Case Report

Primary Non-Hodgkin Lymphoma of the temporal bone: A rare case report

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ABSTRACT

Introduction: Primary lymphoma of the temporal bone is extremely rare, difficult to diagnose and to manage. It is essential that the clinician keeps in mind the possibility of this pathology as a differential diagnosis with the infections resistant to the usual treatment.

Case report: We report a rare case of a diffuse large B-cell lymphoma in a 70-year-old woman, with history of diabetes. The pathological study was in favor of a Non-Hodgkin Lymphoma of the Temporal Bone.

Discussion: Lymphomas defined as malignant monoclonal proliferation of lymphoid cells, are not uncommon in the head and neck region. Literature presents with few cases.

Conclusion: The aim of this article is to report a rare case of a diffuse large B-cell lymphoma with primary mastoid and external auditory canal infiltration without systemic involvement initially presented as a benign ear infection.

1. Introduction

Lymphomas are the second most frequent malignant tumor with an incidence of 2.5% in the head and neck region after squamous cell carcinoma [1]. Knowing that malignant involvement of the temporal bone is rare with an incidence of less than 0.2% among all head and neck cancers and that some authors have reported the involvement of the temporal bone by generalized lymphoma [2]; primary involvement of temporal bone without systemic involvement is broadly rare [3]. Only few cases of primary involvement of the temporal bone by lymphoma were reported in the past 20 years of which only 3 cases have been reported in the last year [4,5]. Lymphomas can be widely classified as Hodgkin’s lymphoma (HL) or non-Hodgkin’s lymphoma (NHL). NHL can be sub-categorized into B-cell lymphoma, T-cell lymphoma, or natural killer (NK) cell lymphoma [6].

The aim of this article is to report a rare case of a diffuse large B-cell lymphoma with primary mastoid and external auditory canal infiltration without systemic involvement initially presented as a benign ear infection. We present a case in accordance with SCARE 2020 criteria [23].

2. Case report

We report a case of an 70-year-old woman with history of diabetes. She was presented to the otolaryngology department with a House-Brackmann grade 5 right facial nerve palsy with a 2 weeks history of right postauricular pain and progressive unilateral hearing loss associated with a fever of 38.5 °C. Clinical examination showed a firm scalp swelling and nontender and margins could not be well identified. Oto-scopy revealed a right inflammatory polyp that completely obstructs the external auditory canal. There were no disorders of the balance notably no spontaneous or provoked nystagmus; also there was no cervical lymphadenopathy.

White blood count was elevated with 16,030 (80% neutrophils, 13% lymphocytes, and 7% monocytes). Computed tomography (CT) of the temporal bones showed a homogenous soft tissue density measuring 60 Hounsfield units, with pseudopagetooid reorganization of the texture of the whole of the petrous bone, with osteolysis of the walls of the middle ear, bone erosion of the second portion of the facial nerve, filling of the mastoid cells, of the external and middle ear. Retention of fluid in the right mastoid air cell system and the middle ear was noted. Noradiological sign raising suspicion of cholesteatoma was seen. (Fig. 1). Magnetic resonance imaging (MRI) demonstrated a heterogeneous enhancing process at the level of the temporal bone (Fig. 2). The lesion was hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging. A large intermediate signal petrous apex mass with extension into the right middle ear cavity and attic along with fluid...
Retention in the mastoid air cells was confirmed on the MRI. No lymph node involvement was present on either CT or MRI.

Intravenous antibiotics and topical ciprofloxacin/dexamethasone drops were continued with no improvement. We carried out an excisional biopsy under local anesthesia of the inflammatory polyp. The histologic examination of the mass revealed a peripheral B-cell NHL: diffuse large cells 60% and follicular 40%, with an immune-proliferative activity (Ki-67 index) of 60%. The immunohistochemical study showed the negativity of CK, EMA, PT, PB, and the positivity of CD79a.

