Case Report
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Primary Diffuse Large B-Cell Lymphoma of the External Auditory Canal with Extensive Local Extension: A Case Report and Literature Review

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Abstract

Introduction:

Lymphoma is the second most frequent type of malignancy in the head and neck region, following squamous cell carcinoma. However, the external auditory canal (EAC) is an extremely rare primary site, with only 13 cases reported worldwide.

Case Report:

We present a case of middle-aged woman presented with a four-month history of swelling at the right EAC, reduced hearing, and intermittent otalgia. Over the following two months, she developed swelling at the right preauricular and cervical regions. Radiological evaluation revealed an enhancing mass at the right EAC with extensive local extension. Histopathological and immunohistochemical analyses confirmed the diagnosis of lymphoma. Determining the primary site was challenging due to the extensive involvement of multiple adjacent structures. However, we believe EAC was the most probable primary site, given that EAC swelling was the initial presenting symptom, although this cannot be established with certainty.

Conclusion:

The diagnosis of EAC lymphoma is challenging due to the rarity of the disease, nonspecific clinical presentation, and its frequent misdiagnosis as a benign lesion or infection at initial presentation. Additionally, obtaining an adequate biopsy sample for histopathological examination and immunohistochemical analysis can be difficult. Extensive involvement of surrounding structures at the time of presentation may further hinder accurate identification of the primary site. There is no well-established consensus on treatment guidelines, and prognosis is primarily determined by the lymphoma subtype, its clinical behaviour, the presence of B symptoms, and staging.

Keywords: External auditory canal, Lymphoma, Chemotherapy

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Introduction

Head and neck (HN) region is the second most common site for lymphoma, with lymphoma being the second most frequent type of malignancy in this area after squamous cell carcinoma (SCC) (1-3). Most cases are non-Hodgkin lymphoma (NHL) arising from lymph nodes, whereas extranodal lymphoma is relatively uncommon, with the oropharynx being the most common site, followed by the nasopharynx, thyroid, salivary glands, and paranasal sinuses (4).

The ear and temporal bone are very rare primary sites, with most cases arising from the mastoid and middle ear cavities (1-3,5). To the best of our knowledge, only 13 cases have been reported arising from the external auditory canal (EAC) (Table .1) (1-3,6-15).

Table 1. Review of reported cases of external auditory canal lymphoma

Authors	Year	Age	Sex	Site	Symptoms	Methods of tissue diagnosis	Imaging	HPE	Treatment
Maiche et al[10]	1991	71yo	M	B	Hearing loss	Biopsy EAC mass (on first attempt)	CT scan (soft tissue in EAC, no bony erosion)	Diffuse centrocytic malignant lymphoma	CTX
Merkus et al[11]	2000	83yo	F	L	Otalgia, otorrhea, FN palsy	Biopsy EAC ulcerative lesion (on first attempt)	CT scan (soft tissue in EAC with erosion of posterior wall)	Anaplastic large cell lymphoma	RT
Shuto et al[12]	2002	49yo	M	B	Incidental finding during examina-tion for other reason	Biopsy of EAC granulation tissue (on first attempt)	CT scan (soft tissue in EAC, no bony erosion)	B-cell small lymphocytic lymphoma	Surgery and CTX
Fish et al[13]	2002	53yo	F	L	Otalgia	Biopsy of EAC inflamed lesion (on third attempt)	CT scan (soft tissue in EAC, no bony erosion)	Diffuse large B-cell lymphoma	Surgery and CTX
Hersh et al[14]	2006	83yo	F	L	Hearing loss	Biopsy of EAC soft tissue lesion (on second attempt)	CT scan (soft tissue in EAC, no bony erosion)	Diffuse large B-cell lymphoma	RT
González et al[15]	2008	53yo	F	L	Non-specific discomfort and gradual hearing loss	Resection of EAC mass (on first attempt)	CT scan (soft tissue in EAC, no bony erosion)	Anaplastic, non-Hodgkin's T cell lymphoma, type Ki-1	Surgery and CTX
Bruschini et al[3]	2013	46yo	M	L	Aural fullness	Excision of EAC polyp (on first attempt)	CT scan (soft tissue in EAC, no bony erosion)	Diffuse large B-cell lymphoma	CTX
Khaw et al[8]	2014	59yo	F	L	Otalgia, otorrhea, hearing loss	Excision of EAC polyp (on first attempt)	CT scan (soft tissue in EAC, no bony erosion)	Mucosa-associated lymphoid tissue lymphoma	CTX
Mettias et al[6]	2018	49yo	F	R	Hearing loss, otalgia, ear bleeding, FN palsy, headache	Biopsy EAC mass (on first attempt)	CT scan (soft tissue in EAC,with bony erosion superiorly) MRI (intracranial extension into posterior fossa)	Diffuse high grade large B-cell lymphoma	CTX
Kim et al[1]	2019	76yo	F	L	Hearing loss, otalgia, otorrhea	Biopsy EAC mass (on first attempt)	CT scan (soft tissue in EAC, with bony erosion inferiorly and extension into parotid space)	Diffuse large B-cell lymphoma	RT
Hemanth et al[2]	2020	68yo	M	R	Hearing loss	Excision of EAC mass (inconclusive findings and need correlation with incision biopsy of neck lump and BMA)	CT scan (soft tissue in EAC, no bony erosion)	Diffuse large B-cell lymphoma	CTX
Chen et al[9]	2021	34yo	M	L	Painless mass in EAC	Biopsy EAC mass (on first attempt)	CT scan (soft tissue in EAC, no bony erosion)	Anaplastic large-cell lymphoma	CTX and RT
Sousa et al[7]	2024	53yo	M	L	Aural fullness, otalgia, otorrhea, FN palsy, vertigo	Excision biopsy of retroauricular lesion (2 biopsies of the EAC lesion were inconclusive)	CT scan (soft tissue in EAC, with bony erosion superiorly) MRI(mass occupying EAC, extending to mastoid, upper cervical fatty tissues, IJV and FN at the stylomastoid foramen)	Diffuse large B-cell lymphoma	CTX

