Case 13382

Eurorad

Multisystemic Langerhans cell hystiocitosis - Head and neck involvement

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Section: Paediatric radiology
Area of Interest: Bones
Procedure: Localisation
Procedure: eLearning
Procedure: Education
Imaging Technique: MR
Imaging Technique: Ultrasound
Special Focus: Hyperplasia / Hypertrophy Neoplasia
Case Type: Clinical Cases
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Patient: 11 months, male

Clinical History:

11 month old baby who presented to hospital with a growing soft-tissue mass in the left temporo-parietal skull associated with cervical lymphadenopathy. Erythematous maculopapular lesions in the retroauricular region, inguinal region and axillary folds were also present. After 6 months of life, he suffered from multiple otitis.
Imaging Findings:

Plain radiograph: Well-defined lytic lesion with a double contour (hole within a hole) indicating button sequestrum (sclerotic centre = residual bone) associated with a growing soft-tissue mass. (Fig. 1).

Sonography: Left temporal soft-tissue lesion invading and destroying the inner and outer table of the skull, leaving multiple hyperechoic coarse calcifications inside, which show a twinkling artefact on colour Doppler US, compatible with residual skull bone (Fig. 2).

MRI: Osteolytic lesion associated with a soft-tissue mass of 3.5 cm approximately. This lesion showed heterogeneous and peripheral enhancement as well as restricted diffusion (Fig. 3).

PET-CT: Performed to identify other LCH lesions. PET-CT is the most sensitive imaging method for detecting LCH lesions and for evaluating response to therapy. In our case, an increased F-18 FDG uptake (SUVmax: 5.0) focus in the left temporal lesion was observed. This finding is in relation to an active and proliferating lesion. Bilateral laterocervical hypermetabolic lymphadenopathy was also observed (SUVmax: 8.5). (Fig. 3)
Discussion:

BACKGROUND

Langerhans cell histiocytosis (LCH) is a rare multi-system disease with a wide clinical spectrum, but the most frequently affected organs are skeleton (80% of cases), skin (33%), and pituitary (25%). Extraosseus disease

includes pulmonary, thymic, hepatobiliary, splenic, gastrointestinal, neurologic, mucocutaneous, head and neck soft-tissue masses and salivary involvement. The disease is more common in the paediatric population. [1]

CLINICAL PERSPECTIVE

LCH is divided into 3 groups (based on the number of lesions, systems involved)
1- Unifocal (localized) form: (70% of cases) Limited to a single or a few bones and may involve the lung.
2- Multifocal unisystem (20% of cases) Multiple bones, reticuloendothelial system, pituitary/hypothalamus.
3- Multifocal, multisystem: (10% of cases) The worst prognosis. Multisystem LCH occurs in two or more organs or body systems or may be spread throughout the body.

LCH may affect low-risk or high-risk organs: Low-risk organs include the skin, bone, lungs, lymph nodes, gastrointestinal tract, pituitary gland. High-risk organs include the liver, spleen, and bone marrow. CNS lesions have their own classification in low and high-risk lesions. [1]

IMAGING PERSPECTIVE

Head and neck involvement in Langerhans cell histiocytosis is common and may include bone and soft-tissue lesions, cervical lymphadenopathy and skin rash.
Craniofacial osseous destruction has been described in association with adjacent soft-tissue infiltration.
The ear is a common site of involvement and its involvement most often manifests as chronic otorrhoea, mastoiditis or otitis (as our patient). Most patients who present ear involvement have multisystem disease. [2]

CT is performed to evaluate the extent of osseous erosion or destruction. Temporal bone destruction (mastoid, external auditory meatus, middle or internal ear) is often associated with an adjacent soft-tissue mass. [2, 3, 4]

MRI shows soft-tissue masses with hyperintense signal on T2-weighted sequences and isointense to hypointense signal on T1-weighted sequences with marked enhancement.
Involvement of the skull base may infiltrate the petrous portion of the temporal bone. [5]

OUTCOME

Biopsy was performed to confirm the diagnosis. Prognosis can be variable (unifocal disease: >95% survival; two organ involvement: 75%; Langerhans cell sarcoma: 50% survival). In head and neck involvement, outcome after chemotherapy is good although recurrences are common, especially in patients with multisystem disease.
Treatment is based on vinblastine, prednisolone and metrotexate. [1, 2]

TAKE HOME MESSAGE

- LCH is a rare multi-system disease

- LCH is divided into 3 groups: Unifocal/multifocal, unisystem/multifocal, multisystem form.

