MAR Oncology & Hematology (2023) 6:1

Case Report

Langerhans Cell Histiocytosis

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Received: 14 June 2023

Published: 01 July 2023

Mr . XYZ

- Age - 24 years
- Sex - Male
- Hailing from Kolkata
- Education – Hotel management student
- Right handed individual

Chief complaints

- Back pain since 1 year

History of presenting illness:

- Patient was apparently normal 1 year ago following which he developed.
- Insidious onset of gradually progressive
- Continuous dull aching type of non-radiating mid and low back pain
- Aggravates on bending forward.
- Relieved on taking rest and with NSAIDs.
- Patient revealed that pain has aggravated significantly since past 3 months, to an extent that patient has difficulty in performing his activities of daily living.

No history suggestive of motor weakness

- No higher mental dysfunction symptoms
- No history suggestive of cranial nerves involvement
- No history suggestive of cerebellar system dysfunction
- No bladder and bowel disturbances

- History of fever since 3 months
- Intermittent
- low grade
- Not associated with chills and rigors
- Relieved with medications.
- History of loss of weight - approximately 20 kgs in past 3 months
- Negative history: No history of cough with expectoration, hemoptysis and breathlessness
- No history of chest pain, syncopal attacks
- No history of pain abdomen, vomiting
- No history of peripheral joint pains or swelling of joints
- No skin lesions.
- For above mentioned complaints, patients visited local hospital 2 months ago in West Bengal , where MRI spine was done which was reported as abscesses at multiple vertebrae and FNAC was done from the abscess site.
- Patient was advised for Anti tubercular drug therapy with the diagnosis of TB SPINE, however for unknown reasons patient discontinued the medication within a week.
- PAST HISTORY Not a known case of type 2 Diabetes mellitus, systemic hypertension, bronchial asthma.
- No history of tuberculosis in the past, no history of exposure to tuberculosis patients in recent past.
- No history of trauma to back.
- Personal historyConsumed mixed diet
- Appetite: Normal
- Sleep: Disturbed
- Bowel and bladder: Regular
- No substance abuse.

- ON EXAMINATION

Young male

- Moderately built and poorly nourished with BMI 15.1kg/m2
- No pallor ,icterus ,cyanosis , clubbing , lymphadenopathy , pedal edema .
- Gibbus in mid thoracic region
- Tenderness present in mid thoracic region
- No neuro-cutaneous markers.
- Vitals:
- Pulse: 80bpm in right radial artery, regular normal volume and character all peripheral arterial pulsations well felt and normal
- BP: 110/70mmhg measured in right brachial artery in supine position no postural variation.
- SPO2: 97% on room air
- RR: 20 cycles per minute
- Temp: 98.1F
- CNS examination Higher mental functions:
- Conscious, oriented to time, place and person
- Memory [immediate, recent, remote] - intact
- No speech disturbances
- MMSE: 30/30
- No cranial nerve dysfunction signs

Motor System Examination:

- No wasting of muscles
- Hypertonia in all 4 limbs
- Power in all 4 limbs : 5/5
- Deep tendon jerks of both upper and lower limbs - exaggerated
- Plantar : Bilateral extensor response present
- No sensory abnormality noted
- Cerebellar system : no dysfunction signs
- Gait : normal
- No autonomic disturbances
- CVS : S1S2 heard –normal
- No S3,S4
- No mumurs
- RS : Bilateral normal vesicular breath sounds heard
- No added sounds.
- P/A : soft , non-tender
- liver and spleen not palpable
- bowel sounds heard
- PROBLEMS : Chronic gradually progressive dull aching , non radiating type of lower back pain S/O vertebral bony pain
- H/O chronic intermittent fever
- H/O chronic weight loss
- Gibbus in mid dorsal spine
- Tenderness in mid dorsal spine

Differential Diagnosis

Spinal Tuberculosis

- Pyogenic spondylodiscitis
- Brucella spondylodiscitis
- Lymphomas
- Leukemias
- Metastasis to vertebrae
- Investigations