Abbreviations: yo – year-old; M - male; F - female; B – bilateral; L – left; R – right; FN – facial nerve; EAC – external auditory canal; BMA – bone marrow aspiration; CT - computerised tomography; MRI - magnetic resonance imaging; HPE – histopathology examination; CTX – chemotherapy, RT - radiotherapy

The diagnosis of primary EAC lymphoma is challenging due to the rarity of the site, the requirement for a large biopsy sample for histopathology examination (HPE) and immunohistochemistry (IHC) analysis, and the fact that the disease may involve other nearby structures at the time of diagnosis. Treatment options include surgery, radiotherapy, chemotherapy, or a combination of these modalities, depending on the extent of the disease and systemic involvement.

Case report

A 55-year-old woman with no known medical illness presented with swelling at the right EAC, progressive hearing loss, and intermittent otalgia of four months' duration. This was associated with painless swelling at the right preauricular and neck regions for the past two months. There was no history of ear discharge, vertigo, loss of appetite, weight loss, fever, night sweats, or a family history of malignancy. She had been treated with two courses of oral and topical antibiotics by her primary care physician and was subsequently referred to our tertiary centre due to the persistence of symptoms.

On examination, a smooth, reddish mass occupied the right EAC and was firm and minimally tender on palpation, without discharge (Figure 1).

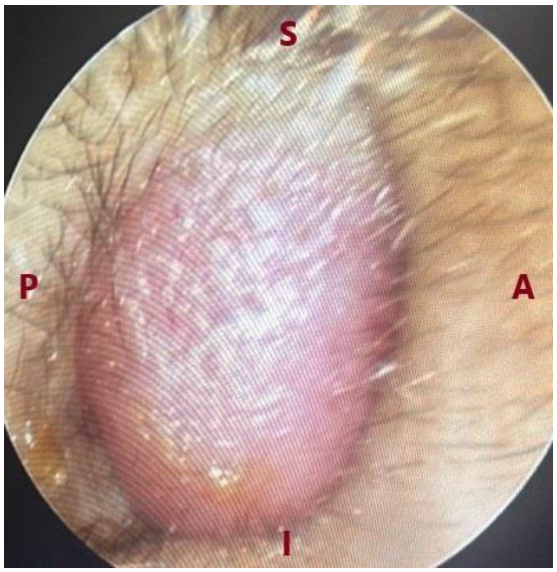

Fig 1. A firm smooth surface reddish mass occupied the right external auditory canal. S – superior; I – inferior; A – anterior; P – posterior

The tympanic membrane was not visualised, being obscured by the mass. The contralateral ear appeared normal, while Rinne's and Weber's tests suggested right conductive hearing loss. Additionally, there was a diffuse firm swelling at the right preauricular region extending to the angle of the mandible (Figure 2).

