International Journal on Oncology and Radiotherapy

Langerhans Cell Histoctiosis (LCH) of the Temporal Bone Misdiagnosed as Parameningeal Rabdomyosacroma: A Case Report

Case Report

Volume 5 Issue 1- 2024

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Article History

Received: January 02, 2024 Accepted: January 18, 2024 Published: January 18, 2024

Summary

LCH is a rare disease seen at age but mostly affecting young children. Its may present with different clinical presentation. Among flat bones skull and especially the temporal bone is the most affected. Early diagnosis of the disease with appropriate radiological exam and subsequent biopsy is very important to begin treatment to avoid further spread of the disease into the cranial fossa. Systemic chemotherapy (vinblastine and prednisone) is required and was effective in our case

Keywords: Langerhans Cell Histocytosis (LCH), Computed tomography (CT), Magnetic resonance imaging (MRI), Temporal bone, Head and Neck

Introduction

Langerhans cell histiocytosis (LCH) is a very rare disease caused by the abnormal proliferation of histiocytes like cells. Its incidence is 8-9 cases/million in the year and affects all age groups mostly in infants between 1-3 years old. [1,2] It may be a single system or multi system disease that can be seen as the solitary lesion to severe multifocal disease. Diagnosis of the disease is done by a correlation of the clinical presentation of the disease with the imaging and the most important is histopathologic features [2]. Most of the cases are presenting as uni or multifactorial bone lesions, visceral organs involvement is less seen [3,4]. Lung, liver, skin, pituitary gland mucous membranes and lymphatic system are among the other affected organs [5]. We report an LHC in a 3 years old female in the right temporal bone that was misdiagnosed as lymphadenopathy in the first clinical examination and diagnosed as rhabdomyosarcoma in contrast-enhanced magnetic resonance imaging (MRI) exam. The diagnosis was confirmed after fine needs aspiration biopsy and histopathology that show LCH.

Case Presentation

A 3 years old female patient admitted to our clinic with swelling and tenderness in the right jaw. Her complaint did resolve although she used antibiotics and analgesics prescribed 15 days before by another medical centre. Physical examination revealed a swelling in the right temporal region upper to right tempormandibular joint measuring 3x2cm, with palpable right posterior auricular and submandibular lymph nodes. No, another pathology was noted during the physical examination. No history of previous trauma and no significant family history. Blood investigations, C-reactive protein, urine analysis and liver function tests were normal. USG examination showed irregular hypoechoic lesion superior to the upper part of the right parotid gland measuring 40x18x13 with minimal vascularization and right submandibular and superior cervical LAP the largest measuring up to 21x9 with cortical thickening and thin hilus. Brain computed tomography (CT) scan showed a destructive lytic lesion with soft tissue component in the right temporal bone. (Figure 1) Contrast-enhanced brain and

maxillofacial MRI exams demonstrated an infiltrative solid lesion in the squamous part and greater wing of the right temporal bone, extending into the temporal muscle, measuring 15mm thick and 25mm in its extra-cranial component, invasion and infiltration of the meningeal and the surrounding tissues with necrotic non- enhanced foci and infiltration of the surrounding soft tissue mass.

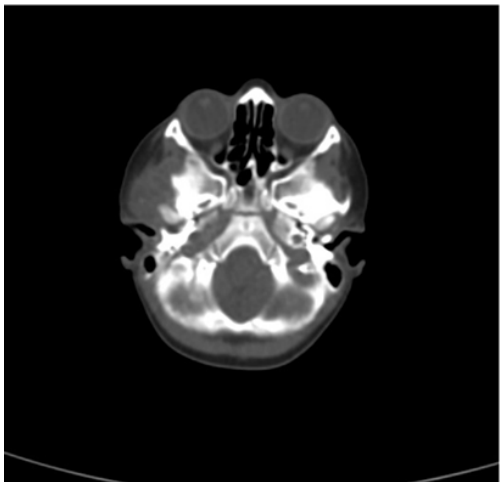

Figure 1: Axial brain CT scan shows a lytic lesion in the right temporal bone with soft tissue component.

