Case 16438

Eurorad

Solitary bone lesion in a 4-year-old girl

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Section: Head & neck imaging
Area of Interest: Bones Paediatric
Imaging Technique: CT
Imaging Technique: MR
Case Type: Clinical Cases
Authors: Elena Zabía Galíndez1, Alicia Duque2
Patient: 4 years, female

Clinical History:

A 4-year-old-girl presented with progressive unilateral jaw pain and swelling. Blood examinations showed increased inflammatory markers.

Imaging Findings:

CT imaging showed a well-defined intramedullary lytic lesion in the right mandible with cortical thinning, focal cortical defects and without reactive sclerosis.

MRI confirmed the lytic bone lesion, hyperintense to muscles on T2-weighted images, isointense to muscles on T1-weighted images and with diffuse contrast-enhancement, and better depicted the infiltration of the lateral pterygoid muscle and the masseter.

Discussion:

Langerhans-cell histiocytosis (LCH) is a rare multisystem disease of unknown cause characterised by clonal proliferation of antigen-presenting mononuclear cells of dendritic origin [1] (Langerhans cells: CD1a, CD207 and S100 positive) [2] and presents a heterogeneous clinical spectrum including:

- Letterer-Siwe disease (up to 10%): fatal acute disseminated leukaemia-like form, affecting infants and newborns and characterised by bone marrow infiltration, hepatosplenomegaly, lymphadenopathy and cutaneous and bone lesions (skull, mandible and long bones). [3]

- Hand-Schuller-Christian disease (15-40%): chronic disseminated form, affecting children over 3 years and characterised by lytic bone lesions (skull, scapula, ribs, pelvis), classically associated with diabetes insipidus and exophtalmus. [3]

- Eosinophilic granuloma (60-80%): chronic localised form affecting children and young adults characterised by single or multiple osseous lesions, most frequently affecting skull, mandible and long bones. Isolated lesions mostly occur in the thoracic spine. [3, 4]

EG (< 1% of all bone tumours) [1] shows Langerhans cells mixed with inflammatory and giant cells without nuclear

atypia and atypical mitosis, which differentiates it from malignant conditions (Ewing sarcoma, bone lymphoma and metastatic neuroblastoma), which may look similar due to the round cells seen in all those cases. [6]

The lesion may be asymptomatic. Clinical symptoms include pain, swelling and tenderness to palpation and depend on the location of the lesion, particularly in the spine.
EG is known as “the great mimicker” as it appears similar to many lesions in different imaging techniques, typically as a well-defined lytic bone lesion without reactive sclerosis frequently surrounded by a hypervascularised soft-tissue mass.
In the skull, a solitary or multiple punched out lytic lesions can be seen. In the mandible the “floating tooth” sign is the result of alveolar bone destruction around its root. In the spine it can progress to vertebral collapse (vertebra plana) with peridural/paraspinal spread.
MRI signal characteristics include hyperintensity on STIR and T2-weighted images, hypo-isointensity on T1-weighted images and diffuse contrast enhancement. [6]

Tumour tissue has to be tested for BRAFV600E [2]

Observation can be considered for asymptomatic lesions. Treatment of single bone lesions includes curettage with or without radiotherapy or intralesional steroid injection. Curettage and bone grafting are indicated to prevent fractures, particularly in lesions near articular surfaces. Corticosteroid injections can be used for isolated lesions not amenable to treatment and for spinal lesions that compromise stability or neurologic status. [2, 7]
Chemotherapy is indicated in cases of skull-based lesions, single system multifocal involvement and multisystem involvement and allogenic haematopoietic-cell transplantation in cases of refractory or relapsed LCH. [2]

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Eosinophilic granuloma, Odontogenic cyst, Osteomyelitis, Primary bone tumours, Lymphoma or leukaemia, Metastases

Final Diagnosis: Eosinophilic granuloma

References:

