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Histiocytosis Otomastoiditis X: About a Clinical Case

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Abstract: Langerhans cell histiocytosis (LCH) or histiocytosis X refers to a group of diseases that are characterized by clonal proliferation of histiocyte with similar characteristics to Langerhans cells, which affect different organs of the human body, including the temporal bone. The present study describes the clinical case of a 42-year-old patient admitted to surgery for acute chronic otomastoiditis on the right temporal bone. Axial Computerized Tomography (CT) revealed petromastoid osteolytic lesions, dehiscence of the lateral sinus and external semicircular canal and a solution of continuity of the tympanic tegmen, but without infiltration of neurological structures. The diagnosis of temporal bone LCH was confirmed with histopathological and immunohistochemical exams. Depending on the type of lesion, there are different therapeutic modalities for LCH of the temporal bone that include surgery, chemotherapy, radiotherapy and even corticotherapy. In this case, the patient was submitted to mastoidectomy the right, followed by corticotherapy (dexamethasone) and chemotherapy (vinblastine). Up to date, he has not yet had any relapse, but is still on follow up.

Keywords: Langerhans Cell Histiocytosis, Temporal Bone, Langerhans Cell

1. Introduction

Langerhans Cell Histiocytosis (HCL) or Histiocytosis X designates a group of diseases that are characterized by the clonal proliferation of histiocytes phenotypically similar to Langerhans cells. It is a rare disease with unknown etiopathogenesis. Genetic, immunological or infectious factors, of viral nature, may predispose to its appearance, but there′s not enough evidence [1]. The estimated annual incidence is approximately 5 cases per million people, mainly in males and pediatric disease age groups, with a peak incidence between 12 months and four years of age [1, 2], so the therapeutic protocols of adults are based on those developed for pediatrics [1, 9, 13].

The clinical presentation, may involve any organ or system. Bone involvement is the most common form of presentation, being in unifocal adults or multifocal children [1, 2, 9] It mainly affects the bones (80% of cases), preferably the cranial ones, with lesions predominantly lytic [10].

According to current recommendations from the Histyocite Society, the spleen, liver, bone marrow, lung (excluding isolated lung involvement) or hematopoietic organs are considered at risk. Anatomical structures are risk sites, which, when affected, may cause morphological and functional alterations the central nervous system [1, 9].

The diagnosis of LCH is based on the immunohistochemical result of the affected tissue samples, with the identification of CD1a and/or CD207 (langerin) markers. The identification of the S-100 protein is suggestive of HCL, but it does not allow diagnosis because it is unspecific [9, 10, 11, 12]. In the temporal bone, lytic lesions on Computed Tomography (CT) contribute to the suspicion. The treatment shall be based on the site and extention of the disease, taking into account the stratification of patients according to clinical risk [2, 7, 12].

The authors present in this article a clinical case of rigth

chronic otomastoiditis by Langerhans cell histiocytosis, with extensive petromastoid lytic lesion with bone continuity solution between the external auditory canal and the posterior fossa, in a 42-year-old patient, as well as a brief literature review the literature on subject.

2. Clinical Case

P. J. M. P., 42 years old, male, Caucasian, smoker, with history recurrent suppurated repetition in childhood, but without complaints in adulthood. In 2017, episodes of intermittent, foul-smelling otorrhea began in the right ear, progressively worsening hearing loss and vertigo with peripheral characteristics, complaints that remitted with systemic and topical antibiotherapy (ceftriaxone and ofloxacin). He was not referred to Otorhinolaryngology until May 2018, when he was attended at the Emergency Department of the Lisbon Hospital Center, due to an exacerbation of the complaints, having been referred to the Otorhinolaryngology service of the Hospital Center of Setúbal Center with diagnostic hypothesis of Acute Right Chronic Otomastoiditis with a possible neoformative lesion and under therapy of ceftriaxone, metronidazole and indomethacin.

At the objective examination, on the right otoscopy only hyperemia of the posterior wall of the external auditory canal was observed, with filling by polypod lesion and purulent otorrhea.