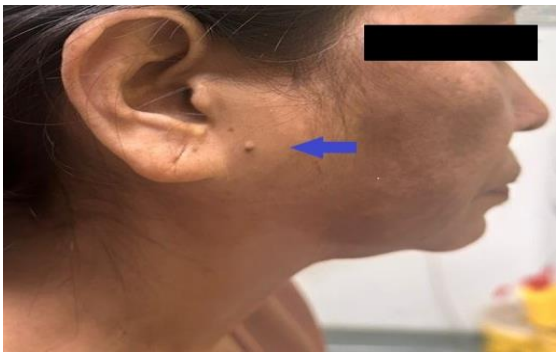

Fig 2. A diffuse firm swelling at the right pre-auricular region extending to angle of mandible

Neck examination revealed an enlarged right level II cervical lymph node, measuring 2 cm x 2 cm. Cranial nerve and other HN examinations were unremarkable. A full blood count showed normal haemoglobin, white blood cell, and platelet levels, while the peripheral blood smear revealed no abnormal cells.

Subsequently, a contrast-enhanced computed tomography (CT) scan of the HN was performed, showing a homogeneous enhancing mass at the right EAC, extending to the middle ear, parotid gland, parapharyngeal space, masticator space, and temporomandibular joint, with erosion of the lateral plate of the pterygoid bone and skull base (Figure 3).

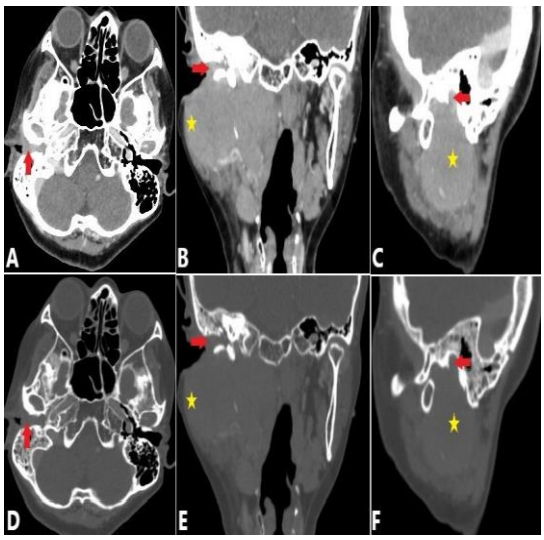

Fig 3. Contrast-enhanced computed tomography scan of the head and neck includes soft tissue (Figures A-C) and bone windows (D-F) in axial (Figures A and D), coronal (Figures B and E) and sagittal views (Figures C and F), showing a homogeneous enhancing mass at the right external auditory canal (arrow), extending to the middle ear, parotid gland (star), parapharyngeal space, masticator space, and temporomandibular joint with erosion of the lateral plate of pterygoid bone and skull base

Additionally, multiple enlarged right cervical lymph nodes were noted, the largest at level II measuring 1.2 cm x 1.9 cm. Pure tone audiometry revealed moderate conductive hearing loss in the right ear, while hearing in the left ear was normal. As the mass was a single continuous lesion, we decided to perform a Tru-Cut biopsy at the most prominent swelling at the preauricular region; however, the histology result was inconclusive. Subsequently, an incisional biopsy was performed at the infra-auricular region, where the swelling was also prominent, to leave a less noticeable scar. We did not perform biopsy in the EAC, as we believed it would be difficult to obtain adequate representative tissue for HPE. The final HPE findings were suggestive of diffuse large B-cell lymphoma (DLBCL). The tumour tissue was composed of atypical lymphoid cells arranged in a diffuse pattern, large in size, with small centrally located nucleoli and scant cytoplasm (Figures 4A and 4B).

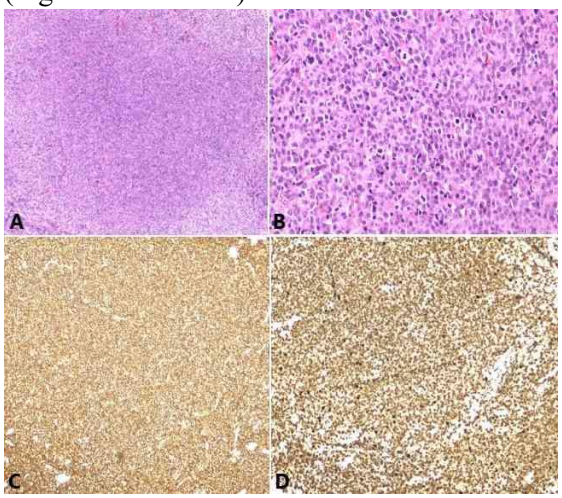

Fig 4. The tumour tissue was composed of atypical lymphoid cells arranged in a diffuse pattern (Figure A, H&E x4), large in size, with small centrally located nucleoli and scant cytoplasm (Figure B, H&E x20). CD20 immunostaining highlights malignant cells (Figure C, x4), and the Ki 67 proliferative index is approximately 80% (Figure D, x4).

