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RESEARCH ARTICLE

ATYPICAL LANGERHANS HISTIOCYTOSIS IN CHILDREN: ABOUT 2 CASES

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Abstract

Langerhans histiocytosis is a rare disorder with a heterogeneous clinical presentation. It is characterized by an abnormal proliferation of Langerhans cells in different tissues. The clinical manifestations depend on the number of lesions and their extension to the soft tissues. The organs affected by the disease, besides the head and the neck, may be the skin, long bones and the digestive system. Imaging confirms osteolytic lesions. The definitive diagnosis is made by biopsy with histological study.

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Introduction:-

Langerhan's histiocytosis is a rare disorder, characterized by an abnormal proliferation of Langerhans cells in different tissues. Clinical manifestations depend on the site and extension of the lesion. Otorhinolaryngologic with hepatic involvement and mandibular localizations are uncommon.

Imaging modalities show bone lesions. The definitive diagnosis is made by biopsy with histological study [1,2]. We report two cases of Langerhan's histiocytosis with mastoid, liver and mandible involvement.

Case Report:

Case report 1:

A 6 years old boy was hospitalized in pediatric department for a bilateral otalgia with purulent otorrhea, resistant to treatment.

The clinical examination found a patient in good general condition, febrile (38°C). The rest of examination revealed mastoid swelling and hepatosplenomegaly. At the otoscope: external auditory canal stenosis with invisible eardrum in both ears. Bacteriological culture was negative. The biological assessment showed an inflammatory syndrome with abnormal liver enzymes.

Computed tomography of the temporal bone shows bilateral mastoid osteolysis destroying the mastoid, extended to the temporal bone (Figure 1).

Abdominal ultrasonography revealed hepatomegaly with periportal hyperechoic images and an hypoechoic collection (Figure 2). We completed with an abdominal scan which shows diffuse periportal hypodensity with hypodense collection enhanced at the periphery after injection of contrast (Figure 3).

The diagnosis of histiocytosis was suggested and confirmed histologically after biopsy of the granulation tissue in external auditory canal.

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The treatment strategy used in this case was vinblastine and prednisolone association. After 3 weeks, the evolution was marked by regression of mastoid swelling, otalgia, otorrahebut there was no amelioration of the liver biochemistry.

Case report 2:

A 7 year old boy was referred to the pediatric department for bilateral jaw swelling evolving gradually since 3 months (Figure 4). Initially, the symptomatology was taken for a swelling of the parotid glands related to viral parotitis symptomatically treated by paracetamol without improvement. Oral examination found an ulceroproliferativemandible process. Cranio facial enhanced CT showed an extensive geographical osteolytic homogeneous mass destroying the mandible (Figure 5, 6). The diagnosis of histiocytosis was suggested and confirmed histologically after biopsy. Histological findings showed a granuloma made of clusters of vacuole dhistiocytes surrounded by lymphocytes and plasma cells.

Immunohistochemistry shows CD1a positivity A surgical curettage of the bone lesion was performed, with regimen of vinblastine and prednisolone. After 2 months, the evolution was marked by a clear regression.

Discussion:-

Langerhan's histiocytosis (LCH) is a rare disease characterized by a proliferation in different dendritic cell tissues belonging to the line of Langerhans cells. The physiopathological mechanism is still unknown [3].

The clinical manifestations depend on the number of lesions and their extension to the soft tissues. The organs affected by the disease, besides the head and the neck, may be the skin, long bones and the digestive system.

The Histiocyte Society proposes a classification based on the age, the number and topography of lesions [4]:

Type 1: unifocal involvement

Type 2: Multi-focal involvement without liver, splenic, pulmonary or spinal cord injury, and aged 2 years and over.

Type 3: Multi-focal involvement with at least one impairment of the following four organs: liver, spleen, lung, marrow, or less than 2 years old.

The involvement of liver, lung, bone marrow makes the prognosis worse.

The diagnosis is based on histopathological findings which show a granuloma made of clusters of vacuole dhistiocytes surrounded by lymphocytes and plasma cells.

Immunohistochemistry shows CD1a positivity [4,5].

We distinguish multiple clinical entities with a common histological substratum, localized eosinophilic granuloma (isolated benign lesion), Letterer-Siwe disease (rapidly progressive multi-organ involvement) and Hand Schuller's disease. Christian (progressive condition with diabetes insipidus, intracranial bone deficiencies and exophthalmos) and transition forms which associates multiples lesions and locations [4].