• **Investigations**	• **Report**
• **WHITE BLOOD CELL COUNT**	• **17000/cumm**
• **HEMOGLOBIN**	• **11g/dl**
• **PLATELET COUNT**	• **4,71,000/cumm**
• **NEUTROPHIL COUNT**	• **90.8%**
• **LYMPHOCYTES COUNT**	• **5.5%**
• **LIVER FUNCTION TEST**	• **WNL**
• **BLOOD UREA ,SERUM CREATININE**	• **WNL**
• **SERUM ELECTROLYTES**	• **WNL**
• **ESR**	• **51 MM/HR**
• **CRP**	• **62MG/DL**

- PERIPHERAL BLOOD SMEAR : Neutrophilic leukocytosis
- BLOOD CULTURE AND SENSITIVITY : No growth after 72 hours of incubation
- ECG : Sinus rhythm , no ischemic changes

USG Abdomen:

Abdominal lymphadenopathy

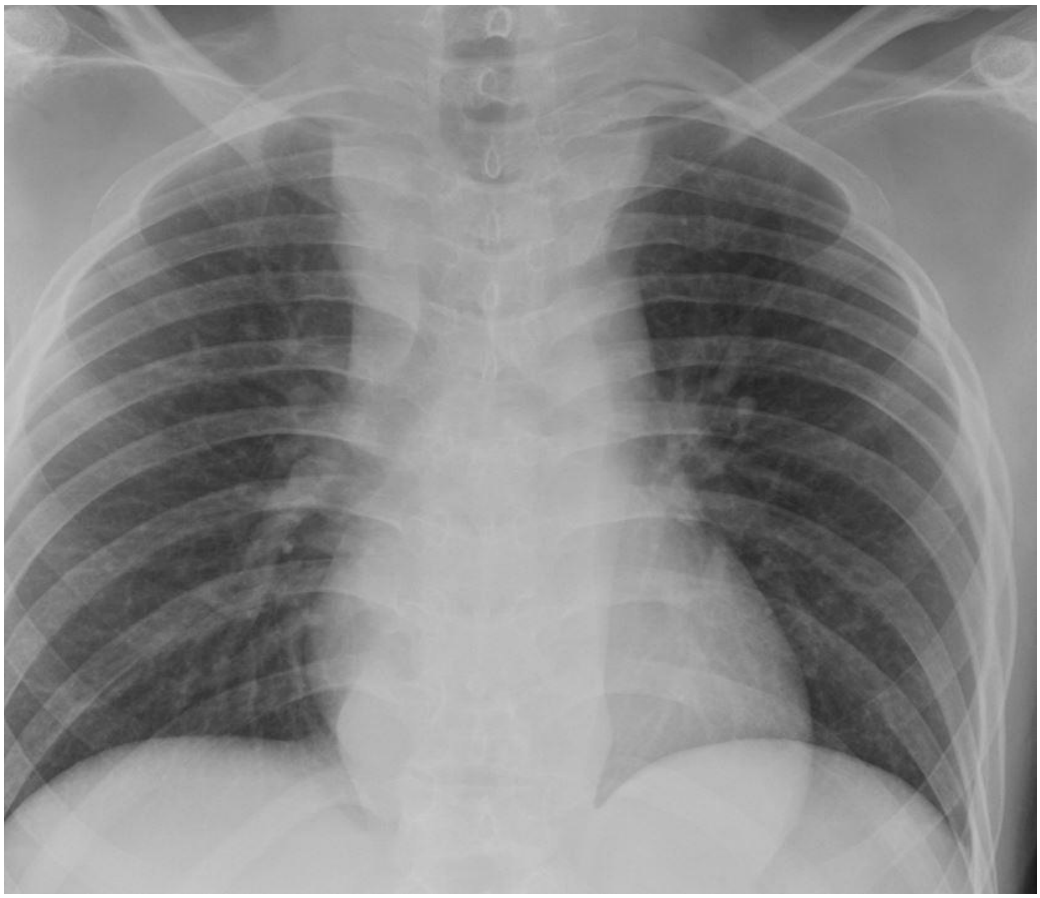
Paraaortic and paracaval lymph nodes

Largest size :11X 7 mm

- 2D ECHO : NO RWMA , EF :60%
- CHEST X-RAY PA VIEW :