- Head and neck involvement in LCH is common (bone and soft-tissue lesions, cervical lymphadenopathies and skin rash)

- Ear involvement is a common and manifests as chronic otorrhoea, mastoiditis or otitis.
Differential Diagnosis List: Multisystemic Langerhans cell histiocytosis - Head and neck involvement., Lymphoma, Rosai-Dorfman (sinus histiocytosis+ lymphadenopathy + soft-tissue lesions of the head and neck + it

does not involve the skin), Mastoiditis, Sarcoidosis, Tuberculosis

Final Diagnosis: Multisystemic Langerhans cell histiocytosis - Head and neck involvement.

References:

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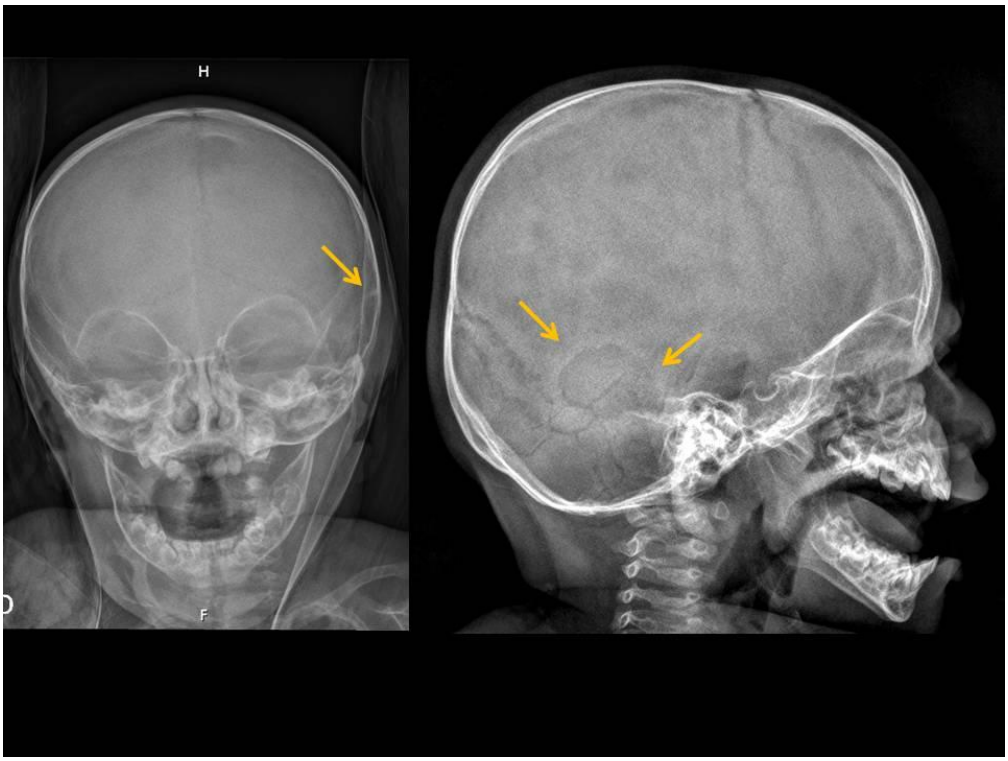
Figure 1

