

## Langerhans cell histiocytosis, classic findings in a paediatric patient

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**Section:** Paediatric radiology

**Area of Interest:** Head and neck Haematologic Oncology  
Paediatric Bones

**Procedure:** Diagnostic procedure

**Imaging Technique:** CT

**Imaging Technique:** Conventional radiography

**Imaging Technique:** Catheter arteriography

**Imaging Technique:** MR

**Special Focus:** Haematologic diseases Case Type:  
Clinical Cases

**Authors:** Bosca-Ramon A, Dualde-Beltran D, Nersesyan  
N, Flores-Casaperalta S, Delgado-Moraleda JJ,

**Patient:** 13 months, male

### Clinical History:

13-month-old nursing infant, whose parents noticed two soft, swollen right frontal and left temporal lesions, presented with fever, pallor, and bilateral cervical lymphadenopathy. Blood tests showed haemoglobin of 5.8 g/dL.

### Imaging Findings:

Ultrasonography (US) showed bilateral cervical lymphadenopathy.

Skull radiographs demonstrated a right frontal lytic lesion with beveled edges, and a right occipital lytic lesion.

Computed tomography (CT) demonstrated the aforementioned lytic lesions in the skull. Right mandibular and left temporal lytic lesions were also seen. There was a uniform collapse with preservation of the adjacent intervertebral disk spaces of the T4 vertebral body (vertebra plana). T9 and T10 also demonstrated partial collapse. Lytic lesions located on 4th and 9th left ribs, clinoid, clivus, odontoid, and T1 spinous process were also seen.

Magnetic resonance (MR) showed the lytic lesions in the skull. The posterior pituitary bright spot was preserved, and a thin pituitary stalk was observed, meaning that the central nervous system (CNS) was not affected.

### Discussion:

Langerhans cell histiocytosis (LCH) is the most common dendritic cell disorder. The abnormal dendritic cells found in this disease imitate the Langerhans cells found in the normal skin and mucosa. The proliferation and accumulation of abnormal LCH cells in various organs results in the clinical disease. [4] Older names that attempt to group the clinical manifestations such as eosinophilic granuloma, Hand-Schüller-Christian disease, histiocytosis X, and Letterer-Siwe disease should be disregarded. The usual age of presentation ranges between 5 and 15 years of age, but the disease may affect any age group. [1]

LCH may affect any organ. Clinical manifestation varies from patient to patient, and ranges from self-limiting to fatal

disease. LCH is divided into unifocal, multifocal unisystem, and multifocal multisystem forms. [1]

Imaging studies have great importance in determining the extension of the disease, especially the involvement of risk organs that indicate a worse prognosis (liver, spleen, and lung). Haematopoietic system involvement (determined by the presence of 2 of the following: anaemia, leukocytopenia, or thrombocytopenia) is also considered a risk organ. [2]

Lytic skull lesions with beveled edges, and vertebra plana are the most specific imaging findings. CT is the technique of choice for evaluating bone involvement. MR plays a key role in determining the involvement of the pituitary gland and CNS, and US is the technique of choice in determining the presence of hepatomegaly, splenomegaly and lymphadenopathy.

The diagnosis is clinicopathological, based on classical clinical findings and histological/immunophenotypic examination of lesional tissue. Curettage of a bone lesion is generally sufficient for pathologic diagnosis. [3]

Treatment varies according to the extent of the disease, and is given to improve survival but also to prevent late sequelae. Although spontaneous resolution can occur, the treatment of choice is with relatively non-toxic chemotherapeutic agents. [2]

In our case the diagnosis was made with cervical lymph node biopsy.

LCH has a varying clinical manifestation, a varying age of presentation and may affect any organ [5]. Considering its relative rarity, LCH remains a disease in which the diagnosis is often delayed or missed, whilst the prognosis may worsen. [2] Radiologists must be familiar with this condition and its imaging key findings.

**Differential Diagnosis List:** Multifocal multisystem Langerhans cell histiocytosis., Leukaemia, Lymphoma, Metastases, Osteomyelitis, Dermoid cyst

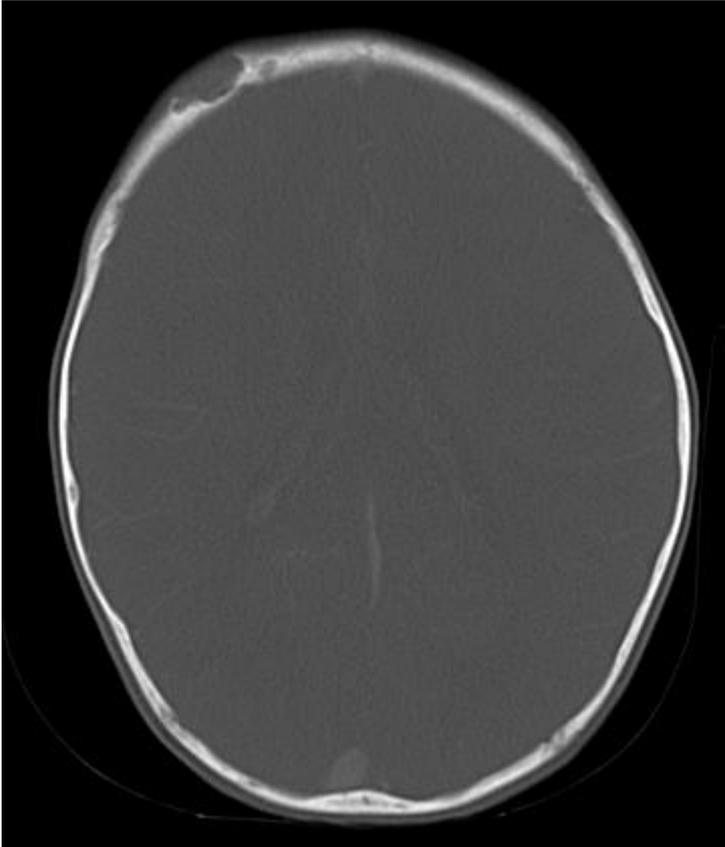
**Final Diagnosis:** Multifocal multisystem Langerhans cell histiocytosis.