External auditory meatus, temporomandibular joint are invaded and destructed by the lesion. Few lymphadenopathy are seen around the parotid gland and in the submandibular region with no evidence of parotid gland invasion. Petrous bone and cavernous sinus are normal. Correlation of the patient age and location parameningeal rhabdomyosarcoma was considered as the first differential diagnosis. (Figures 2,3) No other intracranial lesion was seen. An ultrasound-guided fine needle biopsy was to confirm the diagnosis of Lang- perhaps cell histiocytosis (LCH) was made seeing Fragmented tumour tissue composed of cells with indented nuclei and eosinophilic cytoplasm (HEx40) Langerhans cells which are polygonal cells with eosinophilic cytoplasm and oval nuclei some with longitudinal grooves, and accompanying eosinophils plasma cells and neutrophils. CD1a and Langerin expression in tumour cells were evident (Figures 4-6).

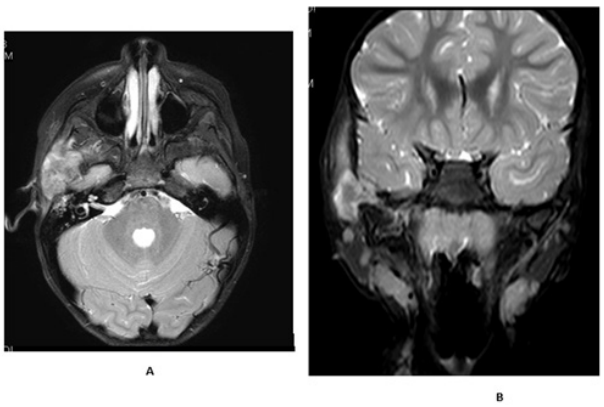

Figure 2:

A. Axial

B. Coronal

T2 weighted MR images showing a heterogeneous hyperintens lesion with soft tissue component in the right temporal bone

PET CT was done for the staging of the disease that showed a soft tissue mass measuring 31x2mm in size with calcification invading the right temporal bone and right lateral sphenoid wing showing pathologic 18F-FDG fixation (SUV maks: 5). Minimal soft tissue 18F-FDG fixation (SUV maks: 7.8) in the nasopharynx properly due to inflammation (Figure 7). The patient was diagnosed as LCH-IV (Single system multiple involvements). Vinblastin and prednisolone protocol was given. A decrease in swelling was seen after 4 weeks. The patient continues to undergo follow-up and no evidence of active disease has been observed.

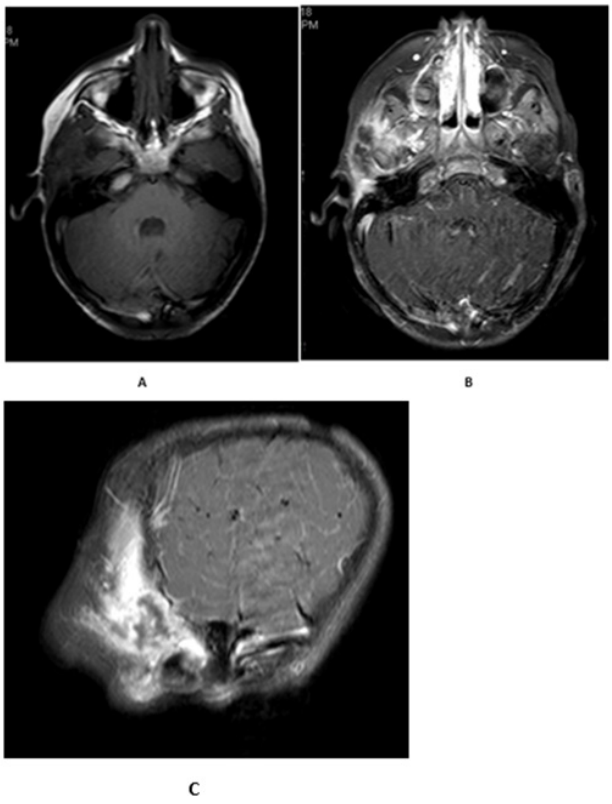

Figure 3: (A) Pre-contast axial T1 weight image shows hypointens lesion in the right temporal bone which Show heterogenous contrast enhancement in post- contrast axial and sagittal T1 weighted image (B ,C)

