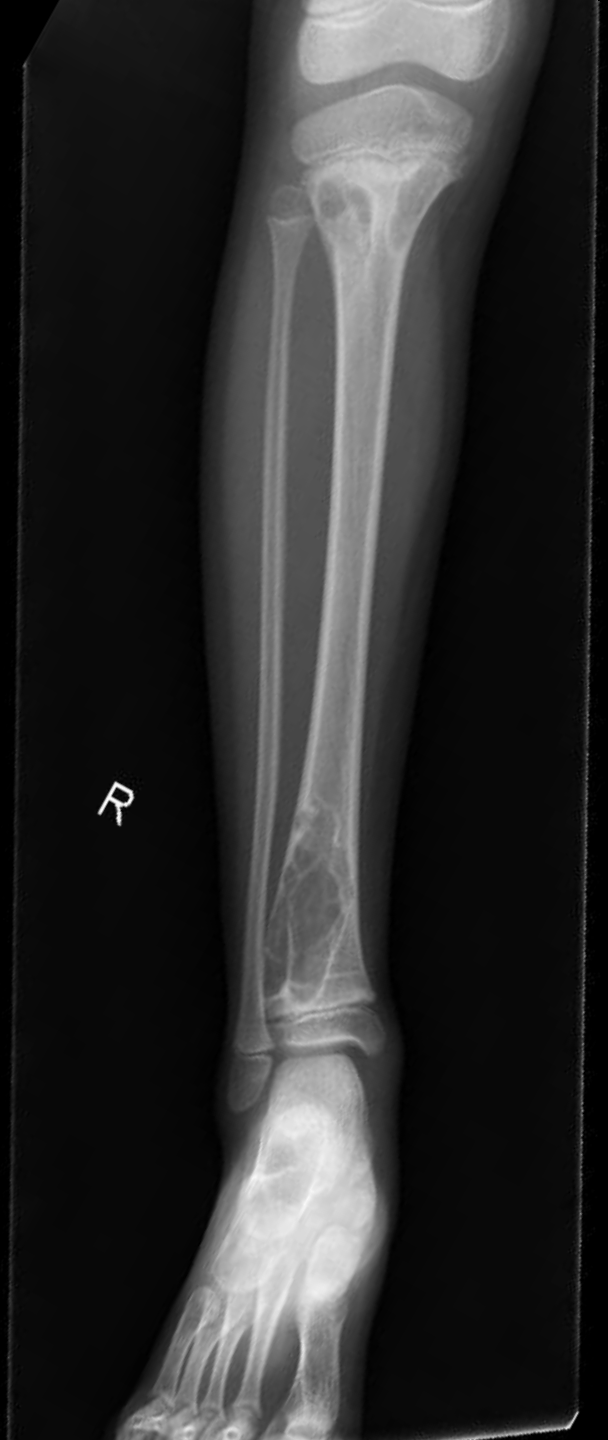
History

- 8M with leg pain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