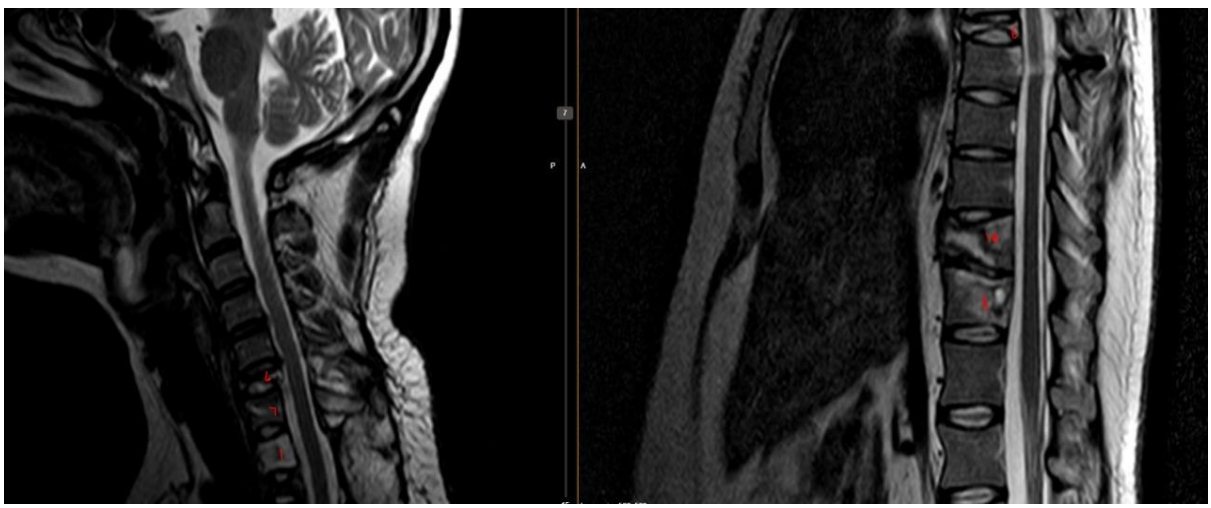
Mediastinal widening

lung fields are clear

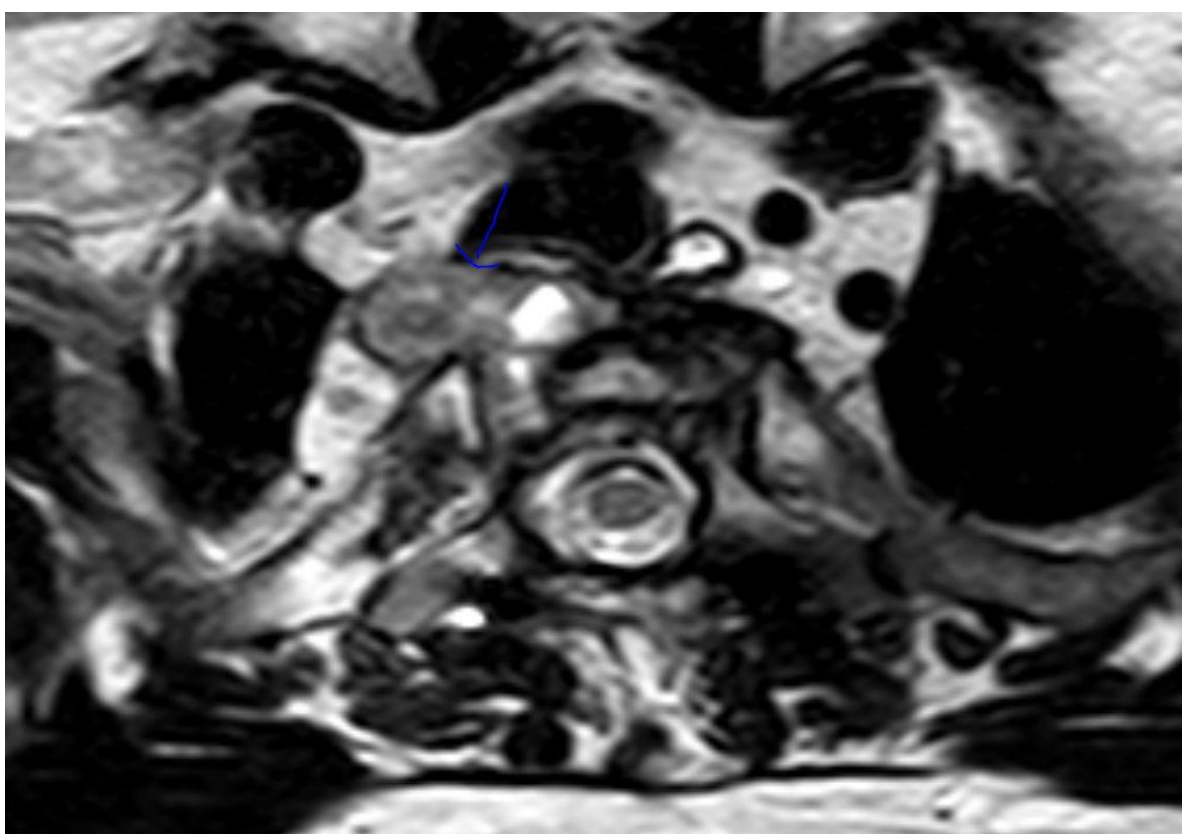


MRI Whole Spine