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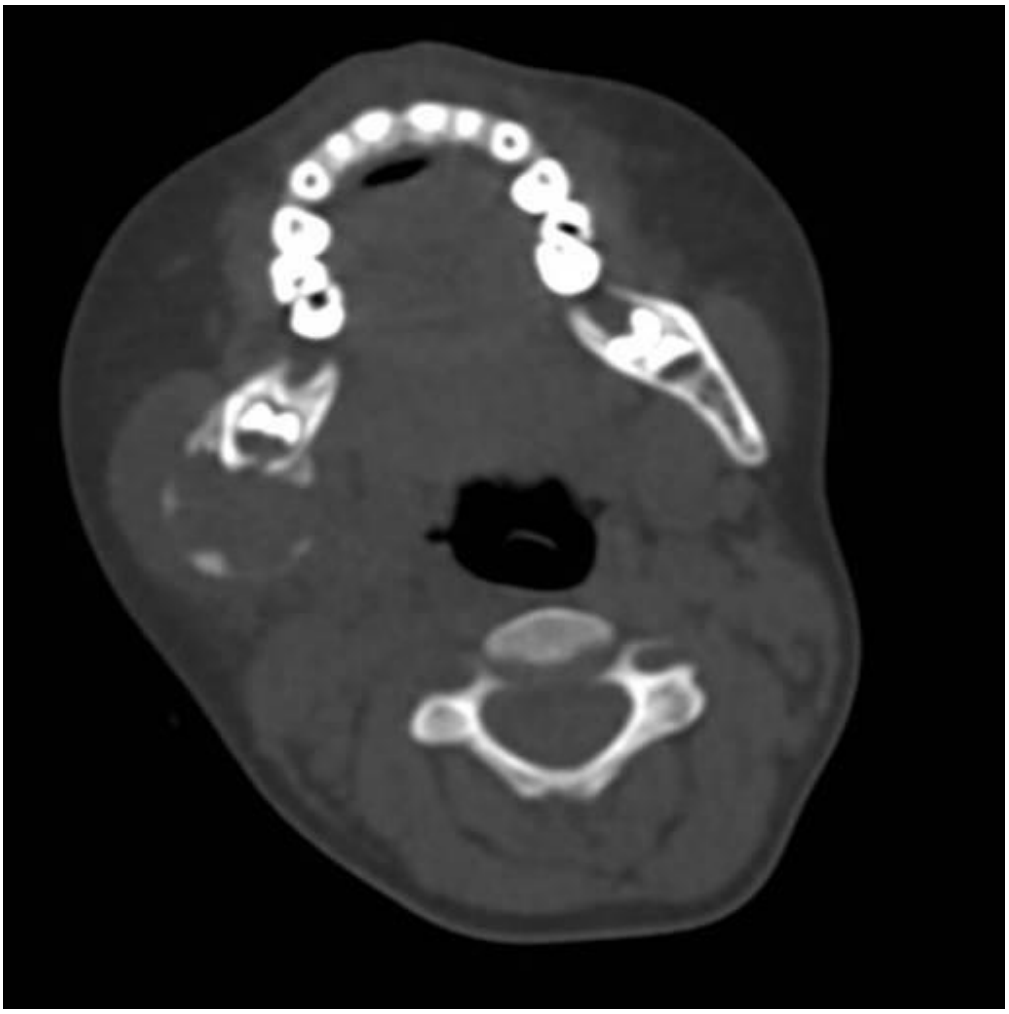
Haupt R, Minkov M, Astigarraga I, et al (2013) Langerhans cell histiocytosis (LCH): guidelines for diagnosis, clinical work-up, and treatment for patients till the age of 18 years. Pediatr Blood Cancer 60 (2):175–184 (PMID:23109216)

