Medical Science

pISSN 2321–7359; eISSN 2321–7367

Primary B-cell lymphoma: A case report of an isolated two masses occurring in the external auditory canal and middle ear cleft of the same ear simultaneously

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To Cite:
Aldhafeeri K, Telmsani L, Telmesani L. Primary B-cell lymphoma: A case report of an isolated two masses occurring in the external auditory canal and middle ear cleft of the same ear simultaneously. Medical Science, 2022, 26, ms227e1674.
doi: https://doi.org/10.54905/disssi/v26i124/ms227e1674

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Peer-Review History
Received: 11 March 2022
Reviewed & Revised: 14/March/2022 to 14/June/2022
Accepted: 15 June 2022
Published: 18 June 2022

Peer-review Method
External peer-review was done through double-blind method.

URL: https://www.discoveryjournals.org/medicalscience

ABSTRACT

We report a 52-year-old woman with primary aggressive B-cell lymphoma occurring as two isolated masses in the external auditory canal and middle ear cleft of the left ear, simultaneously. The patient presented with left ear pain, blockage, decreased hearing, and a mass in the external ear canal. Histopathological examination confirmed the diagnosis, and patient was treated with surgery, chemo & radiotherapy. Primary lymphoma is extremely rare in the ear. As per our knowledge, it's the first case reporting two isolated masses occurring simultaneously in the same ear.

Keywords: primary lymphoma, external ear canal, middle ear cleft, B-cell lymphoma

1. INTRODUCTION

Lymphomas are the 3rd most common malignant tumor of the head and neck, after squamous cell carcinoma & thyroid carcinoma. It accounts for most cases of non-epithelial malignancies in the head & neck, these neoplasms seldom presents in the temporal bone area (Müller et al., 2005). However, Hodgkin and non-Hodgkin lymphoma both could present at Nodal or extra-nodal sites, and when involving extra-nodal sites it mainly involve the following: Waldeyer's ring, paranasal sinuses, nasal and oral cavities and the salivary glands (Alexander et al., 2007). During our review of literature, it became clear that isolated case reports of a primary lymphoma in the ear in an immunocompetent patient are exceedingly rare. In this article, we report a case of B-cell lymphoma originating in the external auditory meatus a mass in the left ear & middle ear cleft, a site rarely reported in the literature.

2. CASE PRESENTATION

A 52-year-old woman, who is not known to have any previous medical illness, presented initially to our outpatient clinic complaining of left ear pain, blockage and decreased hearing without any history of dizziness, tinnitus or facial palsy. Examination revealed a mass in the left ear, arising from the superior posterior wall of the external auditory canal, obscuring the vision of tympanic membrane (figure 1). Right ear exam was normal, and there was no significant cervical lymphadenopathy. Hearing assessment showed right normal hearing and left moderate conductive hearing loss (30 db. in 1, 2 and 4 KHZ). Initial CT-scan of the temporal bone showed soft tissue mass causing complete opacification of the left external auditory meatus pushing the tympanic membrane medially; middle ear and ossicles were free (figure 2).

The patient underwent left ear examination and excisional biopsy under general anesthesia, which revealed intact tympanic membrane and disease-free middle ear cleft. Histopathological examination of the specimen reported as Epithelioid Hemangioma. After two months, the patient presented with the same mass in the external ear canal. CT of the temporal bone was done and showed soft tissue density filling the external ear canal and extending to the supra, post, and infra auricular area. Another soft tissue density filling the lateral, medial and superior attic was observed (Figure 3). MRI showed on T1 contrasted images a high signal intensity mass lesion filling the external Auditory canal extending to the supra and post auricular region with high signal intensity at the dura just above the tegmen tympani suggestive of meningeal irritation, dura looked intact (Figure 4).

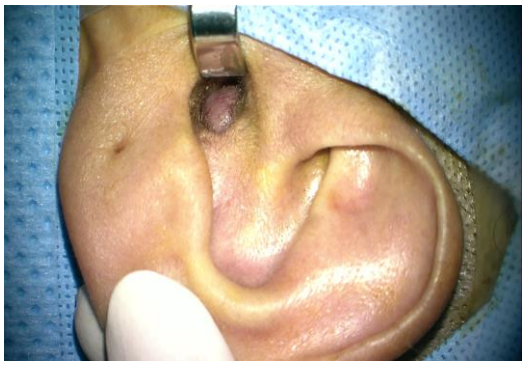

Figure 1 Mass filling the external auditory canal.