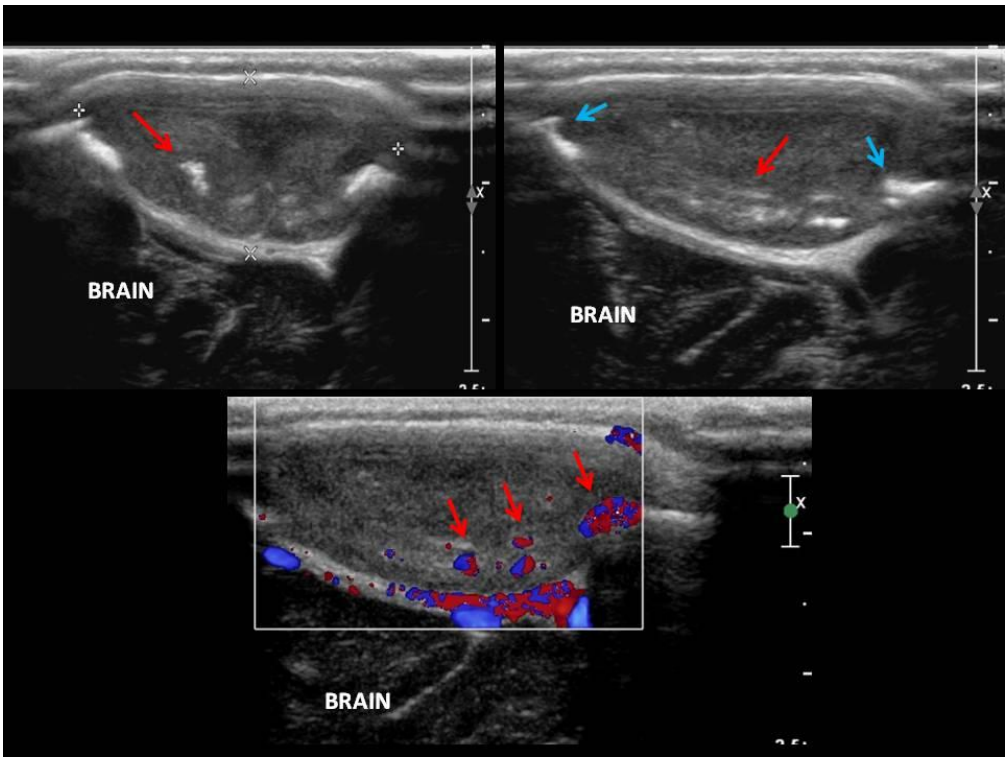
a



Description: Well-defined lytic lesion with a double contour (hole within a hole = button sequestrum) and geographic margins, associated to a growing soft-tissue mass (yellow arrows). **Origin:** Prat-Matifoll J.A, Radiology Department, Vall Hebron Hospital

Figure 2

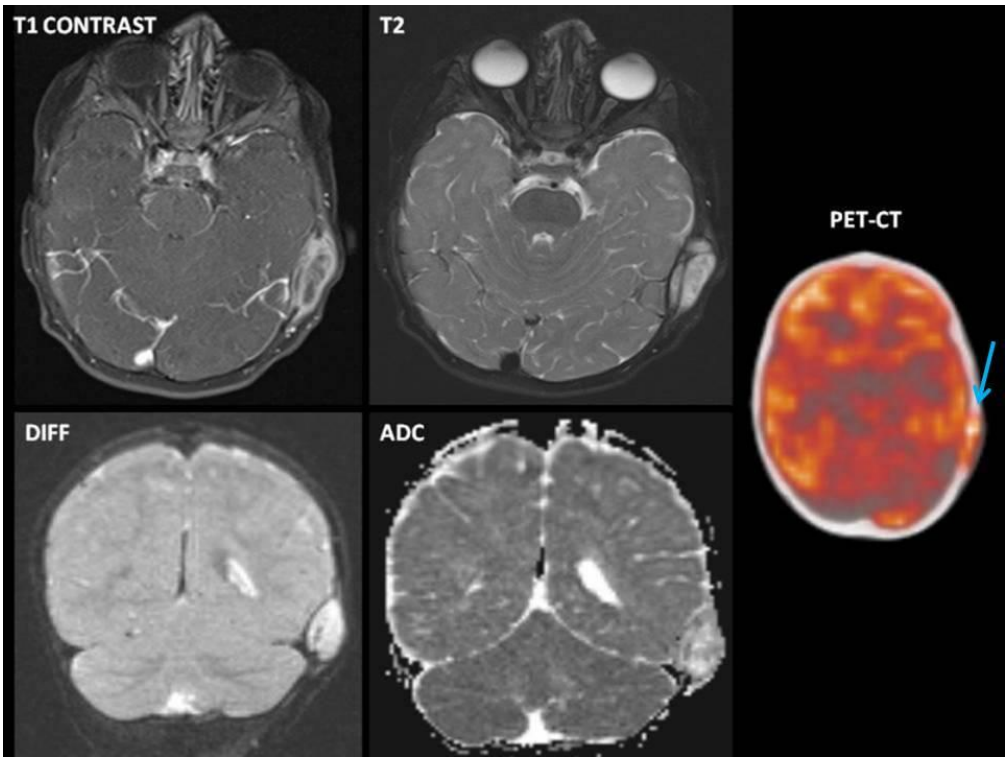
a



Description: Soft-tissue lesion destroying the skull (blue arrows), leaving multiple hyperechoic calcifications inside, which show a twinkling artifact (red arrows). Mild Doppler signal within the lesion was observed. **Origin:** Prat-Matifoll J.A, Radiology Department, Vall Hebron Hospital

Figure 3

a



Description: Osteolytic soft-tissue mass showing an heterogeneous and periferical enhancement as well as a restricted diffusion.

PET-CT showed an increased F-18 FDG uptake (SUVmax: 5.0) (blue arrow) **Origin:** Prat-Matifoll J.A, Radiology Department, Vall Hebron Hospital