C6, D1, D2, D3, D6 and L3 vertebral body collapse with intact disc architecture.



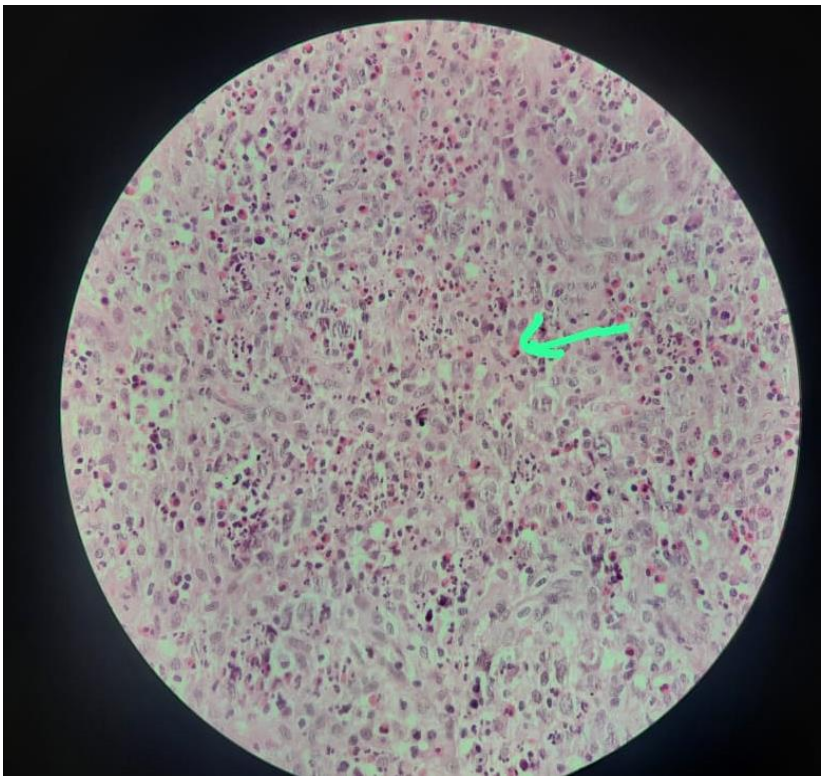
Soft tissue shadow in right psoas muscle