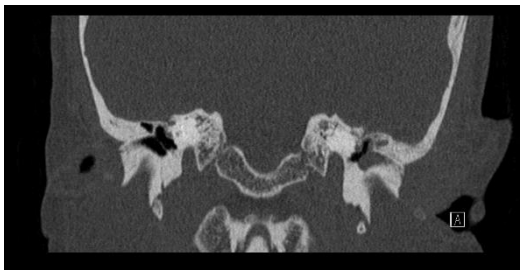

Figure 2 soft tissues mass filling the left external ear canal and pushing the tympanic membrane medially.

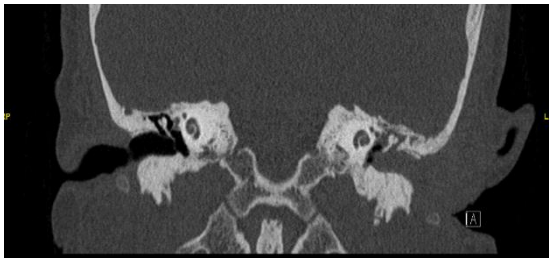

Figure 3 Soft tissue density filling medial and lateral attic, the external auditory canal and extending to supra, post and infra auricular regions.

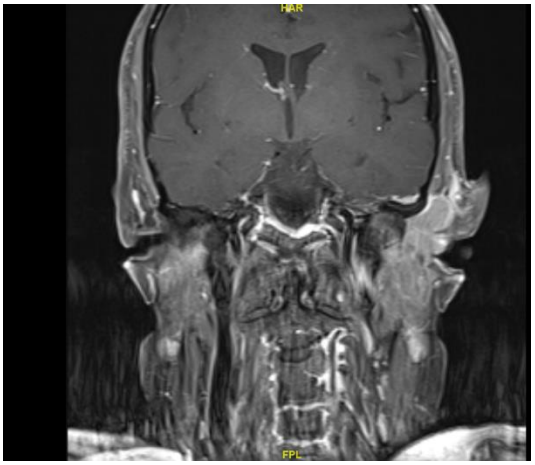

Figure 4 T1 with contrast showed high signal intensity mass lesion filling the external auditory canal and extending to supra and post auricular regions. High signal intensity intracranial on the same side but dura is intact.

At this stage, a high suspicion of lymphoma was reached, and ear exploration was done; Total excision of a well capsulated mass in the external auditory canal (Figure 5). The tympanic membrane was atrophic as it was pushed medially by the mass, it was removed and the middle ear was free of disease with healthy mucosa, exposure of the attic revealed a same mass type filling the attic and ingulfing the head of the malleolus and the body of the incus. The incudo-stapedial joint was dislocated, the head of the malleolus was chopped, and the incus was removed to achieve complete soft tissue removal. The attic roof was grossly intact.

The histopathology exam revealed focal presence of atypical lymphoid cells, the lack of germinal centers and the very high ki67 proliferative index were suggestive of Non-Hodgkin's lymphoma. However, the immunohistochemical findings were inconclusive and may even favor lymphocytoma cutis. Hence; the two specimens were sent to the pathology department of the University of Bern, Switzerland for final diagnosis, reported back as a diffuse infiltrate of small to medium and in part large sized atypical lymphoid cells. Whereas the small lymphoid cells in most part represent reactive lymphocytes, the atypical medium and large lymphoid infiltrate consist of lymphoid cells with a b-cell phenotype with expression of cd20, diffuse and strong expression of CD 10 and nuclear expression of bci-6 as well as bcl-2 and in-situ hybridization for EBV-RNA is negative. In conclusion an atypical b-cell lymphoid infiltrate, accompanied by a reactive t-cell population were observed.

Based on the morphology, the absence of clear follicular structures, the partly nodular, partly diffuse nature of the infiltration with obvious destruction of the underlying normal structures as well as the high mitotic and apoptotic rate, the diagnosis of an aggressive B-cell lymphoma was favored. In post-operative follow up, Pet-scan was done, and it was totally free except for small supra clavicular lymph node. The patient received 3 cycles of chemotherapy and 3 doses of radiotherapy. However, to date she is disease free.

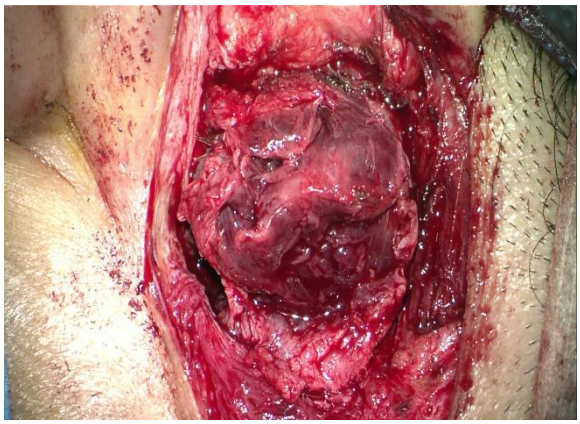

Figure 5 Well capsulated mass was totally dissected and removed.