Frequent mitoses, including atypical forms, were observed. IHC analysis showed that the malignant lymphoid cells were positive for CD20, PAX5, CD10 (>30%), BCL6 (>30%), BCL2 (>40%), and C-Myc (>40%), with a high Ki-67 proliferative index of approximately 80% (Figures 4C and 4D). These cells were negative for CKAE1/AE3, CD3, MUM1, and CD21.

For staging purposes, a bone marrow biopsy and CT scans of the thorax, abdomen, and pelvis were performed. A positron emission tomography-CT scan was not conducted because it was unavailable at our centre. No further disease was identified; therefore, the lesion was considered most likely to originate from the EAC with extensive local extension, corresponding to Lugano classification stage IIE. The patient was referred immediately to the haematology team and scheduled to receive chemotherapy with the R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisolone) regimen. Unfortunately, we were unable to obtain an update on her condition after the completion of chemotherapy, as she continued follow-up at another centre with haematology services.

Discussion

Lymphoma is a heterogeneous group of haematologic malignancies originating from lymphoid tissues and comprises 5-15% of HN malignancies (16). NHL is the predominant type, with approximately 10% presenting as extranodal disease, the majority originating from the area of Waldeyer's ring (17). The EAC is an extremely rare primary site, with only 13 cases reported worldwide to date.

The clinical presentation of EAC lymphoma is nonspecific and depends on the size and extent of involvement of surrounding structures (2,3). The mean age of presentation is 60 years, with no gender predilection. Interestingly, two of the reported cases presented with bilateral disease (10,12). The most common symptoms reported include hearing loss and otalgia, while facial nerve palsy was noted in three cases. Other symptoms included a painless EAC mass, otorrhea, ear bleeding, aural fullness, non-specific discomfort, vertigo, headache, and incidental findings during examination. Early presentation may involve ear discomfort and reduced hearing, which most patients perceive

as benign. Otalgia typically arises as the tumour grows, serving as a warning sign that prompts the patient to seek medical attention, as seen in the presented case. However, due to the rarity of malignant lesions in the EAC, particularly lymphoma, patients are often misdiagnosed with an infection, such as otitis externa, at initial presentation. For instance, Merkus et al. and Fish et al. highlighted diagnostic delays in patients who were repeatedly treated for presumed otitis externa, with the correct diagnosis of EAC lymphoma established only after five months (11,13). Otorrhea and ear bleeding are rare, likely due to the submucosal and subcutaneous growth patterns of lymphoma compared to more common lesions such as SCC, which exhibit exophytic and ulcerative features. Facial nerve palsy is considered a late presentation, indicating that the tumour has already extended beyond the EAC to involve the facial nerve in the middle ear or mastoid bone. The tumour may also extend to the parotid gland through the fissure of Santorini, presenting with preauricular swelling, as seen in the presented case. A similar parotid extension was reported by Kim et al., although the extension was less extensive (1). Adequate tissue biopsy is essential for an accurate diagnosis of lymphoma and its subtype, which typically requires an open biopsy, either incisional or excisional. Tru-Cut biopsy or fine needle aspiration cytology (FNAC) is usually the first-line procedure for HN tumours to obtain tissue or cells for histological and cytological examination, respectively. Unfortunately, these procedures often yield inadequate samples for the diagnosis of lymphoma. In cases of suspected lymphoma, directly performing an open biopsy instead of FNAC or Tru-Cut biopsy may prevent delays in diagnosis. However, thorough assessment and discussion with the haematology team should occur prior to open biopsy, as it may lead to tumour seeding and skin involvement if the lesion proves to be SCC. Additionally, the submucosal and subcutaneous growth characteristics of lymphoma may complicate diagnosis, as it may appear as a benign smooth surface mass on clinical examination, covered by a normal layer of mucosa or epithelium. Thus, a deep tissue biopsy is necessary to obtain a correct representative tissue. Eight of the 13 reported cases were diagnosed following biopsy of an EAC lesion, mass, or granulation tissue (1,6, 9–14). Three cases were diagnosed after excision of an EAC mass or polyp (3,8,15). The remaining two cases required additional procedures: one involved an incisional biopsy of a neck lump with bone marrow aspiration (2), while the other required an excision biopsy of a retroauricular lesion (7). Among the eight cases diagnosed via biopsy, six were confirmed on the first attempt, while two required repeat biopsies—one after a second biopsy and another after a third biopsy (13,14). Identification of the primary site of lymphoma can be difficult in late presentations, where the tumour has already involved adjacent structures, as seen in this case. A thorough history, careful clinical examination, appropriate imaging studies, and a strong understanding of anatomical relationships are essential for determining the site of origin. In this case, we believe EAC was the most probable primary site, given that EAC swelling was the first presenting symptom, although this cannot be confirmed with certainty. Subsequently, the patient experienced progressive hearing loss as the tumour grew and occluded the entire EAC. Otalgia could be attributed to pressure on surrounding structures and bony erosion. Two months later, the patient began to have preauricular swelling as the tumour extended into the parotid gland anteriorly through the fissure of Santorini. The temporomandibular joint was also involved due to its close proximity to the EAC. Through the parotid gland, the tumour extended into the masticator and parapharyngeal spaces anteriorly and medially, respectively. Medially, the tumour also extended into the middle ear through the tympanic membrane. Despite extensive involvement of the parotid gland and erosion of the skull base, there was no cranial nerve palsy, particularly of the facial nerve. This finding may suggest an expansile mass like lymphoma rather than an infiltrative mass as seen in SCC.