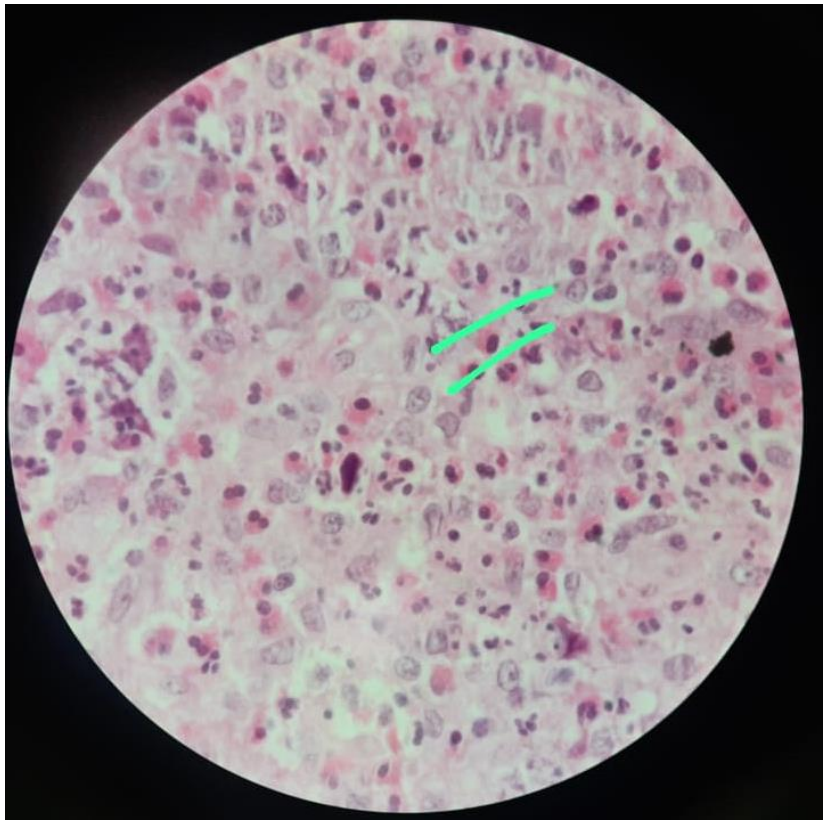
Features	Spinal TB	Pyogenic spondylodiscitis	Spinal metastasis	Brucella spondylodiscitis
Common region	Thoracolumbar	Lumbar	Thoracic	Lumbar
Predilection	Disc and bodies Major soft tissue lesions	Disc and bodies Less soft tissue involvement	Pedicles, lamina, posterior body wall	Disc and bodies Minimal soft tissue involvement
Risk factors	Exposure to TB	Diabetes, immunosuppression	Systemic malignancy	Unpasteurized milk
Clinical Pointers	Chronic back pain, constitutional symptoms	Acute severe back pain, fever	Night pains, constitutional symptoms.	Fever, backache, malaise