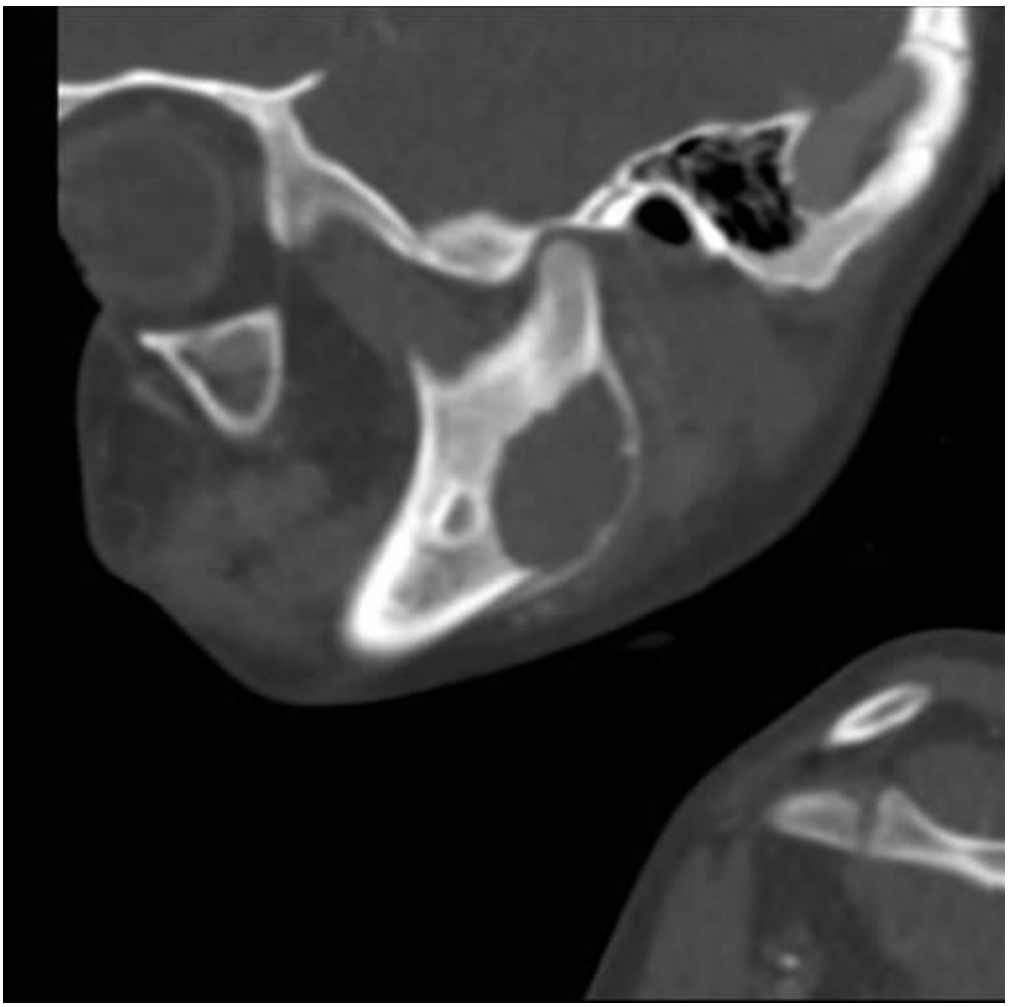
Figure 1

a



Description: Axial (a) and sagittal (b) CT images on bone window setting and axial CT image on soft-tissue window setting (c) show a well-defined intramedullary lytic lesion in the right mandible, isodense to the skeletal muscles, with cortical thinning and focal cortical defects and without reactive sclerosis.
Origin: Department of Radiology. Hospital HM Montepríncipe, Madrid, 2019