After reviewing the CT scan, we decided to perform a biopsy at the parotid region instead of the EAC, as it was likely to yield a more adequate representative sample. Small samples are often difficult for pathologists to interpret lymphoma and its subtype, necessitating repeat biopsies. This may result in unnecessary delays in diagnosis. A CT scan is the most commonly

employed imaging modality and is ideally performed prior to biopsy to assess the nature of the tumour, its extension, and the appropriate site for biopsy. The majority of cases showed very localised disease with no bony erosion (9 out of 13 cases). Magnetic resonance imaging may be indicated if the CT scan reveals extensive bony erosion and intracranial extension. All reported cases of EAC lymphoma were NHL, with DLBCL being the most common subtype (7 out of 13 cases). Other reported subtypes include diffuse centrocytic malignant lymphoma, anaplastic large cell lymphoma, small lymphocytic lymphoma, and mucosa-associated lymphoid tissue lymphoma. The HPE and IHC analysis of our presented case also support a diagnosis of DLBCL, making it the eighth reported case. Generally, DLBCL is considered an aggressive tumour and is the most common subtype of lymphoma (16). It presents as localised disease in 25% to 30% of cases and has a very good prognosis in limited disease, with a 10-year overall survival rate of 70% to 80% and a 5-year progression-free survival rate of 80% to 85% (18,19). The presence of B symptoms and high stage is considered poor prognostic indicators (6). The consensus on treatment guidelines for EAC lymphoma is not well established due to the limited number of cases. Treatment options include surgery, radiotherapy, chemotherapy, or a combination of these modalities. Chemotherapy was the most commonly employed treatment; six cases received chemotherapy alone, three cases received it combined with surgery, and one case received combined chemotherapy and radiotherapy. Surgery is indicated for small, localised epithelial lesions and is part of excision biopsy (3). All reported cases required adjuvant chemotherapy following surgery (12,13,15). Radiotherapy alone was used in three cases and is believed to be adequate for localised disease and lymphomas with more indolent clinical behaviour (1,11,14).

Conclusion

Primary lymphoma of the EAC is exceedingly rare, with no well-established consensus on treatment guidelines. Diagnosis is often challenging due to its nonspecific clinical presentation, frequently resulting in initial misdiagnosis of a benign lesion or infection. Furthermore, obtaining an adequate tissue sample for HPE and IHC analysis may be difficult. Extensive involvement of adjacent structures at presentation can further obscure the identification of the primary site. In the present case, the EAC was considered the most likely primary site, as EAC swelling was the initial presenting feature; however, this cannot be confirmed with certainty. Clinicians should maintain a high index of suspicion and include lymphoma in the differential diagnosis of an EAC mass.

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