Skeletal involvement is one of the most common localization in LCH especially in the pelvis, skull, ribs, vertebra and long bones. Jaws are the site of 7,9% of localizations. In this case, the patient present with swelling, local pain, mucosal ulceration and destruction of alveolar bone. The lesion is unique in 70 to 75% of cases. Mandibular localizations are considered common. They represent 20.8% of non-odontogenic tumors. Mandibular involvement is much more common than that of the maxillary.

Local complications may include pathological fracture, ocular compression and dental instability, functional outcomes are usually limited [6].

Because of the destructive potential of LCH, it can be confused with osteomyelitis or malignant lesion.

X-Rays show osteolytic destructive lesions. CT scan may reveal, beside osteolytic lesion, a periosteal reaction. On MRI, a focal lesion with extensive soft tissue and marrowedema is most commonly found as hypointense areas in T1W images and hyperintense area on T2W and STIR images with enhancement. An endosteal rim of low signal intensity can be found in LCH, which may be an early sign of healing [7].

Liver involvement in children with LCH is associated with high mortality rate. They present with hepatomegaly (due to direct infiltration) and liver dysfunction. CT scan of the liver shows periportal hypodensities, hypodense nodules in parenchyma centered by hepatic vessels, periductular fibrosis and micronodular biliary cirrhosis.

MRI can show abnormal periportal signal and biliary stenosis and dilatations [8]. In our case, we found a hypodense collection in addition to periportal hypodensities.

The treatment of LCH still depends on the extent and the severity of disease at diagnosis. The LCH Study Group adopted a stratification system, which separated LCH patients into two major categories [4,9]:

1. 'Single-system' LCH: subdivided further into single and multiple sites.
2. 'Multisystem' LCH: defined as involvement of two or more organs at diagnosis with or without organ dysfunction; This group was further divided into a 'low-risk' and a 'risk' group.

Low-risk patients (20% of patients), have an excellent prognosis and are characterized by the absence of involvement of 'risk' organs such as liver, lungs, spleen or haematopoietic system.

'Risk' patients (80% of patients) have at least one or more risk organs involved and a high mortality rate.

When LCH affects liver, treatment is aggressive due to the irreversible damage of cholestasis. It consists of systemic chemotherapy [8].

The Histiocyte Society considers an initial 6-week course of therapy with vinblastine and prednisone to be effective treatment with minimal toxicity and is therefore suggested for all patients with MS-LCH; regardless of risk organ involvement [4,10].

Conclusion:-

The diagnosis of Langerhans histiocytosis is often delayed due to a multitude of differentials, and to this day, no screening test has been approved. Computed tomography with contrast is considered the tool of choice for analysis of bone and soft tissue of mandibular histiocytosis. Rapid diagnosis can improve prognosis, especially in liver involvement.

Conflict of interest:

The authors have no conflict of interest

Source of support:

None

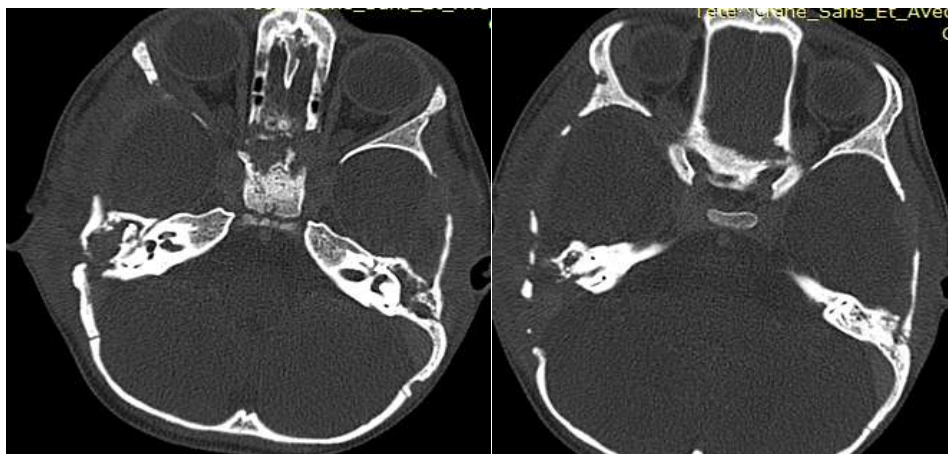


Figure 1:- Computed tomography of the temporal bone showing bilateral mastoid osteolysis destroying the mastoid, extended to the temporal bone .