3. DISCUSSION

Non-Hodgkin's lymphomas are lymphoproliferative disorder with diverse biological and clinical behavior (Ansell et al., 2015). Yet, it could present as a nodal mass, or it could have an extra-nodal presentation. Primary Extra-nodal disease is more common in Non-Hodgkin's lymphoma (NHL) than Hodgkin's Lymphoma (HL), as it accounts for 23% of NHL cases, whereas it accounts of only 4% of HL cases (Clark et al., 1983). Although the head and neck consider the second most common site of extra-nodal lymphomas after the abdomen; primary involvement of the ear is very rare. Diffuse large B-cell lymphoma is the most common type of NHL (Shen et al., 2018). Staging of the disease is determined by Ann Arbor staging classification, based on the number of involved lymph node and organ sites (Armitage et al., 2012). Most lymphomas in the ear & temporal bone are due to invasion from contiguous locations or metastasis foci. However, lymphomas that arise primarily from ear are very rare (Armitage et al., 2012). Therefore, clinical & histological features have not been extensively characterized (Dreyling et al., 2014).

Several factors have been associated with increased risk of middle ear Non-Hodgkins lymphoma, such as epstein–barr virus (EBV), human immunodeficiency virus (HIV), irradiation, immunosuppression and organic toxins (Pasha et al., 2018, Zapater et al., 2010). Few studies have linked hepatitis c virus with increased risk of few lymphoma subtypes (De Sanjose et al., 2008). Our patient has not been investigated for hepatitis c virus antibodies and was HIV- and EBV- negative. Symptoms commonly associated with temporal bone neoplasms tend to be site specific. The first one who described the clinical picture of middle ear lymphoma was Malick and colleagues (Malik et al., 1979). Patients may present with any combination of typical ear complaints, with the most common being recurrent otitis media, otalgia, conductive hearing loss and Facial nerve involvement (Fish et al., 2002; Merkus et al., 2000).

Our patient presented with otalgia and hearing loss but did not have any degree of facial palsy. Otorhinolaryngologists should consider the diagnosis of Non-Hodgkin's lymphoma of the head & neck when clinically and radiologically suggested, but a definite diagnosis has to be histopathological. Extra nodal lymphoma is extremely rare, with GIT being the most common extra-nodal site (Pasha et al., 2018). Moreover, a retrospective studies has been done and reports that incidence between 1.9 to 11.4 cases per year (He et al., 2009; Picard et al., 2015). We found only 31 reported cases of primary lymphoma in the ear in the international literature, Men (64%) being more affected than women (36%). The age of diagnoses ranged from 2 – 83 years with a tendency for paediatric and elderly populations (Alexander et al., 2007).

The diagnosis of B-cell lymphoma is made by either Fine-needle Aspiration which may differentiate B-cell from T-cell lymphoma, or by open biopsy which is usually done to determine the diagnosis prior to treatment. However, other studies such as CBC, BUN, LDH, CT/MRI of neck and chest and bone scan are essential in the diagnosis of any type of Lymphoma (Pasha et al., 2018). Primary management of diffuse large b-cell lymphoma is CHOP such as Doxorubicin, Vincristine, Cyclophosphamide, Prednisone, followed by radiotherapy agents to involved sites. However, low-grade B-cell lymphoma and MALT syndrome may be observed (Pasha et al., 2018; Fish et al., 2002).

The overall 5-year survival rate of H&N in general has improved to nearly 62%. However, in NHL grading is important in the evaluation of the prognosis of the disease. Smaller and nodular masses are associated with better prognosis. On the other hand, larger, diffused, higher grading, and CNS or BM involvement are associated with better prognosis (Pasha et al., 2018).

4. CONCLUSION

The incidence of Non-Hodgkin's lymphoma is rare in the ear. To our knowledge, it's the first case of two isolated masses occurring in the external auditory canal and the middle ear cleft on the same ear simultaneously. However, the presence of benign or malignant tumors should be excluded in the ear infections which are unresponsive to Antibiotic by imaging. Yet, a biopsy should be obtained for the optimum goal of confirming the diagnosis. Moreover, early diagnosis and treatment is the main stay cornerstone for good prognosis and outcome in primary ear lymphoma.

Acknowledgement

We thank the participants who were all contributed samples to the study.

Informed consent

Written & Oral informed consent was obtained from all individual participants included in the study. Additional informed consent was obtained from all individual participants for whom identifying information is included in this manuscript.

Funding

This study has not received any external funding.

Conflicts of interest

The authors declare that there are no conflicts of interests.

Data and materials availability

All data associated with this study are present in the paper.

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