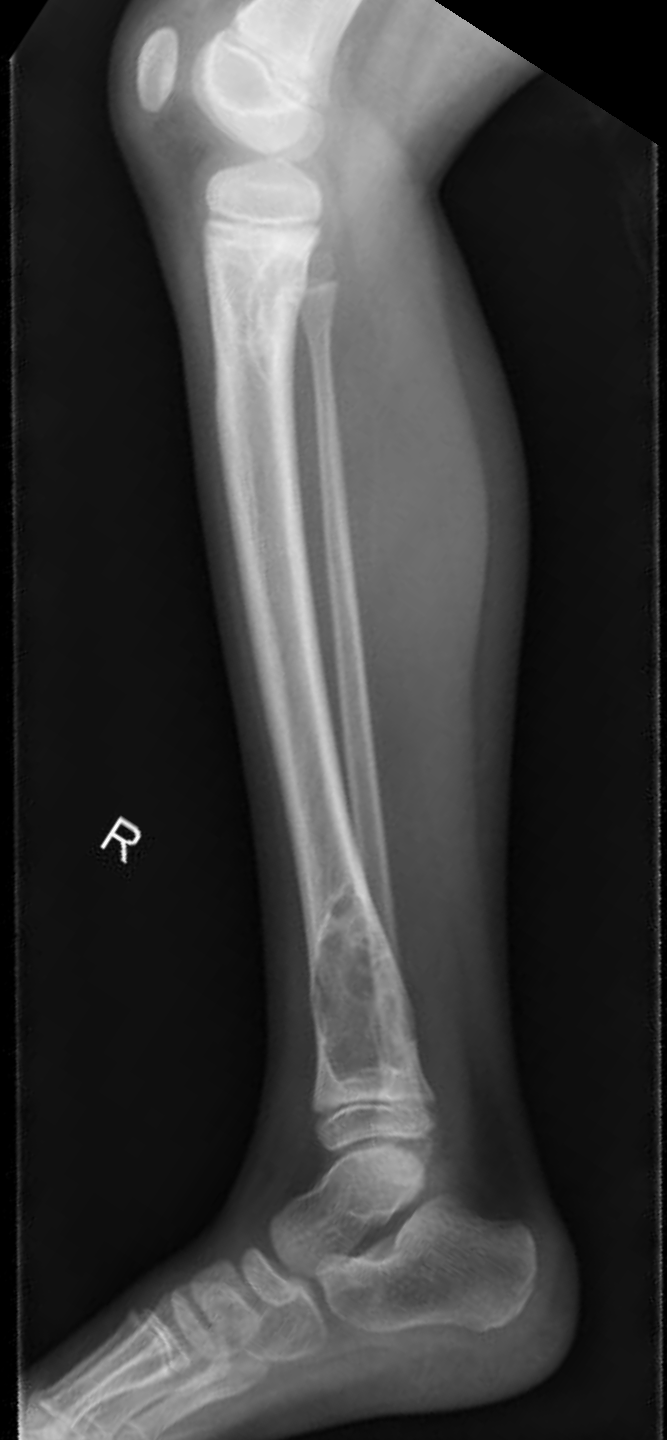
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Sag T1

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