Biopsy Was Taken from D6 Level

Showed numerous eosinophils



Langerhans cells with grooved coffee bean nucleus

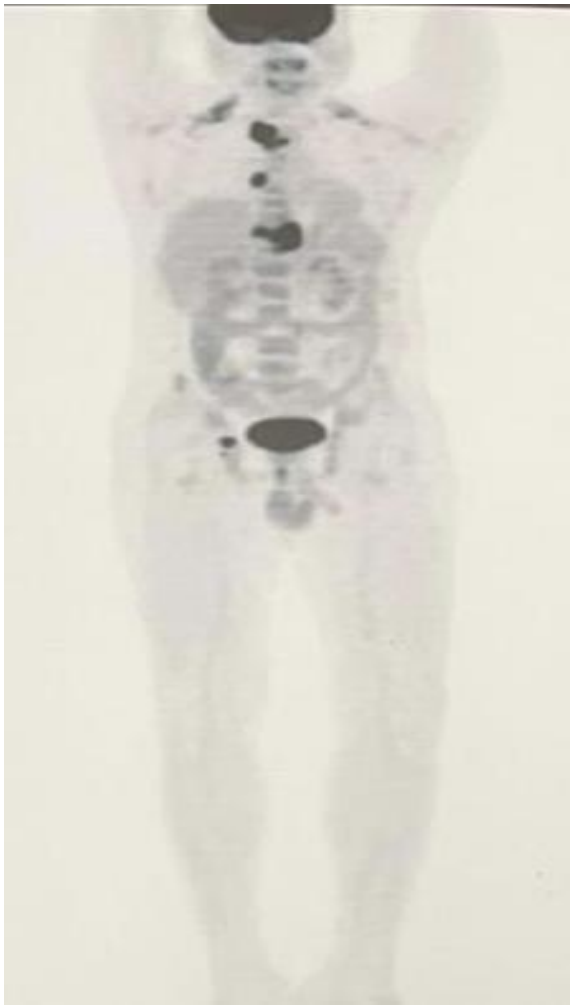


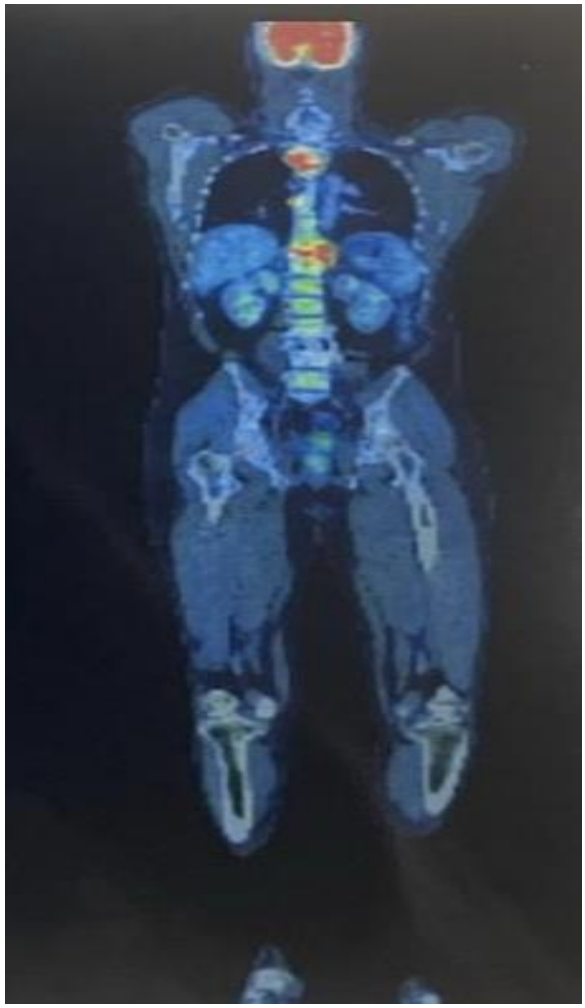
Immuno Histochemistry

- Histocytes are labelled by CD 68
- Langerhans cells are labelled by CD 1a and S100

PET CT scan:

Multiple metabolically active osteolytic skeletal lesions , supra and infradiaphragmatic lymphadenopathy

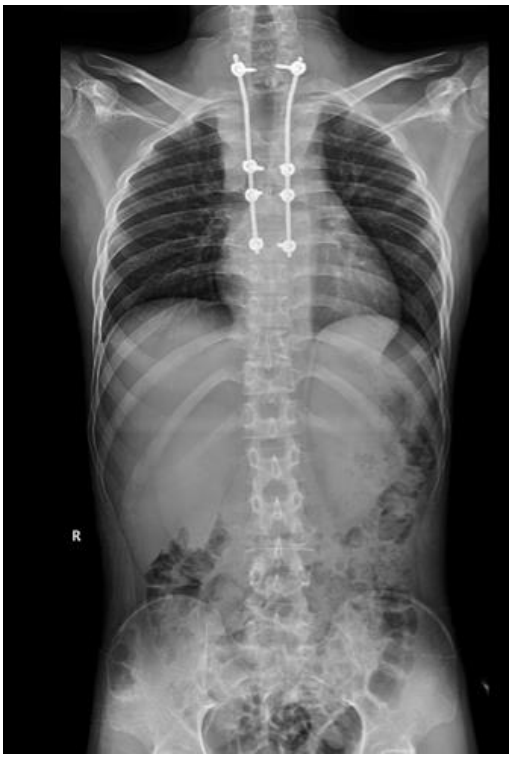




With the above findings , patient was diagnosed with

Langerhans histiocytosis.

Patient was initially taken for D1,D3 and D6 decompressive laminectomy ,C7,D4,D5,D8 transpedicular screw and rod fixation.



- Treated with INJ VINBALASTINE 7MG IV BOLUS WEEKLY for 4 weeks
- Oral PREDNISOLONE 20MG 1-1-1 for 4 weeks and gradually tapered over a period of 2 weeks.
- The patient was followed and presently in remission.
- Langerhans Cell Histiocytosis: Group of multiple-systemic diseases involving bone marrow, internal organs, skin, and mucosae, characterized by a histiocytic proliferation of granuloma-like aspect .