#### References:

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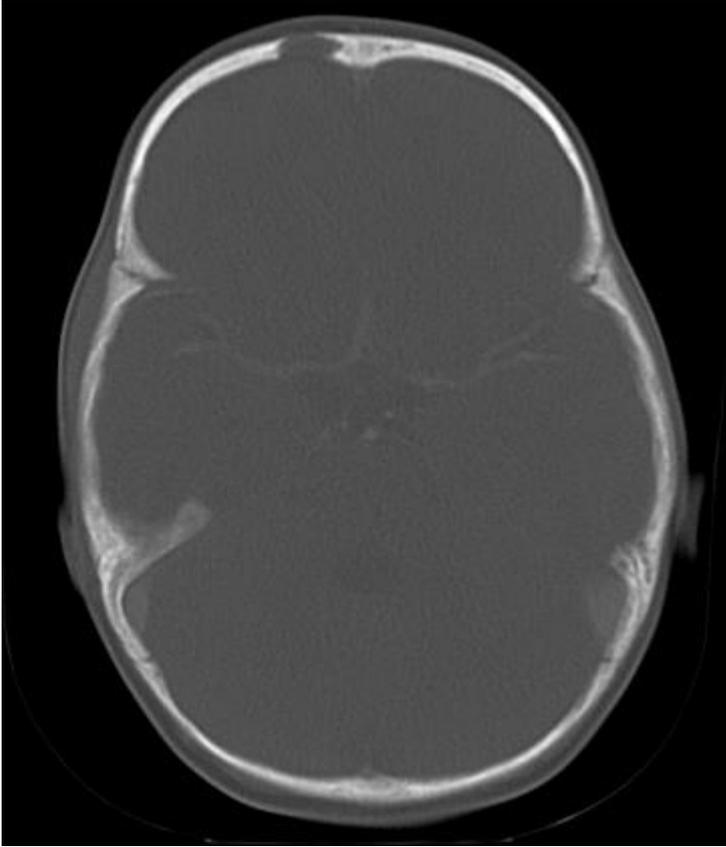
## Figure 1

a



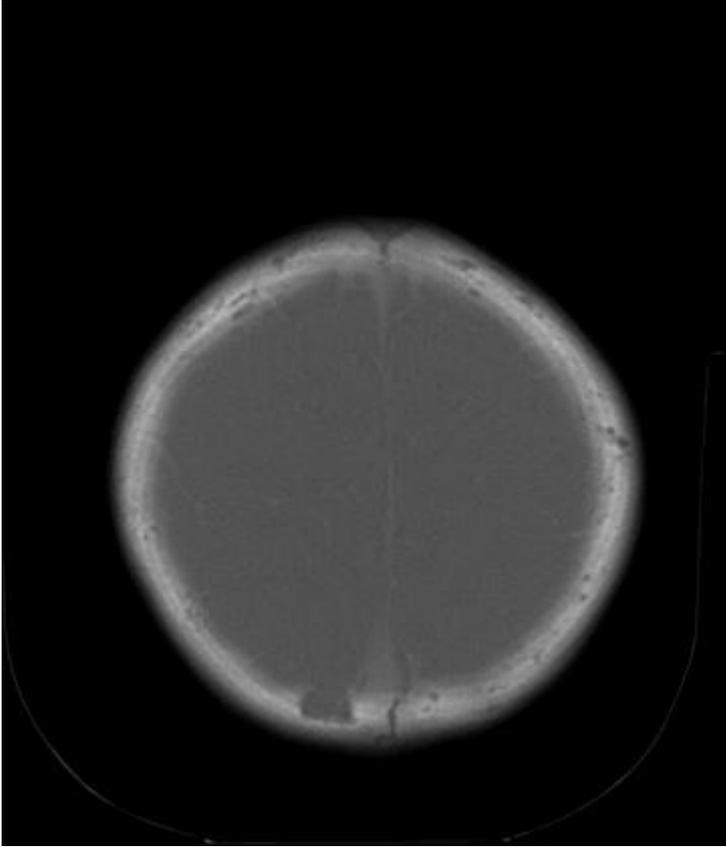
**Description:** Axial CT shows right frontal lytic lesion. Note the asymmetric destruction of the inner and outer cortices, which results in a characteristic beveled edge. **Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