Figure 2:- Abdominal ultrasonography revealed hepatomegaly with periportalhyperechoic images and anhypoechoic collection.

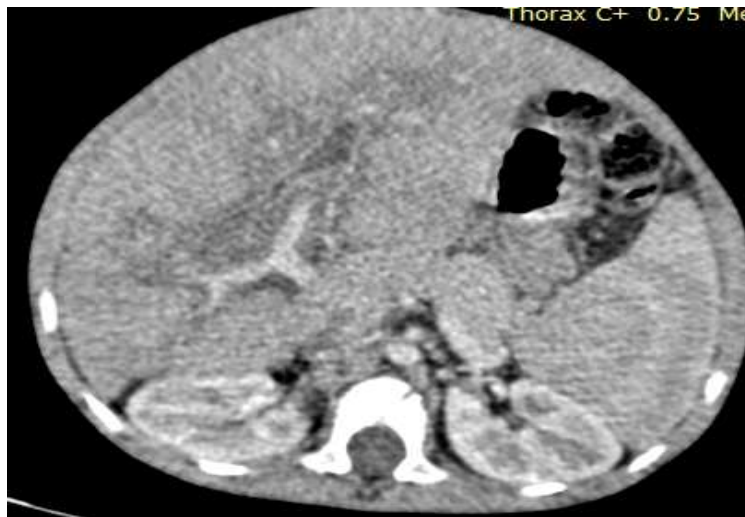


Figure 3:- Abdominal scan which showing diffuse periportalhypodensity with hypodense collection enhanced at the periphery after injection of contrast medium.



Figure 4:Photo showing jaw bilateral swelling.

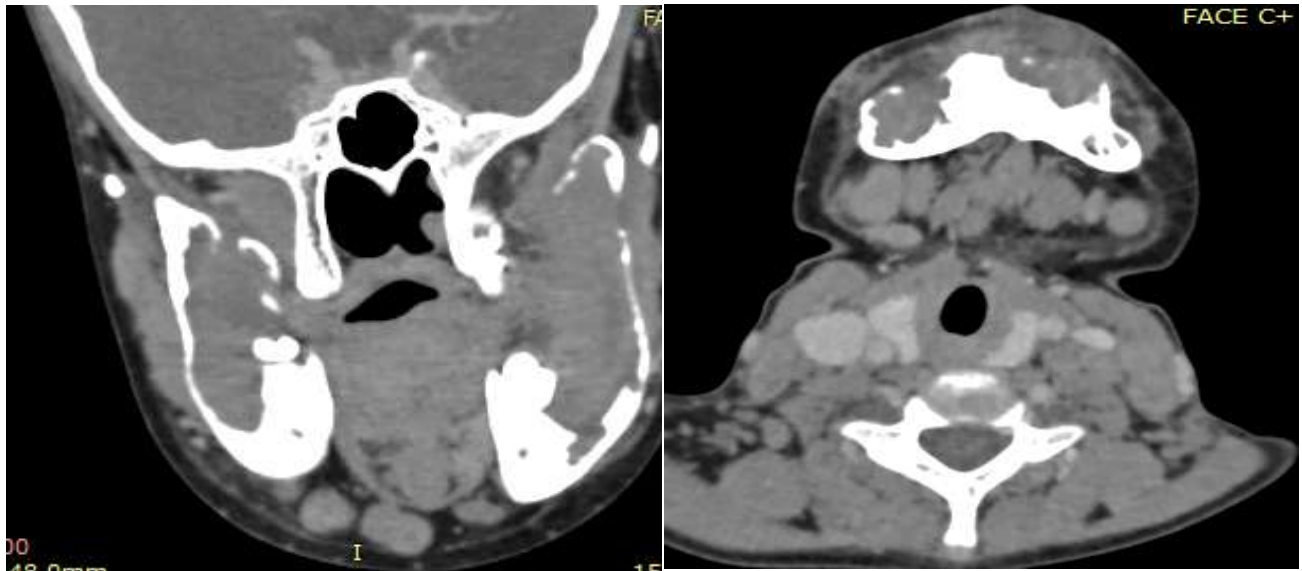


Figure 5:- Cranio facial enhanced CT showing an extensive geographical osteolytic homogeneous mass destroying the mandible.



Figure 6:- Volume rendering CT showing an extensive osteolytic mass destroying the mandible.

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