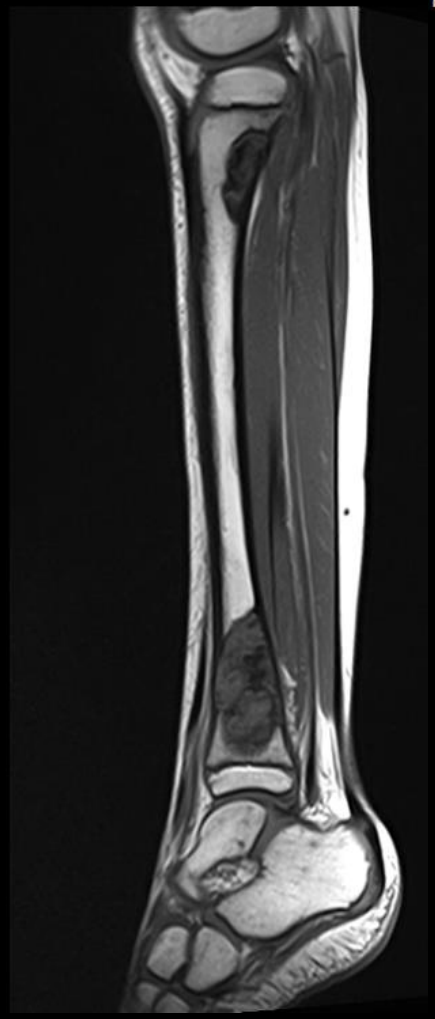

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