**b**



**Description:** Axial CT shows another right frontal lytic lesion. **Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

**c**



**Description:** Axial CT shows right occipital lesion. **Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

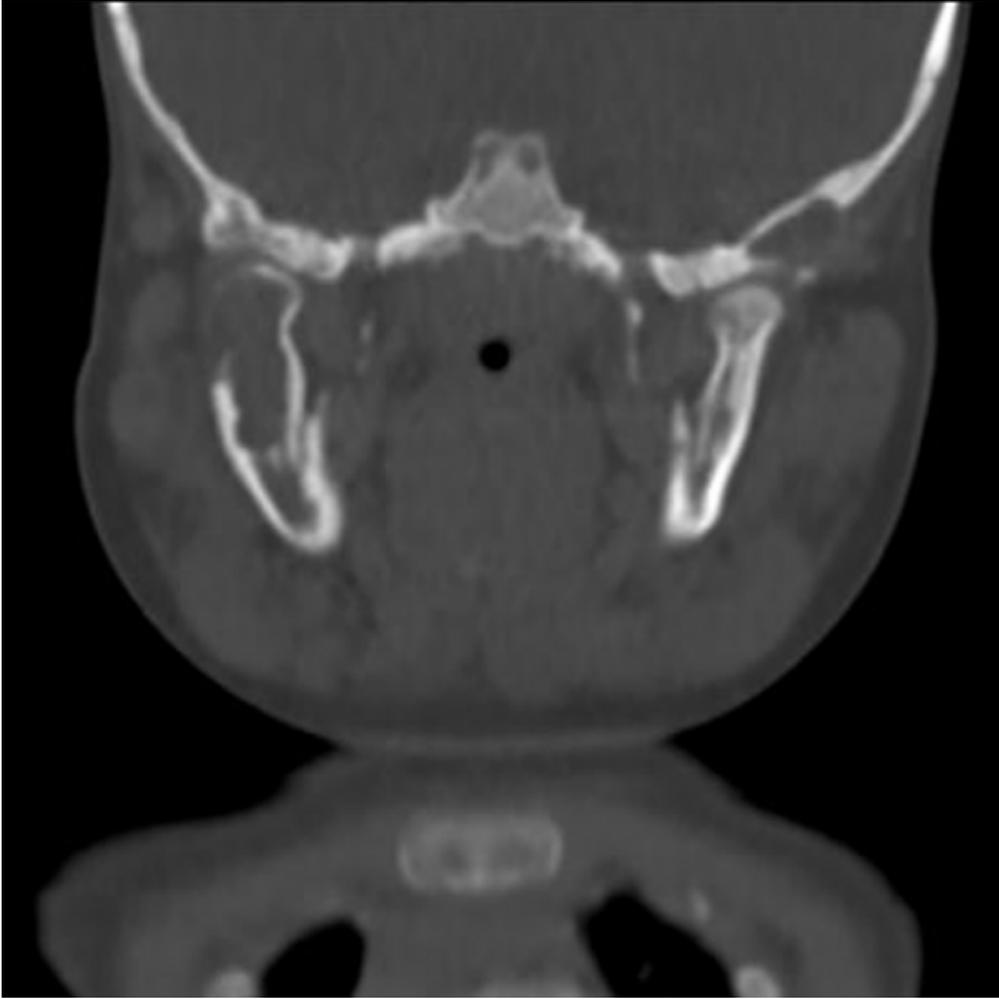
## Figure 2

a



**Description:** Axial CT shows right mandibular lytic lesion. **Origin:** Antoni Bosca, Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

**b**



**Description:** Coronal CT shows right mandibular and left temporal lytic lesions. **Origin:** Antoni Bosca, Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