Of the complementary exams, we highlight the Simple Tonal Audiogram on the right with transmission hearing loss of 5-45 dB under 2Khz and sensorineural over 2Khz frequencies with descending curve to 70 dB at 8Khz (Figure 1). The CT of the ears revealed an extensive right petromastoid osteolytic lesion, with wide bone continuity solution between the external auditory canal and the posterior fossa, with involvement of the sinodural plate and the vestibule aqueduct, dehiscence of the lateral sinus and the external semicircular canal, solution of continuity of the tympanic tegmen and otomastoid filling, but without infiltration of the neurological structures (Figures 2 and 3).

At the end of May 2018, he underwent right mastoidectomy to collect specimen for pathology since the observable lesions were only inflammatory polyps, with complete filling of the mastoid by a friable and bleeding polypoid mass which anatomopathological result was inconclusive. (Figure 4). In view of the aggressive and progressive characteristics of the lesion, and given the strong suspicion of malignancy, the patient was referred to an Otorhinolaryngology appointment at the Portuguese Institute of Oncology in Lisbon.

In that institution, in August 2018, he underwent right subtotal petrosectomy, posterior labyrinthectomy and radical mastoidectomy with placemente of abdominal fat graft to close the wound (Figures 5 and 6). The postoperative period was uneventful, the pathological anatomy of the tissue removed intraoperatively revealed that it was a Langerhans cells histiocytosis, confirmed by immunohistochemistry with positive S100 and CD1a markers (Figures 7 a, b, c).

The patient was followed up by Hematology to screen for cutaneous complications of the disease, due to the appearance of multiple scaly lesions on the posterior surface of the left leg. Postoperative examinations carried out to control the disease after one year revealed expectable sequelae, showing no new alterations or signs of recurrence, so the patient remains under observation and therapy with dexamethasone and vinblastine.

