Langerhans Histiocytosis of the Temporal Bone: A Case Report

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Abstract

Langerhans histiocytosis is a pathology characterized by clonal proliferation of Langerhans cells. It is a rare condition in children and young adults, with great variability in its clinical presentation and the organs it affects, bone involvement being the most common. Isolated temporal bone involvement remains rare and it is commonly misinterpreted as otomastoiditis, delaying diagnosis and appropriate therapeutic management. On imaging, it is characterized by lytic lesions of variable aggressiveness, which are not pathognomonic. Only the histological examination allows the definitive diagnosis to be made. Sectional imaging plays an important role in the diagnostic approach. We report a case of isolated involvement illustrating the diagnostic approach in imaging.

Keywords: Langerhans histiocytosis; Temporal bone; Retroauricular mass; Children; Imaging

Introduction

Langerhans histiocytosis is a pathology of the reticuloendothelial system characterized by clonal proliferation of Langerhans dendritic cells [1]. It is a rare condition in children and young adults, presenting a varied polymorphism, ranging from a simple eosinophilic granuloma to generalized, life-threatening multiorgan lesions [2]. Bone involvement is more common and imaging allows characterization of bone lesions as well as a diagnostic approach, although the definitive diagnosis is based on histological examination. We report this case in order to illustrate the interest of imaging in the diagnostic workup of this pathology in children.

Patient and Observation

This is a two-year-old girl who presented with painful, progressive left retroauricular swelling with otorrhea. A CT scan of the petrous pyramid of temporal bone was performed, showing a lesional process of the left petrous pyramid (Figure 1A,B); and a retroauricular soft tissue infiltration which is enhanced after injection of contrast product (Figure 2A), with extensive bone lysis (Figure 2B). Additional MRI imaging was performed, demonstrating a tissue-like mastoid filling without endocranial extension. The patient underwent a biopsy and the histology confirmed the diagnosis of Langerhans histiocytosis of left temporal location. The other paraclinical examinations, namely the abdominal ultrasound and the thoraco-abdomino-pelvic scanner were normal.

Discussion

Langerhans Histiocytosis (LH) is a group of conditions that have in common tissue infiltration by Langerhans cells or dendritic cells originating from the reticuloendothelial system and organized in to granulomas. It is a systemic disease of children and young adults, with a peak in frequency between one and three years, male patients being predominantly affected [3]. It is a rare condition, with great variability in its clinical presentation and the organs it affects. Its seriousness lies in the possibility of damage to certain organs called “at risk” and therefore the possibility of death or sequelae [4]. Bone involvement is the most common and is often monostotic rather than polyostotic, preferentially affecting the axial skeleton and primarily the skull, namely the vault and the base [5]. Temporal bone involvement has been described in 15 to 61% of all cases of Langerhans cell histiocytosis [2]. Most authors state that otologic involvement generally occurs during multisystem involvement, while it remains isolated in 5 to 25% of patients [6]. The lesions of the temporal bone are located more in the mastoid and in the middle ear than at the tip of the petrous pyramid.

Clinically, Langerhans histiocytosis is characterized by its polymorphism, which often delays diagnosis. The most common otologic symptom is drug-resistant otitis media and retroauricular swelling. The appearance on otoscopy is similar to that of otomastoiditis, this also contributing to the diagnostic delay [6].

Computed tomography is a very sensitive examination in the exploration of temporal involvement and is used for its spatial resolution to analyze the extent of the lesions and to guide the bone biopsy necessary for diagnosis [7,8]. Langerhansian histiocytosis generally results in mastoid tissue filling with osteolysis of the petrous pyramid extending to the ear drum, and is accompanied by damage to the soft tissues, which is enhanced after injection.
of contrast product [6]. However, involvement of the ossicular chain and the inner ear is not as common as might be expected due to the extensive bone damage usually seen. This typical aspect was found in our patient. Magnetic resonance imaging, because of its multiplanar capacity and its high contrast resolution, makes it possible to better specify the extent of the tumor as well as its extension in the soft tissues adjacent to the bone such as the brain temporal lobe and the ponto-cerebellar angle [7,8]. Thus the mass appears as a discrete T2 hypointense and as an intermediate T1 signal, with a clear uptake of contrast after administration of gadolinium [9].

The main radiologic differential diagnosis of such an aggressive osteolytic lesion is rhabdomyosarcoma, although the soft tissue component is larger there. An advanced cholesteatoma is less of a diagnostic trap because of the mismatch between the petrous involvement and that of the ossicles, which is moderate or absent. Other differential diagnoses include mastoiditis, lymphoma and metastases [9,10]. However, extensive osteolysis as well as extensive soft tissue damage is usually not seen in mastoiditis. Metastases, as with rhabdomyosarcoma, should be considered in the presence of neurological manifestations such as deafness, vertigo or damage to the cranial nerves which rarely occur in histiocytosis [11].

Thus the radiological appearance of Langerhans histiocytosis is not always typical, even typical, it is not pathognomonic. Only histological examination can make the definitive diagnosis. Its discovery requires a radiological extension workup in order to assess the degree of dissemination of the condition and to better specify the therapeutic indications.

These are skeletal x-rays or even cross-sectional examinations, targeted according to clinical or biological evocative signs, namely a cervico-thoraco-abdomino-pelvic scan, a brain MRI, an ultrasound or even a hepatobiliary MRI. Bone scintigraphy has a debated role, because while it allows an examination of the whole body, it is less sensitive than x-rays, except for certain locations such as the base of the skull and of course does not allow an accurate anatomical analysis.

Conclusion

Langerhans histiocytosis is renown by its clinical polymorphism. The bone involvement, particularly of the temporal bone is characterized by lytic lesions of varying aggressiveness. Sectional imaging plays an important role in the diagnostic approach. Computed tomography confirms the presence of osteolysis, assesses the extent of the lesion and helps in biopsy guidance. In addition, MRI can be used to determine the extent of the tumor and its intra-cranial extension.

Conflicts of Interest

We declare not to have any conflicts.

Author’s contribution

Germaine NIYITANGA, MD: Conception of the work, Drafting the work, Author corresponding
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