c



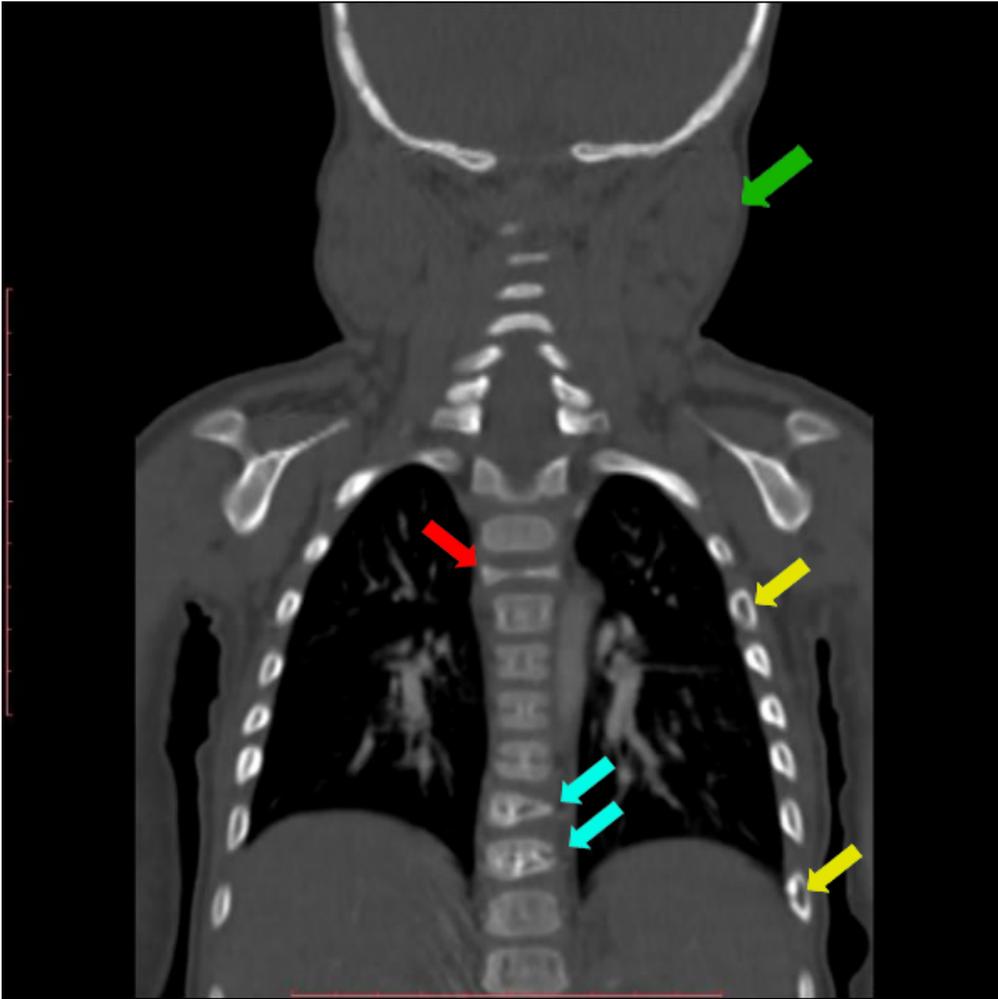
**Description:** Coronal CT shows right mandibular and left temporal lytic lesions. **Origin:** Antoni Bosca, Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

d



**Description:** T4 uniform collapse (vertebra plana) is seen. Note the preservation of the adjacent intervertebral disk spaces. T9 and T10 partial collapse, 4th and 9th left rib lytic lesions and bilateral cervical lymphadenopathy are also seen. **Origin:** Antoni Bosca, Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

e



**Description:** T4 uniform collapse (vertebra plana) is seen. Note the preservation of the adjacent intervertebral disk spaces. T9 and T10 partial collapse, 4th and 9th left rib lytic lesions and bilateral cervical lymphadenopathy are also seen. **Origin:** Antoni Bosca, Department of Radiology, Hospital Clínic Universitario de Valencia, Valencia, Spain.

f



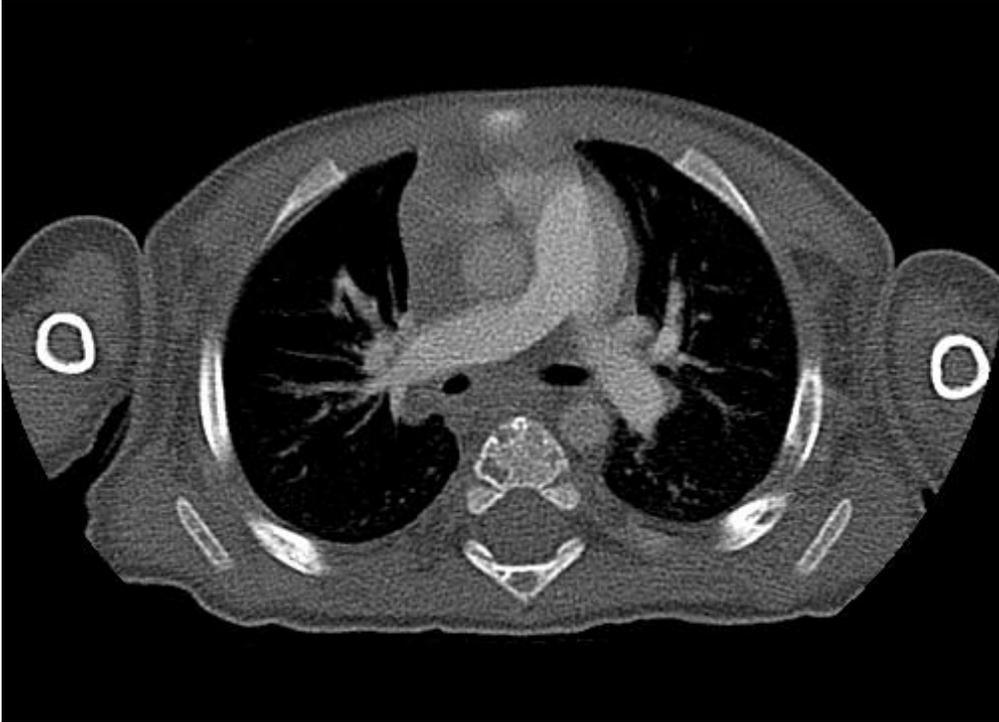
**Description:** Sagittal CT shows vertebra plana. Lytic lesions on clinoid, clivus, odontoid, T9 and T10 vertebral bodies, and T1 spinous process are also present. **Origin:** Antoni Bosca, Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