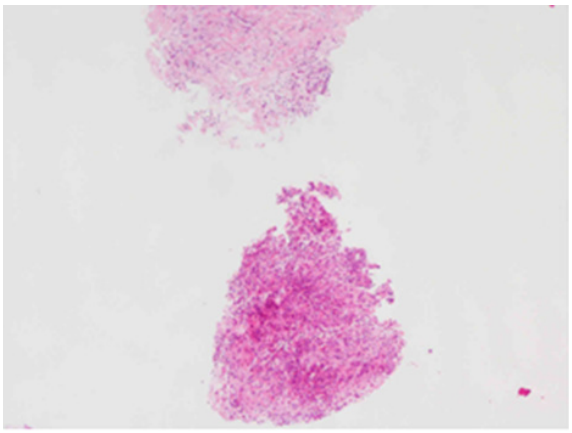

Figure 4: Fragmented tumor tissue composed of cells with eosinophilic cytoplasm (HEx40)

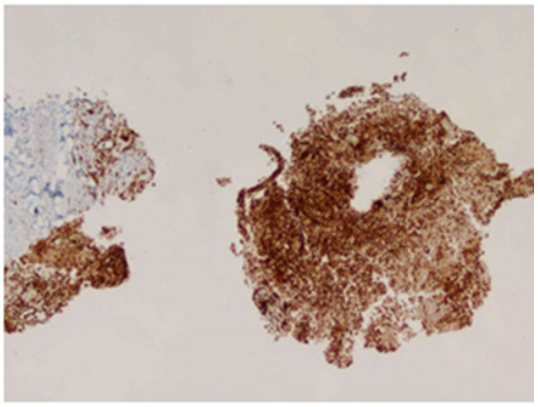

Figure 5: CD1a and Langerin expression in tumor cells.

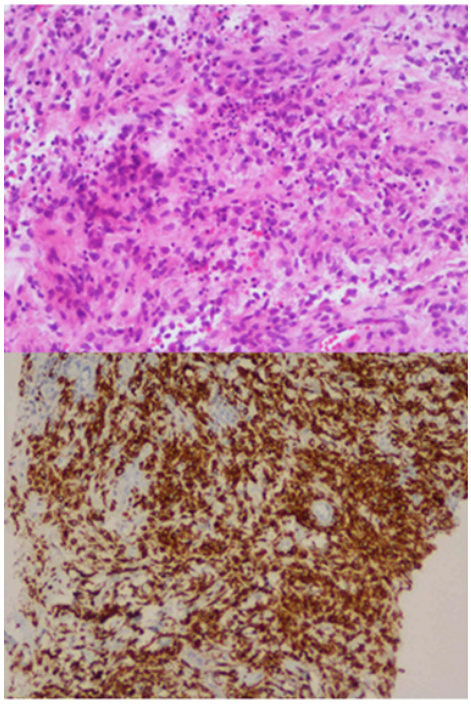

Figure 6: Langerhans cells which are polygonal cells with eosinophilic cytoplasm and oval nuclei some with longitudinal grooves, and accompanying eosinophils and neutrophils (HEx200).

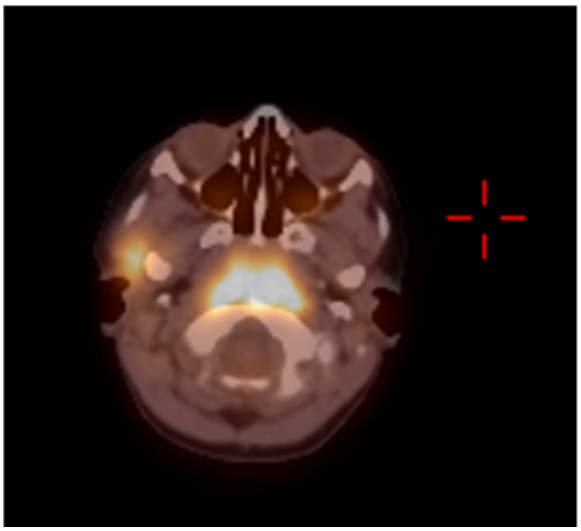

Figure 7: PET CT scan shows a soft tissue mass in the right temporal bone and right lateral sphenoid wing with pathologic 18F-FDG fixation (SUV maks: 5) .Minimal soft tissue 18F-FDG fixation (SUV maks: 7.8) in the nasopharynx properly due to inflammation.