b



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c

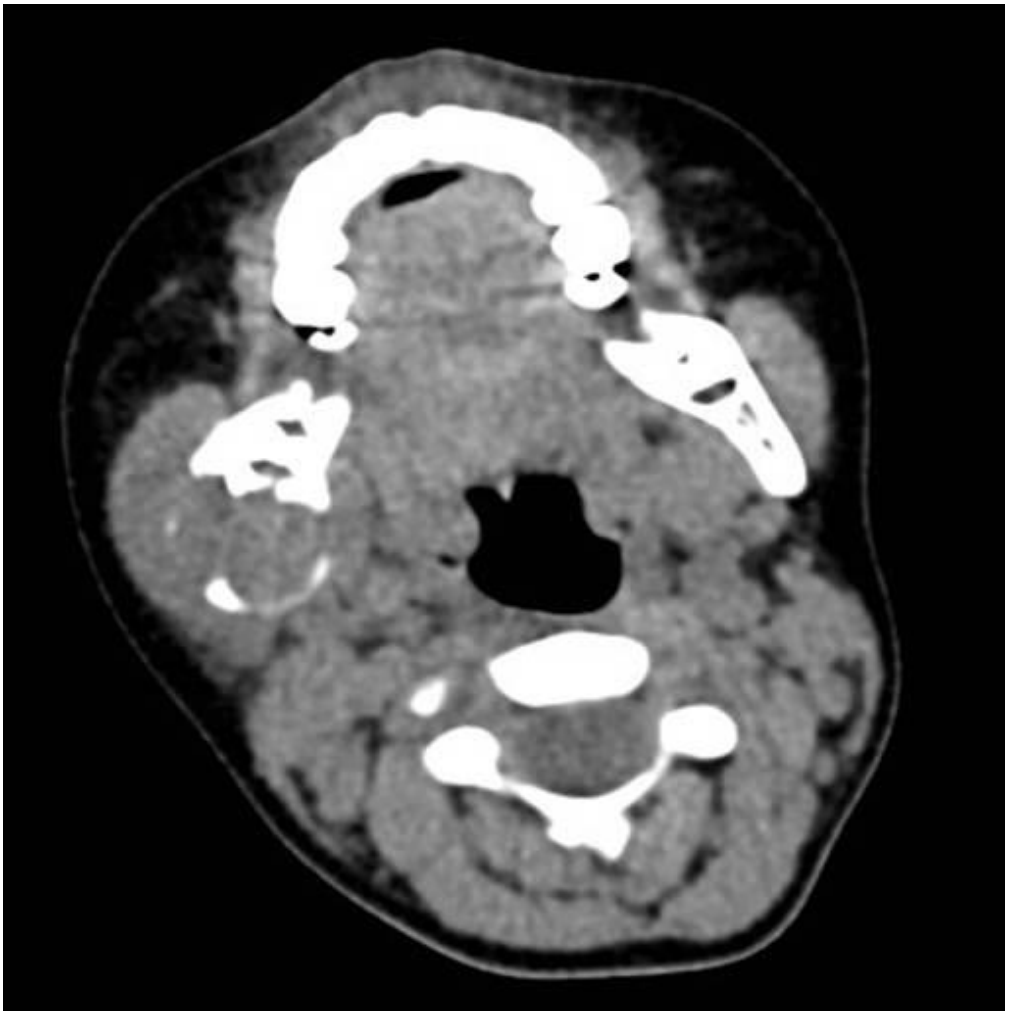

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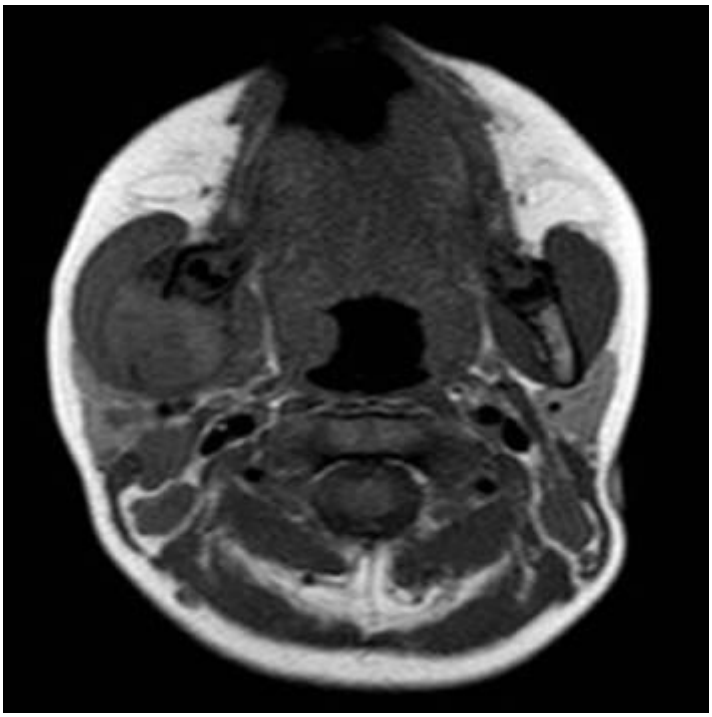
Figure 2