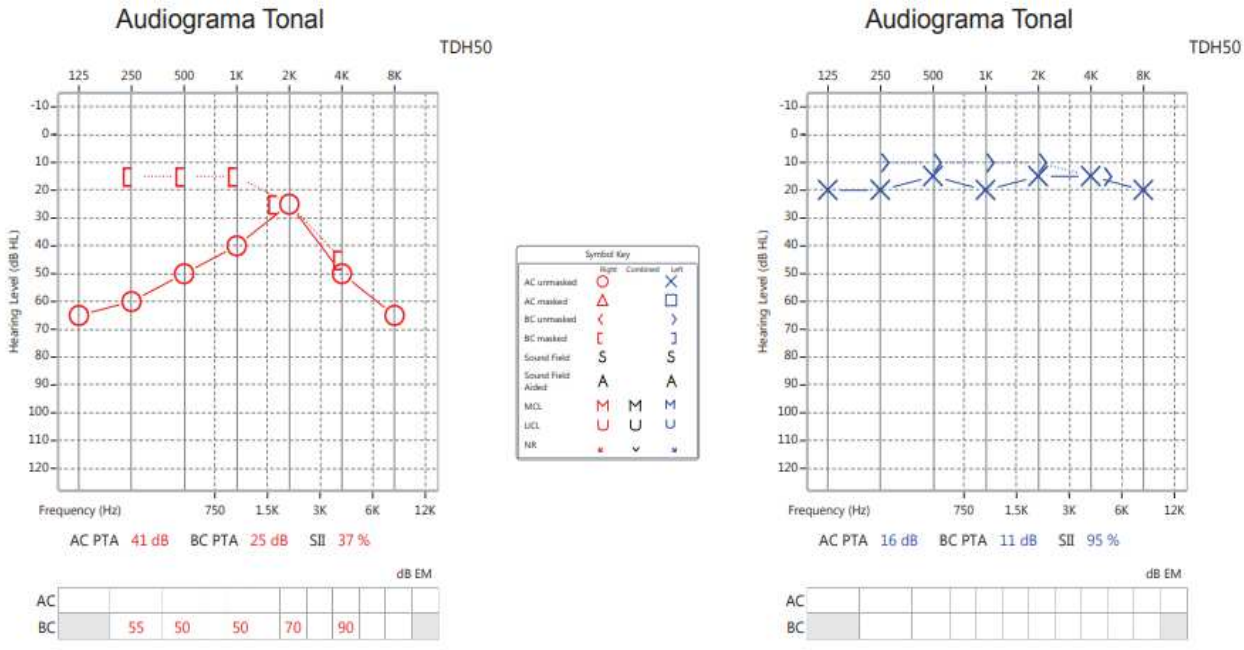

Figure 1. *Simple Tone Audiogram. Right ear: Transmission hypoacusis of 5-45 dB under 2Khz and sensorieneural over 2Khz with a descending curve to 70dB at 8 Khz. Left ear: Normal.*

Preoperative CT.

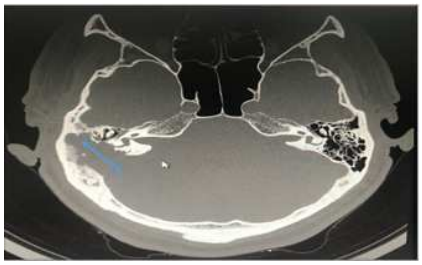

Figure 2. *Section axial view shows right petromastoid osteolytic lesions, with coalescence of mastoid cells and plaque erosion synodural (blue arrow).*

Figure 3. *Coronal section, with tympanic tegmen dehiscence and signs of otomastoiditis (blue arrow).*

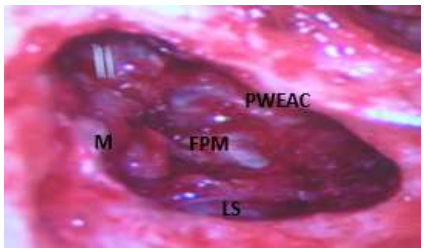

Figure 4. *Intraoperative right mastoid cavity. PWEAC: Posterior Wall of the External Auditory Canal. M: Meninge. LS: Lateral Sinus. FPM: Friable Polypoid Mucosa.*

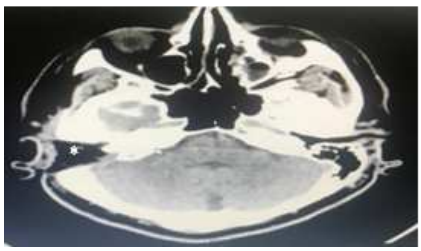

Figure 5. *Ear CT. Empty cavity of right post-subtotal petrosectomy (asterisk).*

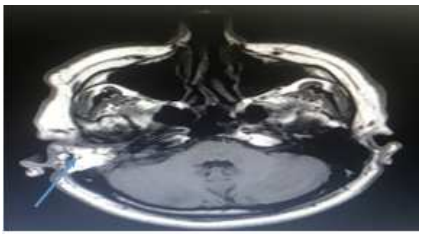

Figure 6. *Magnetic resonance imaging of the ears. Post subtotal petrosectomy status on the right, with filling of the emptying cavity by fat and muscle (blue arrow).*

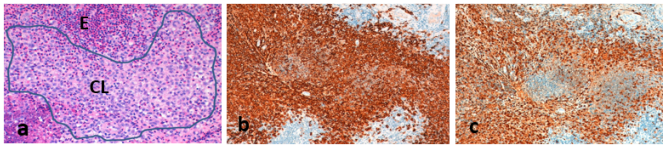

Figure 7. *a. Diffuse proliferation of Langerhans cells (cells with clear eosinophilic cytoplasm, with thin chromatin nuclei, indented and with slits), accompanied by an abundant population of eosinophils (E). b and c. Immunohistochemical studies showing positivity for S100 and CD1a cells.*

3. Discussion

LCH also known as Histiocytosis X, Letterer-Siwe, Hand-Schuëller-Christian disease or eosinophilic granuloma is characterized by neoplastic proliferation of histiocytes and produces clinical syndromes with varied manifestations [1, 2, 4, 6, 7]. It presents itself in several clinical forms, which can be unifocal or multifocal or even multisystemic. Skull bones are frequently affected, and according to the literature, temporal bone involvement is present in 14 to 61% of the cases; being bilateral in 30% of those [1, 2, 4, 6, 7, 9]. Skin lesions are observed in 50% of cases, and this may be the only manifestation of the disease associated with systemic involvement [1, 2].