g



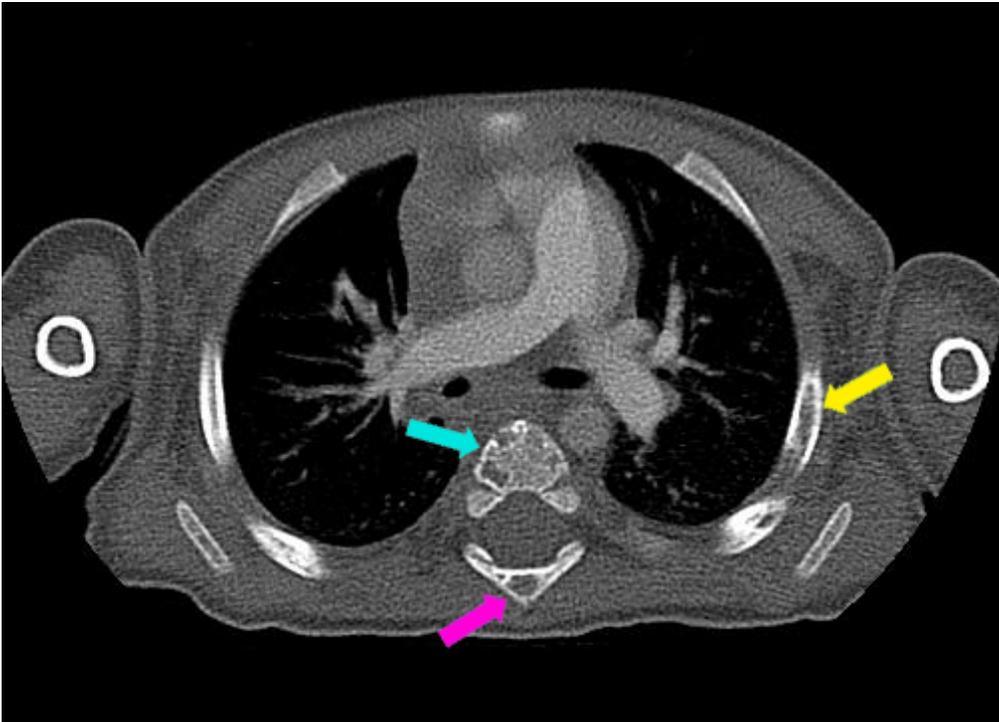
**Description:** Sagittal CT shows vertebra plana. Lytic lesions on clinoid, clivus, odontoid, T9 and T10 vertebral bodies, and T1 spinous process are also present. **Origin:** Antoni Bosca, Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

h



**Description:** Axial CT image shows irregular lytic lesions in a midthoracic vertebral body, spinous process, and left ribs. **Origin:** Antoni Bosca, Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

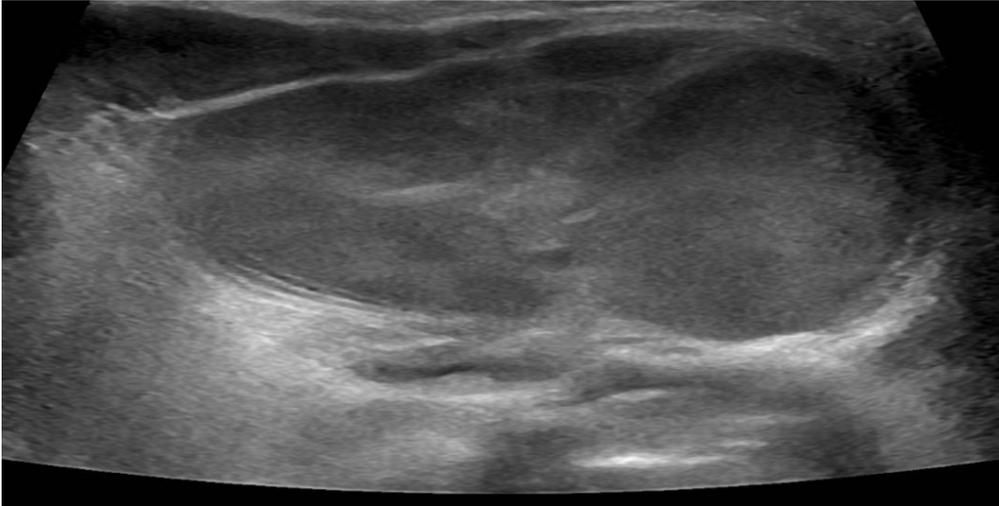
i



**Description:** Axial CT image shows irregular lytic lesions in a midthoracic vertebral body, spinous process, and left ribs. **Origin:** Antoni Bosca, Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