a



Description: MRI images show the lytic mass, isointense to the muscles on T1-weighted MRI images (a) and hyperintense to the muscles on T2-weighted images (b) **Origin:** Department of Radiology. Hospital HM Montepríncipe, Madrid, 2019

b

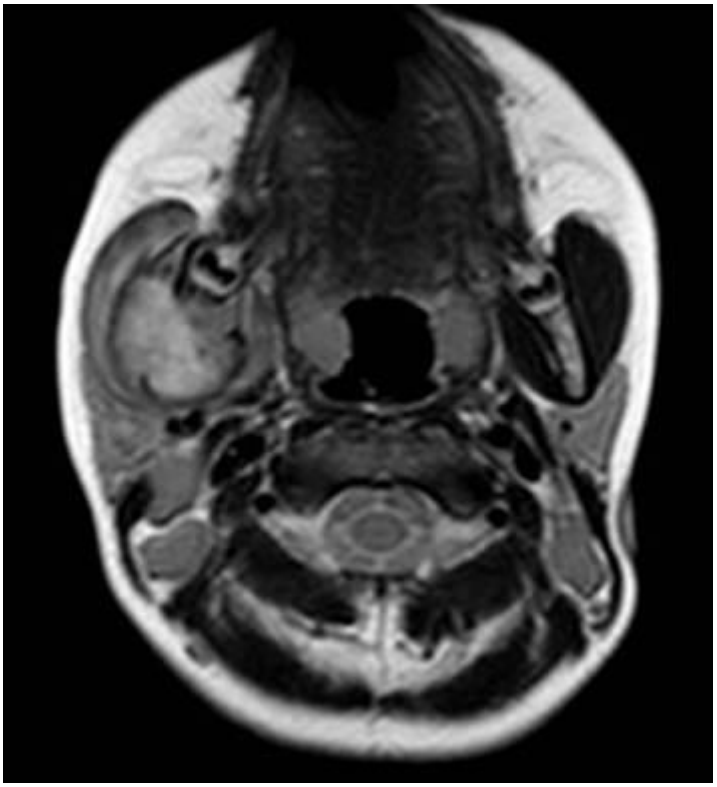


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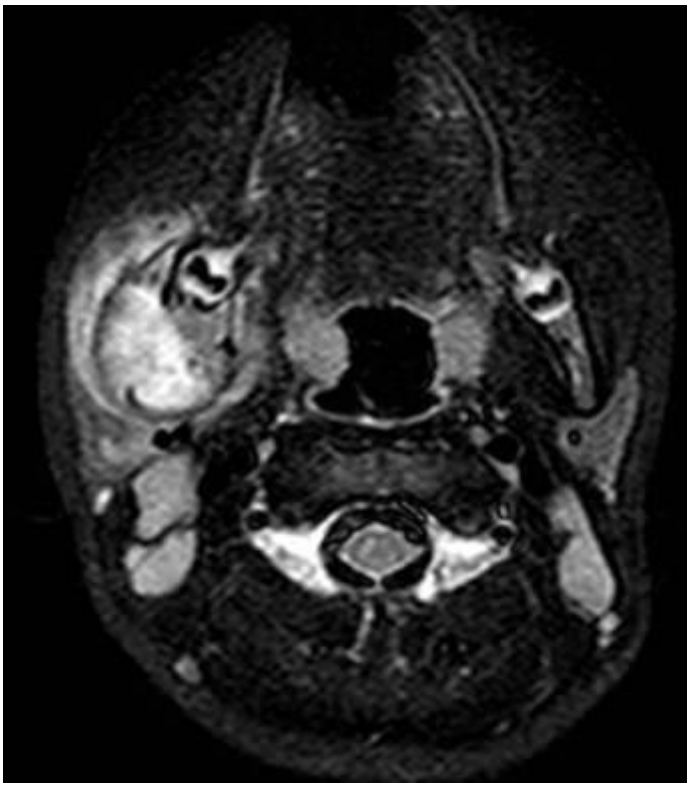
Figure 3