Studies performed by CT have greater sensitivity for assessment of the extention and progression of the disease[2]. The diagnosis is based on clinical manifestations and histopathological and immunohistochemical evidence. Histologically, it is characterized by inflammatory infiltrate, with eosinophils, histiocytes (sometimes forming multinucleated forms), neutrophils and small lymphocytes, where Langerhans cells are observed, which vary in size from 10 to 15 µm and are recognized by their nucleus, with indented slits and vast, eosinophilic cytoplasm [1, 2, 3, 4, 5, 6, 7]. The diagnosis is based on the identification of typical cell morphology, but the definitive diagnosis is based on positivity of the lesional cells for CD1a and/or langerin (CD207). Birbeck granules (rod-shaped cytoplasmic structures with a vesicle on its end, resembling a tennis racket), are old criteria based on electron microscopy, so aren′t recommended, as langerin confirms the presence of them. LCH cells are also positive for the S100 protein, but because other types of macrophages and histiocytes can also stain with this marker, this criterion is not sufficient for diagnosis [1-11].

There is some controversy in the treatment of this disease, particularly in cases where the manifestations are mainly located in the head. The involvement of the temporal bone is easily confused with other inflammatory, infectious and neoplastic lesions of the ear, often causing delay in the diagnosis and treatment of the pathology [1, 2, 7].

The main objectives of treatment are centered on improving symptomatic manifestations and preventing complications [2]. There are several therapeutic approaches to temporal bone LCH, ranging from surgery, corticotherapy, chemotherapy and radiotherapy in focal lesions [1-7].

Most publications advocate surgery in combination with

another form of treatment, particularly chemotherapy, as the best solution [1, 2, 5]. It is important to verify whether ear or temporal bone involvement is isolated or is a manifestation of systemic disease [1, 2]. About 10 to 20% of patients show spontaneous regression of the disease [5]. Being a disease with a wide variety of clinical evolitions that can range from spontaneous remission to severe fatal disease, in the absence of reaching organs considered at risk (liver, spleen, lungs, hematopoietic system) the prognosis is generally favorable [1, 7].

Treatment depends on the presentation and extent of the disease. The Histiocyte Society showed that the therapeutic protocol carried out in a period of 12 months reduces the rate of recurrence of the disease [12]. Patients with or without involvement of risk organs may have a variable clinical evolution. Those with multifocal bone lesions are known to have favorable prognosis (100% survival), but with high tendency for disease recurrence (30-50%) and permanent sequelae. The same is true on patients with involvement of special sites (vertebral lesions with intraspinal soft tissue extension) and central nervous system risk lesions [12-15].

The last protocol of the Histiocyte Society advocated the association of prednisolone and vinblastine as an effective treatment with a minimal rate of toxicity; therefore, it is the standard initial therapy for all patients in whom systemic therapy is indicated [12-14].

In the case described by the authors, the therapy consisted of a combination of surgery followed by corticosteroid therapy (dexamethasone) and chemotherapy (vinblastine) on an outpatient regímen basis. This option was chosen based on the location and limited extention of the disease, as in the case of multicentric lesions, treatment should be more aggressive. The patient is followed up on Hematology appointments for clinical and laboratory control of the disease evolution and screening for possible recurrences or new manifestations [12-15].

4. Conclusion

The etiology of Langerhans Cell Histiocytosis is still controversial; temporal bone involvement is present in about 4 to 25% of cases. Depending on the type of injury, treatment options include surgery, chemotherapy, radiotherapy and even corticosteroid therapy; The prognosis is variable from spontaneous remission to death.

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