## Figure 3

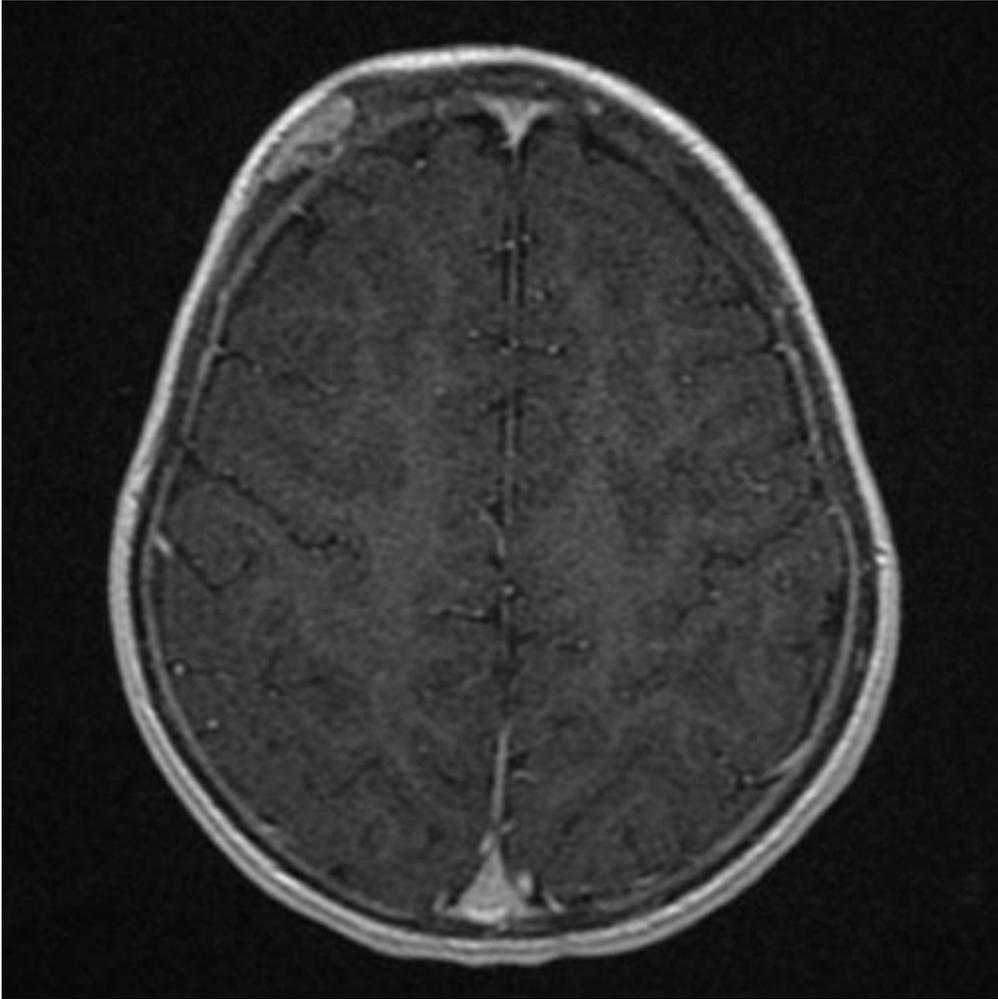
a



**Description:** Ultrasound showed bilateral cervical lymphadenopathy. The largest lymph node was on the right, measuring approx. 8 x 2 cm **Origin:** Antoni Boscà. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

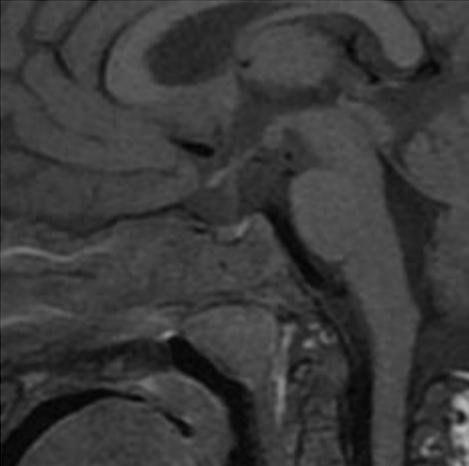
## Figure 4

a



**Description:** Axial 3D FSPGR T1-weighted MR image shows skull lesion, with typical asymmetric destruction of the inner and outer cortices. **Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

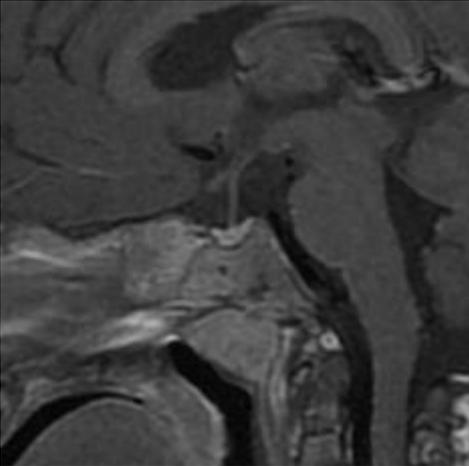
**b**



**Description:** Sagittal FSE T1 weighted MR image shows preserved posterior pituitary bright spot.

**Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

**c**



**Description:** Sagittal FSE contrast-enhanced T1-weighted MR image shows a normal thin pituitary stalk. **Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