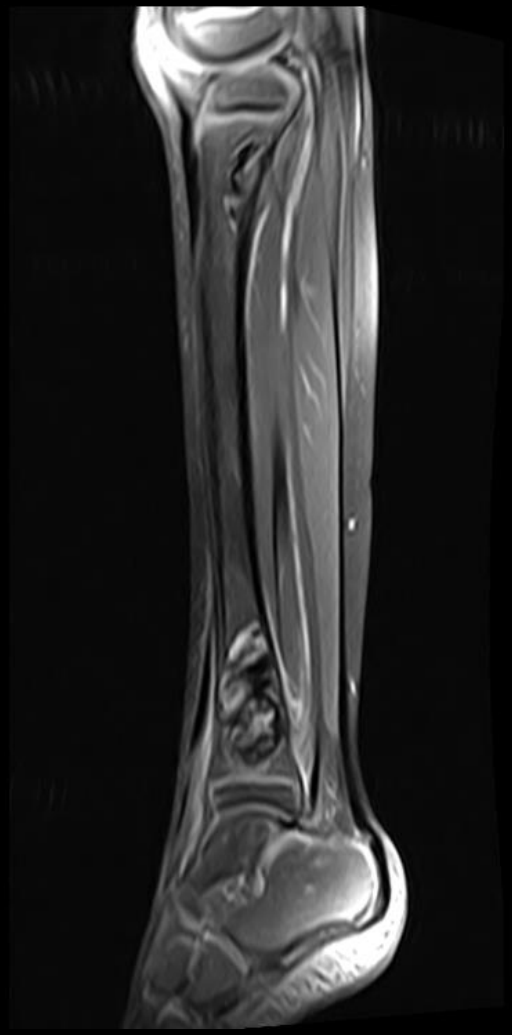

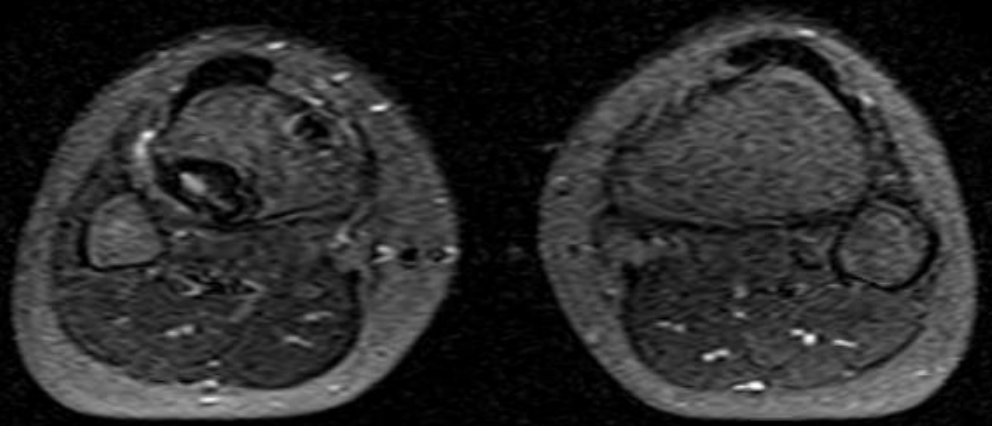

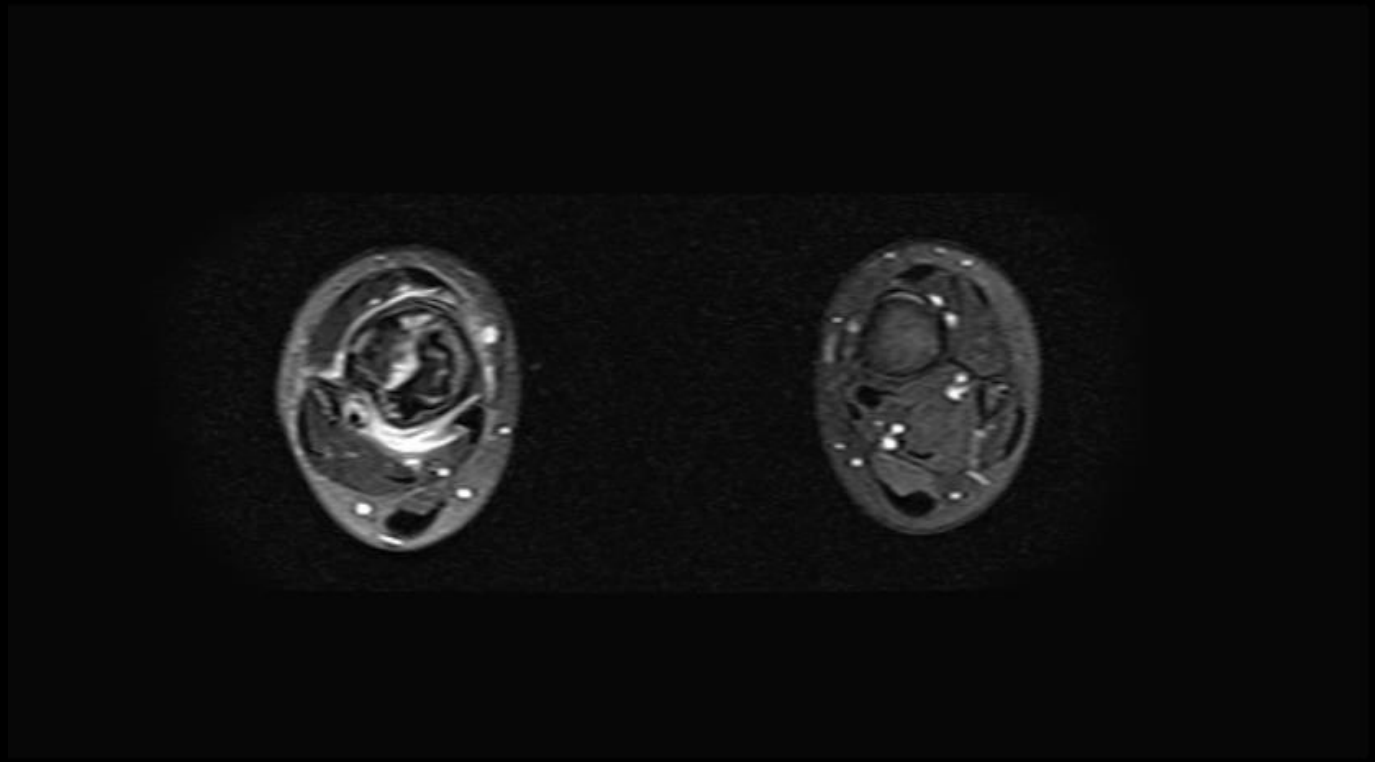

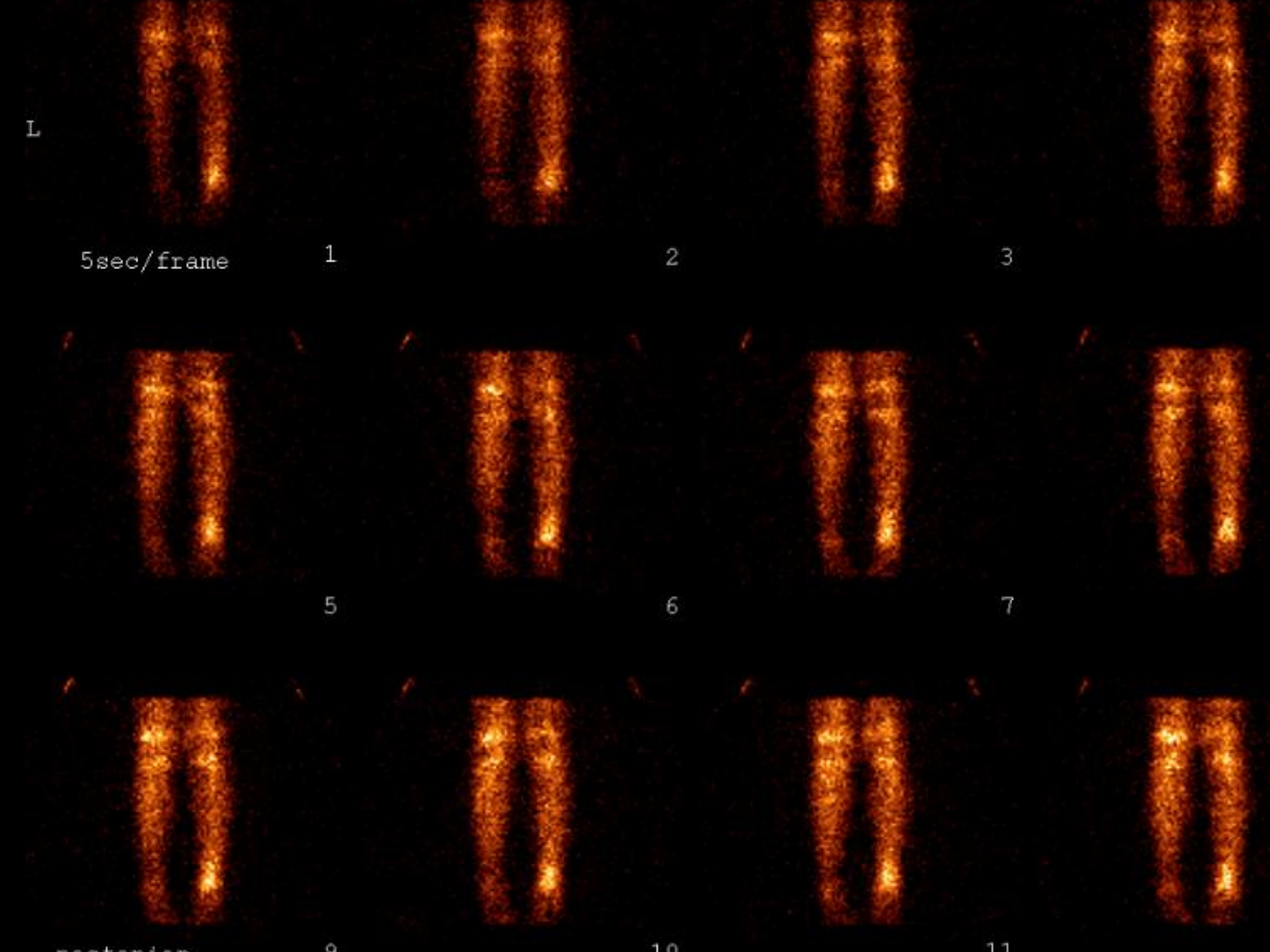
