

Mastocytosis

- Excessive accumulation of mast cells in one or more organs.
- According to the World Health Organization classification, three clinical entities fall under the mastocytosis umbrella:
 - Cutaneous mastocytosis,
 - Systemic mastocytosis (with or without cutaneous manifestations)
 - Mast cell sarcoma. This article deals primarily with systemic mastocytosis.
- Indolent mastocytosis
- Normal life expectancy

Primary radiographic findings

- 2 distinct appearances of osseous structures
 - Osteosclerosis
 - Osteoporosis
- Secondary radiographic findings
 - Pathologic fracture, particularly in spine
- Low sensitivity and specificity for detection of systemic mastocytosis

Bone involvement in mastocytosis

- Either lytic, sclerotic or mixed
- Diffuse involvement tends to be more common
- Usually, the radiographic appearance is that of sclerosis involving primarily the axial skeleton and the ends of the long bones.
- In some patients, there is generalized osteoporosis with the risk of pathological fractures.

Non skeletal CT findings in 19% of patients in one study

- Hepatosplenomegaly
- Retroperitoneal lymphadenopathy
- Periportal lymphadenopathy
- Mesenteric lymphadenopathy
- Thickening of omentum and mesentery
- Ascites
- Less common findings
 - Hepatofugal portal venous flow
 - Budd-Chiari syndrome
 - Cavernous transformation of portal vein
 - Ovarian mass

MASTOCYTOSIS

(CLONAL MAST CELL DISEASE)

↑↑↑ NUMBERS of INCORRECTLY FUNCTIONING MAST CELLS

2 CATEGORIES:

**CUTANEOUS
MASTOCYTOSIS**

**SYSTEMIC
MASTOCYTOSIS**

MAST CELLS ACCUMULATE in:

SKIN

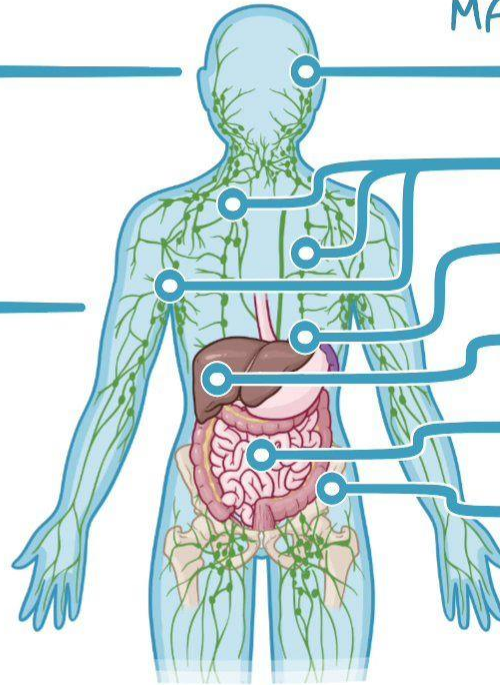
LYMPH NODES

SPLEEN

LIVER

GI TRACT

BONE MARROW



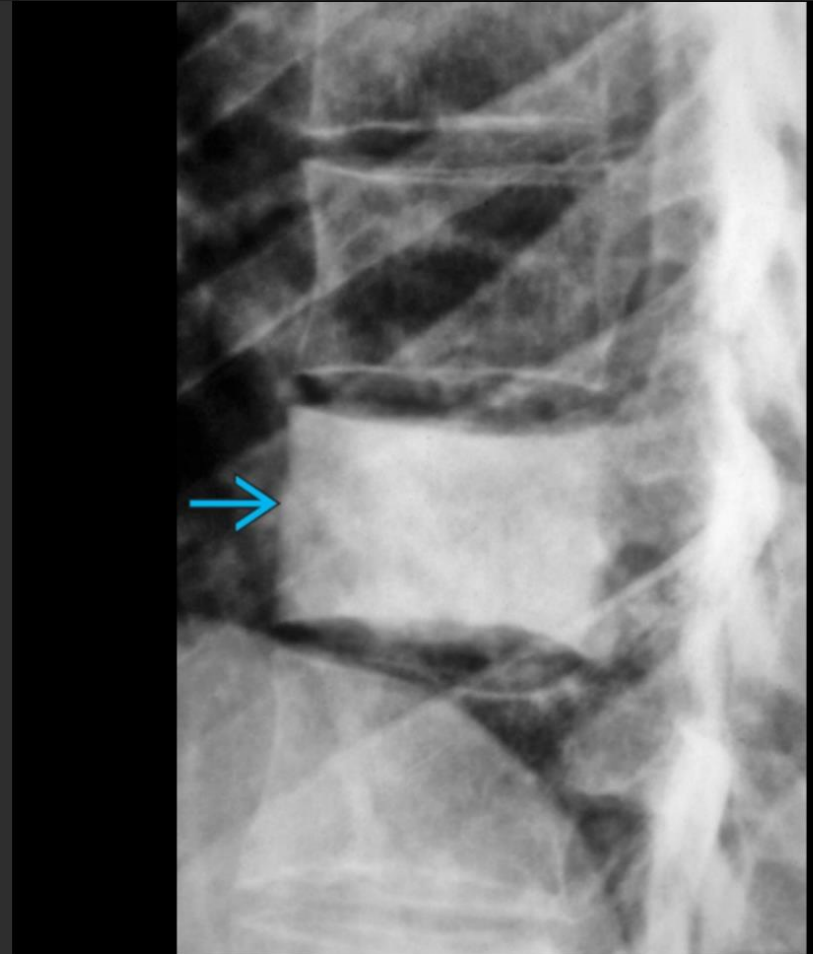


Mastocytosis

Systemic Mastocytosis



focal spots of sclerosis on a
background of diffuse
osteopenia



[View Full Screen Image](#)

Lateral view of the thoracic spine shows a classic ivory vertebra → with dense bone replacing the entire vertebral body with no change in size. Focal sclerosis, such as this, is one appearance of **mastocytosis**.



[View Full Screen Image](#)

AP radiograph shows diffuse and severe osteoporosis with multiple fractures. The appearance is nonspecific but was biopsy-proven **mastocytosis**. It is unusual for childhood **mastocytosis** to have osseous abnormalities.