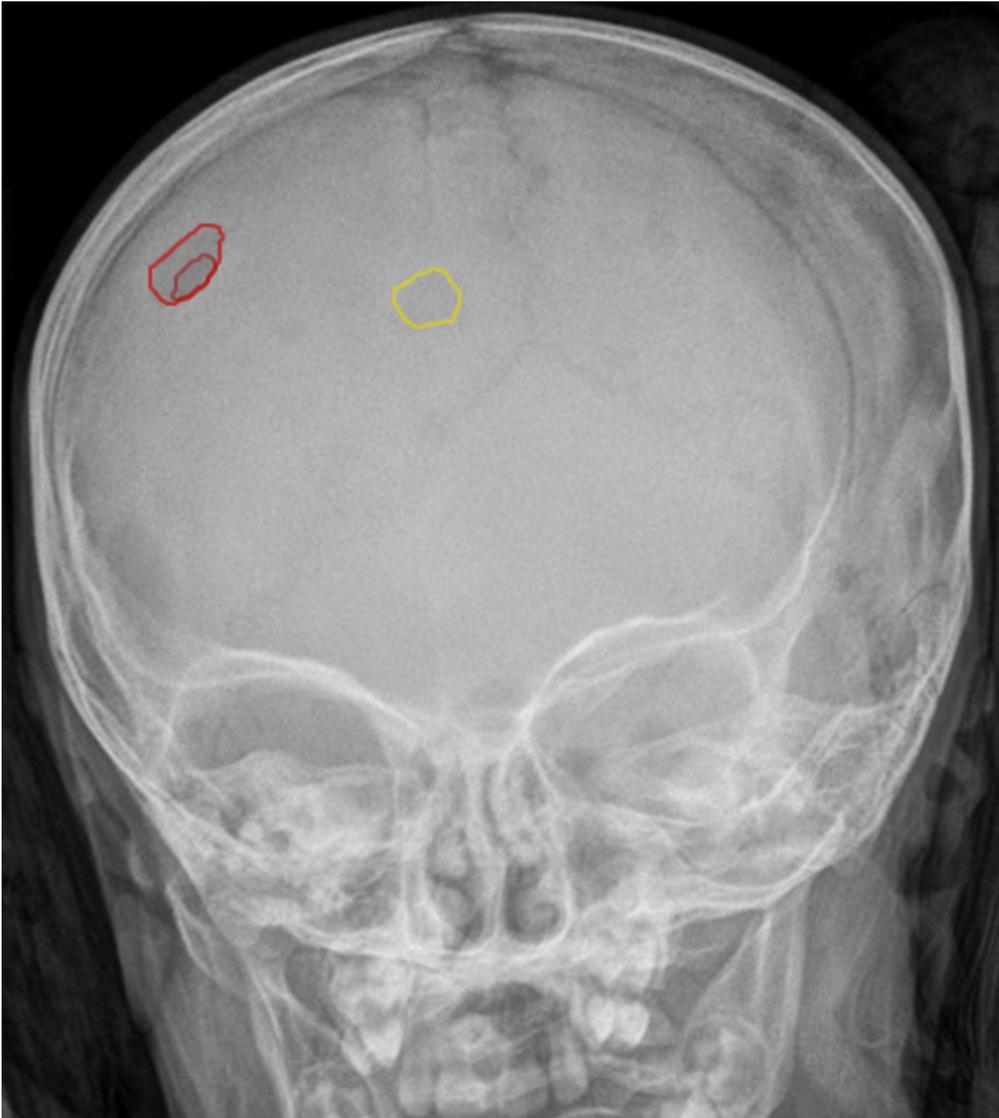
## Figure 5

a



**Description:** Right frontal lytic lesion with beveled edges (hole within a hole sign). A right occipital lytic lesion is also seen. **Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

b



**Description:** Annotated image. Right frontal lytic lesion with beveled edges (hole within a hole sign). A right occipital lytic lesion is also seen. **Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