Types:

Eosinophilic granuloma: localized disease

Hand–Schuller–Christian disease: disseminated form

Letterer–Siwe disease: disseminated and fatal form

Epidemiology

- Incidence:

4-5 cases per million per year in children <15years

1-2 cases per million per year in adults

- Male preponderance
- 5–10 years of age, adults can also be affected.
- ETIOLOGY: The exact etiology of LCH still is unknown.
- Langerhans cells belong to the normal human cellular environment, especially the mononuclear-phagocytic system, their transformation to a pathologic growth pattern remains unclear.
- SITES: Multisystem disease that can affect any organ either in the isolated or in the disseminated forms.
- Within the skeleton, the cranial vault is the preferred site, but the mandible, the rest of the axial skeleton, and the long bones of the extremities follow closely.

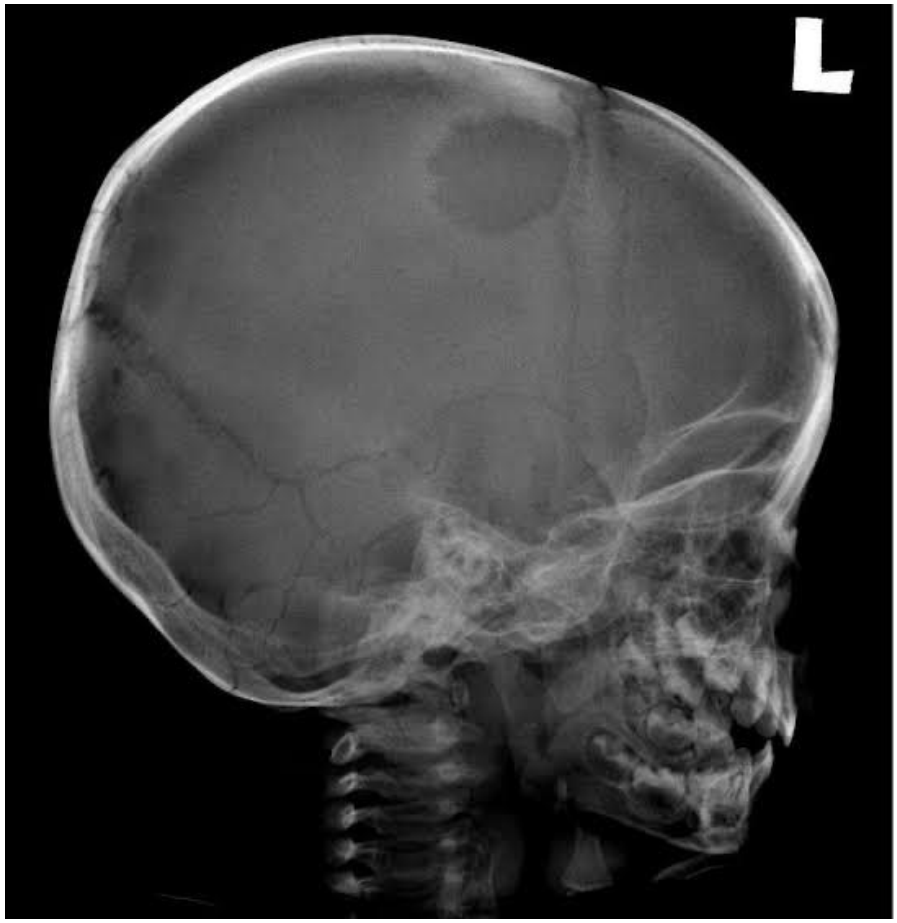
Clinical Features

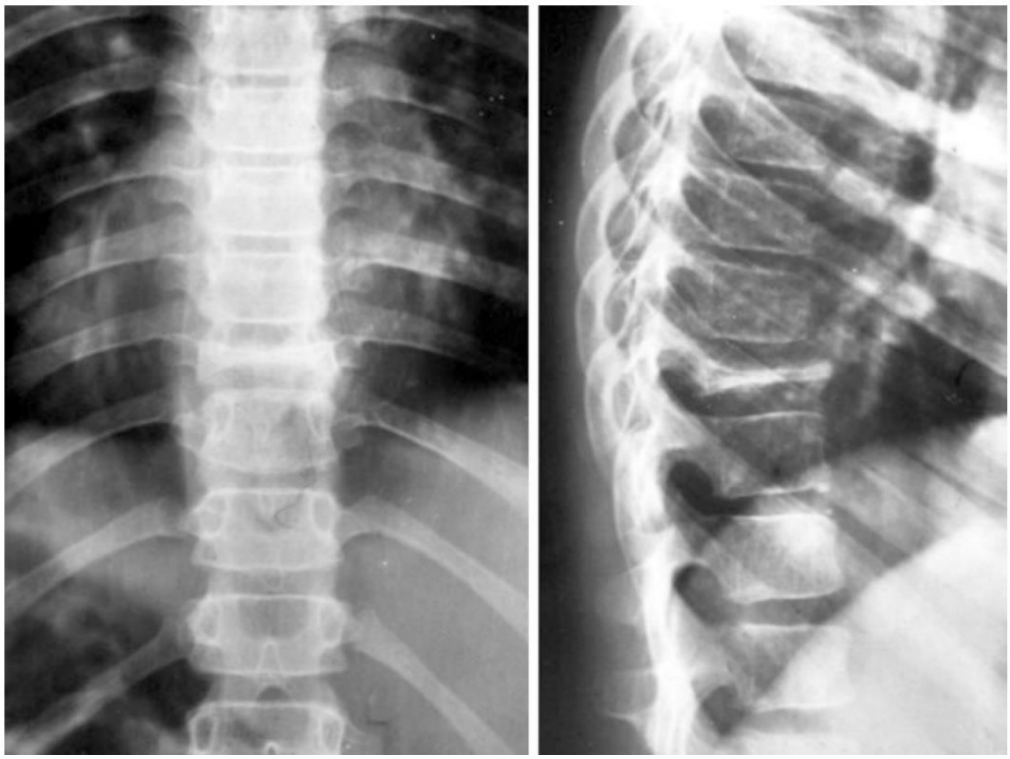
Pain and swelling over the area of the affected bone are usually present.

- Neurological symptoms may result from spine involvement.
- Mandibular lesions may cause loosening of teeth.
- Diabetes insipidus in 15–20 % of patients.
- Hepatosplenomegaly may occur.
- DIAGNOSIS

When the diagnosis of LCH is suspected, a complete skeletal survey must be done.

- In the cranial vault, lesions typically have a beveled or "hole-in-hole" appearance because of the difference in lesion diameter in the two bone tables.
- Vertebral involvement may produce a "vertebra plana," the total collapse of the vertebral body, with preservation of the intervertebral disks, but the lesion may also be expansile and lytic.