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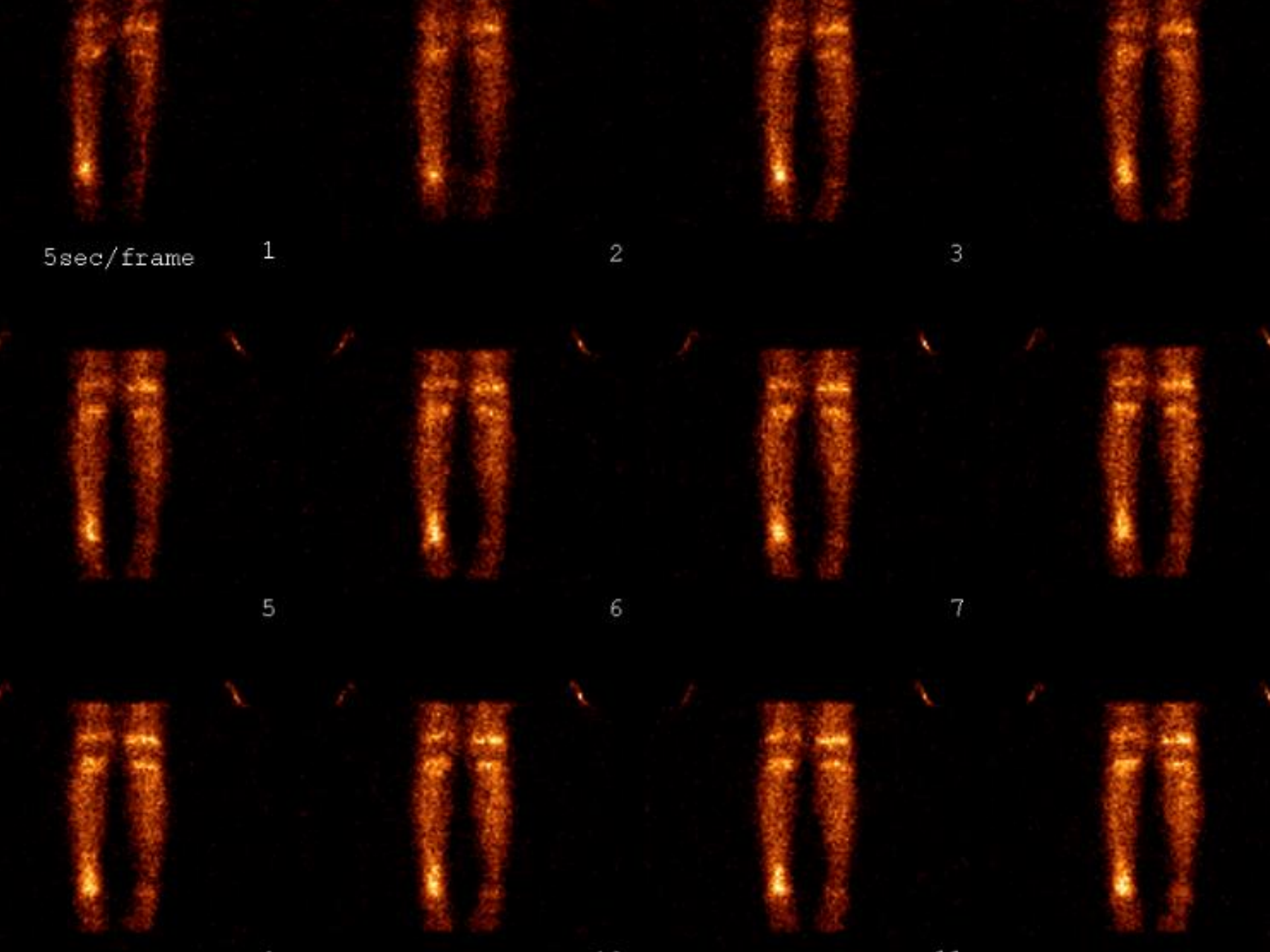