c



**Description:** Right frontal and occipital lytic lesions. **Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

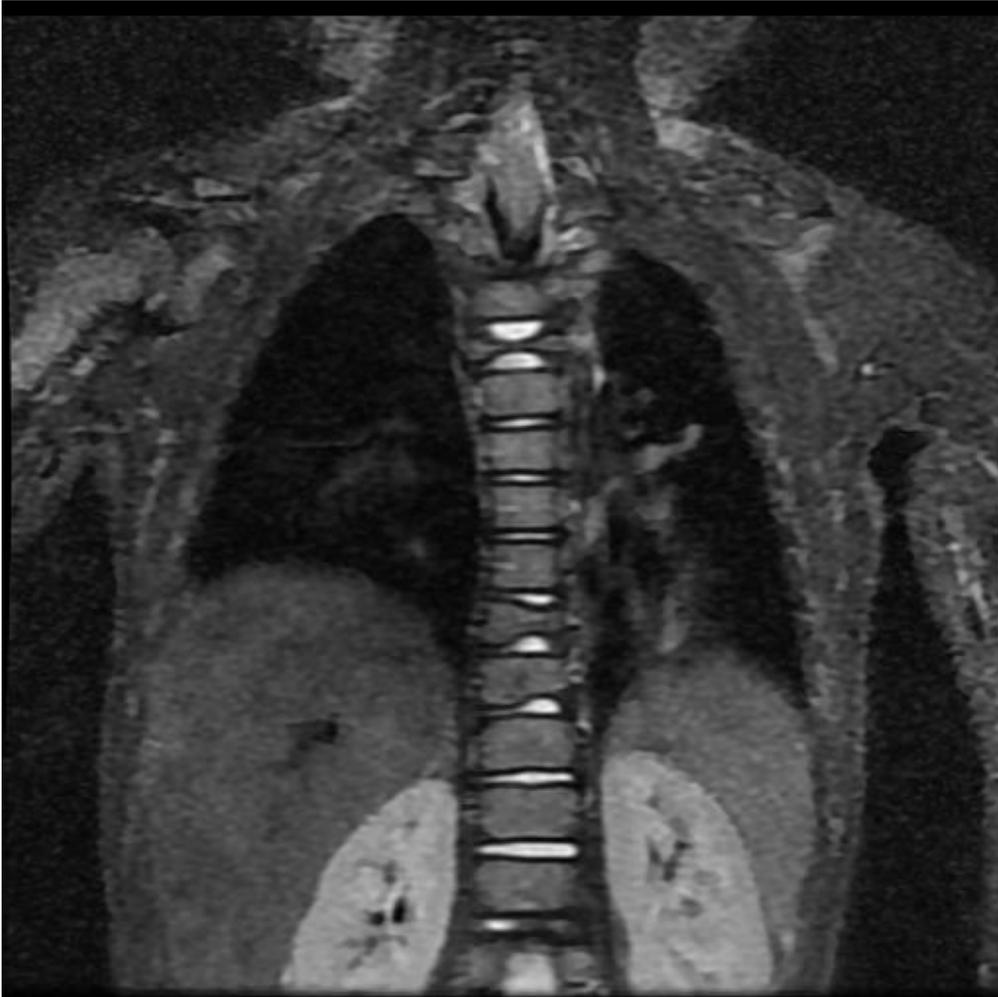
d



**Description:** Annotated image. Right frontal and occipital lytic lesions. **Origin:** Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

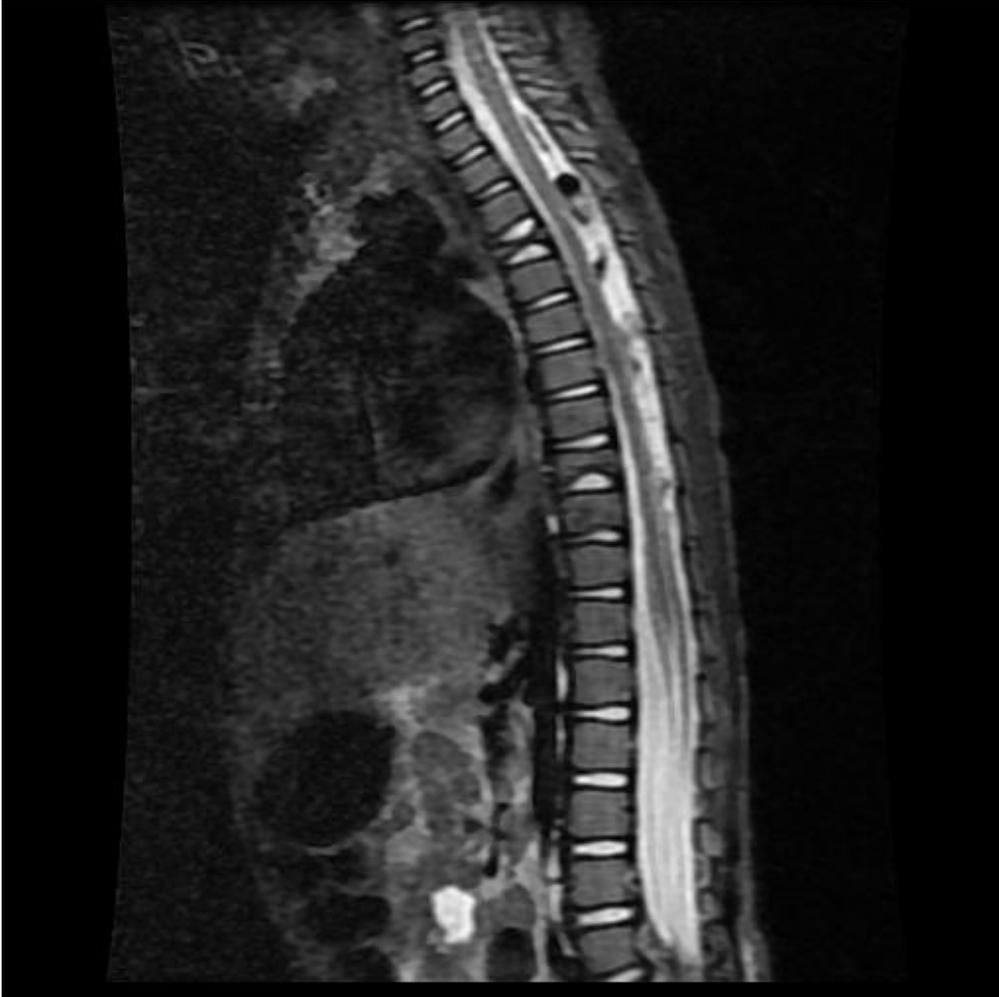
## Figure 6

a



**Description:** Coronal STIR MR shows T4 uniform collapse (vertebra plana). T9 and T10 partial collapse. **Origin:** Image origin: Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain.

b



**Description:** Sagittal STIR MR shows T4 uniform collapse (vertebra plana). T9 and T10 partial collapse. **Origin:** Image origin: Antoni Bosca. Department of Radiology, Hospital Clínico Universitario de Valencia, Valencia, Spain