a



Description: Axial (a) and coronal (b) fat-suppressed T2-weighted MRI images better depict the extension of the lesion and the involvement of the lateral pterygoid muscle and the masseter. **Origin:** Department of Radiology. Hospital HM Montepríncipe, Madrid, 2019

b

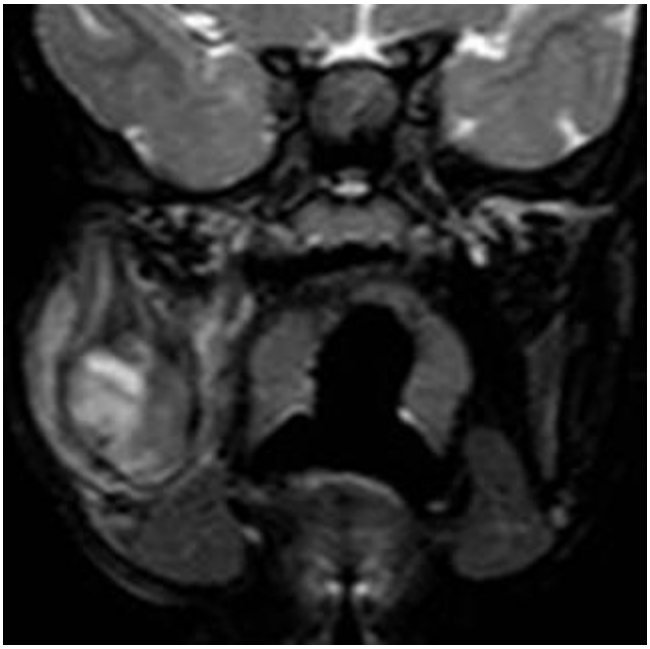


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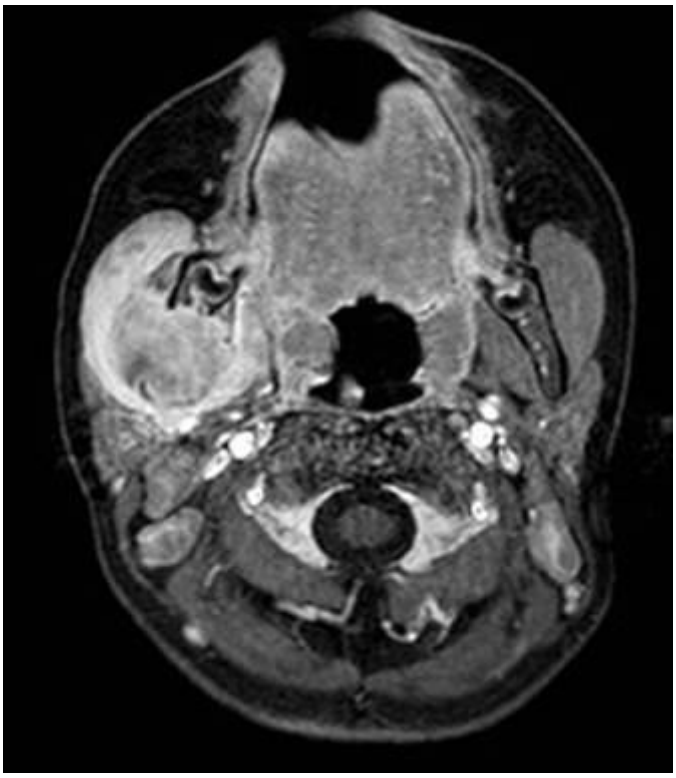
Figure 4

a



Description: Post-gadolinium contrast enhanced MRI imaging shows the diffuse contrast enhancement of the mass and the infiltration of the lateral pterygoid muscle and the masseter. **Origin:** Department of Radiology. Hospital HM Montepríncipe, Madrid, 2019