

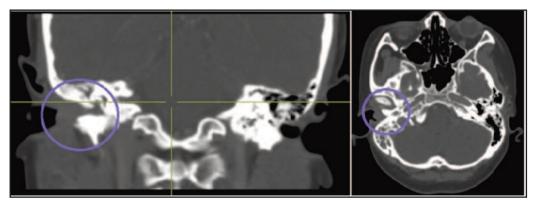


Fig. 1b: CT scan coronal view (left) and axial view (right) images soft tissue component occupying the right middle ear till external auditory canal (blue circles).

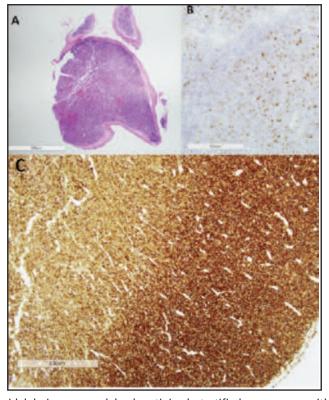


Fig. 2: A; Section shows a polypoidal lesion covered by keratinized stratified squamous epithelium infiltrated by homogenous population of malignant cells arranged in diffuse pattern (H&Ex40). B; The tumour cells show focal positivity for TdT (IHC:x200) immunohistochemistry stain brown color. C; Tumour cells show strong diffuse positivity for LCA (IHC: x100). H&E- haematoxylin and eosin, TdT- Terminal deoxynucleotidyl transferase, IHC- immunohistochemistry, LCA- leukocyte common antigen. Both TdT and LCA are expressed in leukaemia/lymphoma cells.

After establishing the diagnosis of relapsed of B-cell acute lymphoblastic leukaemia, she was started with chemotherapy; remission induction phase. She was on Hyper CVAD regime which consists of cyclophosphamide, vincristine, doxorubicin (which is also called adriamycin), dexamethasone, methotrexate and cytarabine. She responded well to the chemotherapy.

DISCUSSION

Aural polyps are frequently associated with the inflammatory process of the middle ear. The clinical findings of a polyp are described as well-circumscribed, soft and fleshly mass at the ear canal. Commonly, an inflammatory aural polyp is responsive to topical antibiotic with steroid ear drops. However, due to chronic inflammation as in chronic otitis media or cholesteatoma, the treatment may not be effective and it could further lead to cranial nerves palsies. When the presentation of aural polyps is associated with symptoms such as bleeding, earache, multiple cranial nerves neuropathy or sometimes headache, malignancy should be considered. There was a case reported for right facial nerve palsy in acute myeloid leukemia. However, mandibular branch (V3) of the trigeminal nerve involvement has never been described.

The mandibular nerve is a mixed motor and sensory nerve which exits the cranium via the foramen ovale to the infratemporal fossa. Thus, it can be affected by any lesion along its course. As in our patient's CT findings, the lesion was seen in the infratemporal fossa and it involved the structures within it, i.e. the middle and lateral pterygoid muscles. It was likely the posterior division of the mandibular nerve was affected as it consists mostly of sensory fibres; hence, it explained the paresthesia over the mandibular nerve distribution in this patient.

Histopathological study remains as the gold standard to establish the diagnosis, and to assist in the subsequent management of the patient. As there were reports that pathologies in patients presenting with aural polyps may include tumours, we should reasonably consider this in the differential diagnosis.^{2,3} Direct transcanal tissue biopsy or simple aural polypectomy; under local or general anaesthesia, can be done using direct microscope vision or an endoscope. In view of the coexistence of chronic infection, multiple biopsies may be required to confirm the disease. Superficial biopsies may show only inflammation or dysplasia, which can be deceptive. As in our case, transcanal tissue biopsy turned out to be successful. Thus, it helped to minimise the probability of more extensive surgery.

SUMMARY

Leukaemia with ear involvement is indeed rare, though there are some reported cases. Its presentation might be similar to an inflammatory pathology of the middle ear and can be misleading. Malignancy needs to be ruled out in a case of an aural polyp which does not respond to an adequate medical therapy.

REFERENCES

- Acharya R, Tulachan B. Acute mastoiditis with facial nerve paralysis in acute myeloid leukemia. Nepalese Journal of ENT Head and Neck Surgery 2012; 2(2): 13-4.
- Selcuk A, Ensari S, Cetin MA, Sak SD, Dere H. Ceruminous gland carcinoma of the external auditory canal presenting as chronic otitis media. B-ENT 2007; 3(4): 195-9.
- Cheng YF, Shiao AS, Lien CF. Pediatric external canal cholesteatoma with extensive invasion into the mastoid cavity. Int J Pediatr Otorhinolaryngol 2005; 69(4): 561-6.