Assessment of extension made of cervico-thoraco-abdominal scanner and cerebral scanner did not show any lesion making suspect other localizations of lymphoma. The patient was subsequently referred to the Hematology department for osteomedullary biopsy and treatment. The rituxan, cyclophosphamide, doxorubicin, vincristine, and prednisone chemotherapy protocol were commenced with subsequent positive clinical response after the initial treatment.

At two years, follow-up evaluation showed stable complete remission within localized recurrence or any systemic involvement. Otoscopic examination did not reveal any anomaly but the hearing loss remained stationary.

3. Discussion

Lymphomas defined as malignant monoclonal proliferation of lymphoid cells; are not uncommon in the head and neck region (incidence of 2.5%) [2], and involve Waldeyer’s ring, nasal cavity, paranasal sinuses, salivary glands, thyroid gland, and orbit [7]. The mastoid, middle ear most commonly, and the external/internal auditory canals are involved in a lesser degree [8]. All ages can be affected but this malignity but it may have a tendency to appear in adult onset and with male predominance (male-to female ratio ranges from 1.5 to 2:1) [1]. Some authors reported that typically the most common symptom of primary lymphoma of the temporal bone is conductive hearing loss, however sensori neural hearing loss occurs when cranial nerve VIII is involved [9]. Other reported that the most common initial symptom is localized pain, secondarily patients may present with a palpable mass, or rarely facial nerve palsy [10–13], this is due to the fact that the nerve sheath is resistant into tumor invasion and occurs when the bony facial canal is broken by the tumor and nerve fibers are involved by the tumor cells [14]; and affect usually the geniculate ganglion [15]. Therefore all of these malignant disorders are misdiagnosed and treated as external otitis. In our particular case, the right facial nerve palsy was due to erosion of the second bony segment of the right facial nerve canal.

HRCT play an important role in the diagnosis [16], it shows extensive irregular permeative osteolytic destruction of the temporal bone with soft tissue opacification of the mastoid air cells and erosion of the intercellular septae [2]. This osteolytic lesions of the temporal bones should be considered with a high degree of suspicion for malignancy in general or aggressive infective pathologies [17]. There by, HRCT and contrast enhanced MRI scans are important for a better evaluation of malignancies involving the temporal bone [2]. HRCT provides details on the type of bone destruction, middle ear condition, ossicular chain evaluation, exact site/extent of the intratemporal bony facial canal and the integrity of the tegmen tympani. The main role MRI is to identify meningeal and brain parenchymal invasion, to give detailed mapping about the entire extent of extracranial soft tissue involvement as well as the perineural spread on the facial nerve [18,19]. Therefore, any abnormal radiologic findings that are suspicious for a neoplasm should be biopsied. The cells of primary bone lymphoma may be mixed in appearance so that osteomyelitis is suspected [20].

Delgado and al [21]. described that a single biopsy could not be sufficient in some cases. Thus, Fish and al. [22], reported that incisional biopsy may cause a misdiagnosis or a delay in diagnosis.

4. Conclusion

Lymphoma of the Temporal Bone is extremely a rare tumor in current practice, it is also a difficult tumor to be diagnosed, beside that Literature presents with few cases. It is a diagnosis to think of him face to an infection resistant to the usual treatment. The diagnosis is confirmed after a biopsy with anatomopathological examination.

Ethical approval

I declare on my honor that the ethical approval has been exempted by my establishment.

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Author contribution

Chaker Kaoutar: Corresponding author writing the paper.
Ahmed Brahim Ahmedou: Corresponding author writing the paper.
Yousef Oukessou: study concept.
Sami Rouadi: study concept.
Redallah Abada: study concept.
Mohamed Roubaï: correction of the paper.
Mohamed Mahtar: correction of the paper.

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Guarantor

DR AHMED BRAHIM AHMEDOU.
DR CHAKER KAOUTAR.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

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Declaration of competing interest

The authors declare having no conflicts of interest for this article.

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