Discussion

Langerhans cells histiocytosis was previously named as histiocytosis. It a disease with unknown etiology although some reported that it due to uncontrolled proliferation of Langerhans cell histocytes like cells and eosinophils as a result of genetic mutation and infections [5-7]. It has three subtypes, uni focal, multifocal and systemic (Abt-letterer site disease). Affected age group ranges from 5-15 years old with morbidity 5-6 per millions [2, 5]. The unifocal form is the most commonly seen form (70% of all patients) which is usually seen between 5 - 15 ages and is limited to single bone or lung. The multifocal form is also called the chronic recurrent (seen in 20% of all patients) affects bone and other reticuloendothelial systems like (liver, spleen, skin, lymph nodes) and it affects ages between 1-5. The least seen form is the fulminate form (The multifocal multi-system) and it is fetal affecting 10% of patients. It's seen in first two years old age in which involvement of the reticuloendothelial system with anaemia and thrombocytopenia [5-7].

Involvement of organs like lung, spleen, liver and hemopoietic system the disease is considered as a high-risk disease [2-5]. About 50% of the previously reported cases were a single systemic disease which is mostly confined to bone although one skin and lymph nodes involvement were reported too. The bony lesion is seen especially in flat bones (skull, mandible ribs pelvis and spine) in which lytic lesions are the most common radiologic presentation [5]. Skull, head and neck were reported to be involved in 60% of the previous cases. Mastoid bone is the most affected part of the temporal bone.When its affected by swelling pain dizziness and otorrhea are seen [6,7].

Clinical presentation and prognosis of the disease depend on the involved organs, patient age and organ dysfunction. It ranges from self-limited to fetal when multiple major organs are involved. Children less than 2 years old and multi-system are poor prognostic criteria [5,8]. Diagnosis is based on a combination of clinical presentation, radiologic features and the most import histopathologic and immunohistochemistry [2,7]. Lytic bone lesions are the most seen radiologic findings (seen in 80% of patients) The radiologic features of LCH in flat bones like in case of skull which is the most affected are seen as well define lytic lesion with (punched out appearance) and hole-hole appearance or bevelled edge can be seen due to asymmetric involvement of the bone sides of the bones. No periosteal reaction is present. In long bone beside lucent lesion, the new bone formation can be seen [8,5]. CT shows lytic lesions like x-ray, still, they can help in showing the nature of the lesion and its soft tissue component is present and evaluation of the bony structures in 3D reformats. Beside it can show the changes in the other organs.

Magnetic resonance (MR) imaging, helps in evaluating the soft tissue component the which appears hyperintense on T2-weighted images and hypointense-isointense on T1-weighted images, with heterogeneous enhancement in the post-contrast study [5]. Moreover, its shows relationship and extension of the lesion to surrounding soft tissue and vessel. PET CT show the systematic and the extent of distal disease spread and involvement of the other organ. It is the most functional imaging modality in multifiocal LCH and its used to follow up response to treatment [5]. Involvement of the temporal bone was reported in 15 -61% of all LHC [7]. Very few cases were reported in the brain and skull. In such cases osteoma, meningioma, EG and gliomas rhabdomyosarcoma, lymphoma and metastasis are among the differential diagnosis.

Lymphoma, leukaemia, multiple myeloma, and for multiple lesions should be considered in DDX of skull lesions. Epidermoid and dermoid cysts for single lesions must also be considered [9,10]. Here we reported the case of a 3-year-old female patient with as LCH-IV (Single system multiple involvements)in the right temporal bone that was initially rhabdomyosarcoma based on MRI imaging findings but was later diagnosed as LCH by histopathogic examination. Lack of pre-

vious trauma or accident history together with the lack of antibiotics treatment response like in our case need further radiologic examinations. Treatment of the disease depends on the type of the disease, patient age and the affected organ. For single skull or bone lesion curettage with local steroid injection and radiotherapy show to give result in previous cases. While multi-agent chemotherapy is used in the multi-systemic form of the disease [1,2,5,11]. In our case, A vindesine and prednisone-based regimen was selected for treatment and the patient was shown to progress well. However, the patient continues to undergo close follow-up to assess for signs of recurrence. Chemotherapy and oral steroid are used in multiple skull lesions to prevent CNS complications [6,9,10].

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