- The disease may involve the supporting structure of the teeth, producing the "floating-teeth" appearance.





- Histopathology : Soft, semi-liquid, yellowish- gray tissue, with areas of hemorrhage or necrosis with abundant eosinophilic granulocytes, sometimes forming eosinophilic abscesses.
- Langerhans cells are the diagnostic cells.
- Immunohistochemical panel

CD1a	**+**
S100	+
Langerin	+

Treatment

Surgery

- Chemotherapy
- Radiotherapy
- Bisphosphonates

Treatment

Surgery:

Corrective surgery can be necessary in children with bony lesions located in epiphyseal plates to prevent growth retardation.

Vertebra plana with spinal cord compression and associated neurological dysfunction requires extensive reconstructive interventions.

Chemotherapy

Vinblastine – prednisolone induction therapy:

Vinblastine 6mg/m2 intravenously once every three weeks plus prednisolone 40mg/m2 daily by mouth for 4 weeks ,then tapering over 2 weeks

Other drugs:

Mercaptopurine 50mg daily orally can also be used .

Cytarabine 100mg over 30 minutes daily for 5 days

Prognosis

Patients with solitary or a few isolated bone lesions have an excellent prognosis with and, sometimes, without treatment.

- Signs of worse prognosis and possible death of the disease are:

– Young age at diagnosis, less than 3 years of age

– Involvement of more than three bones

– Hematologic manifestations

– Hepatosplenomegaly

Conclusion and Take Home Message

LCH spine may closely resemble Pott's disease .

- Clinicians should be aware of LCH and has to be considered in the list of differential diagnosis when approaching a case of chronic back pain in children and young adults.

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