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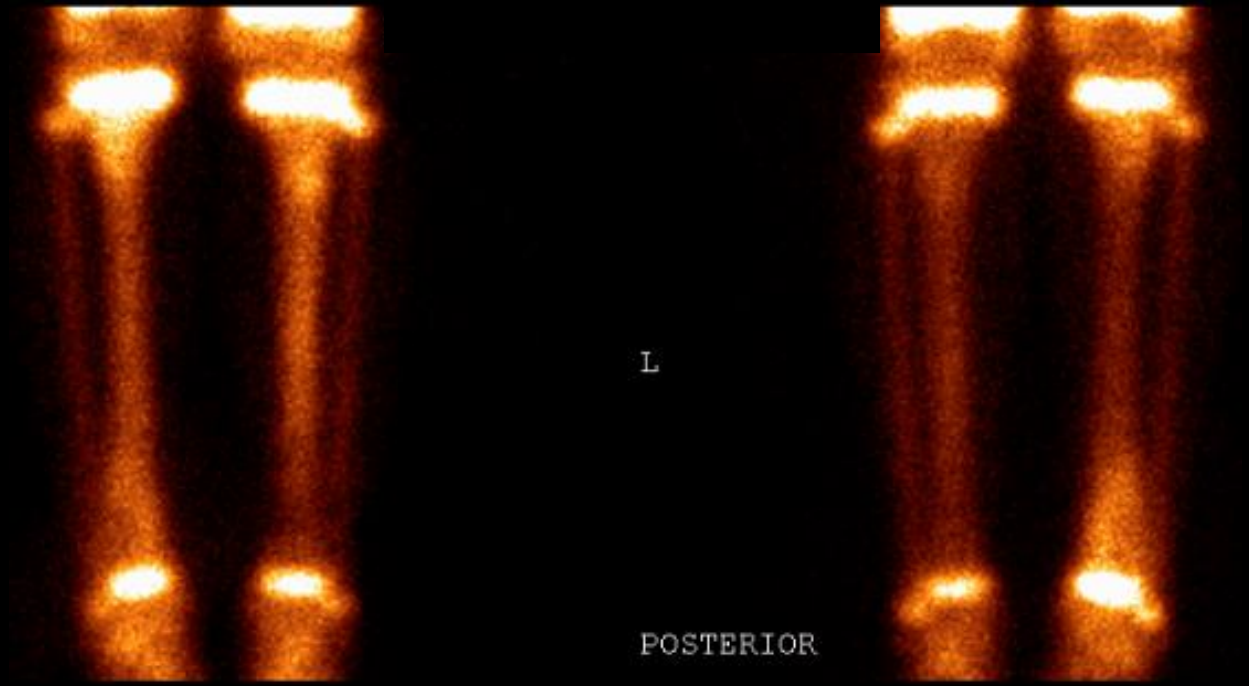
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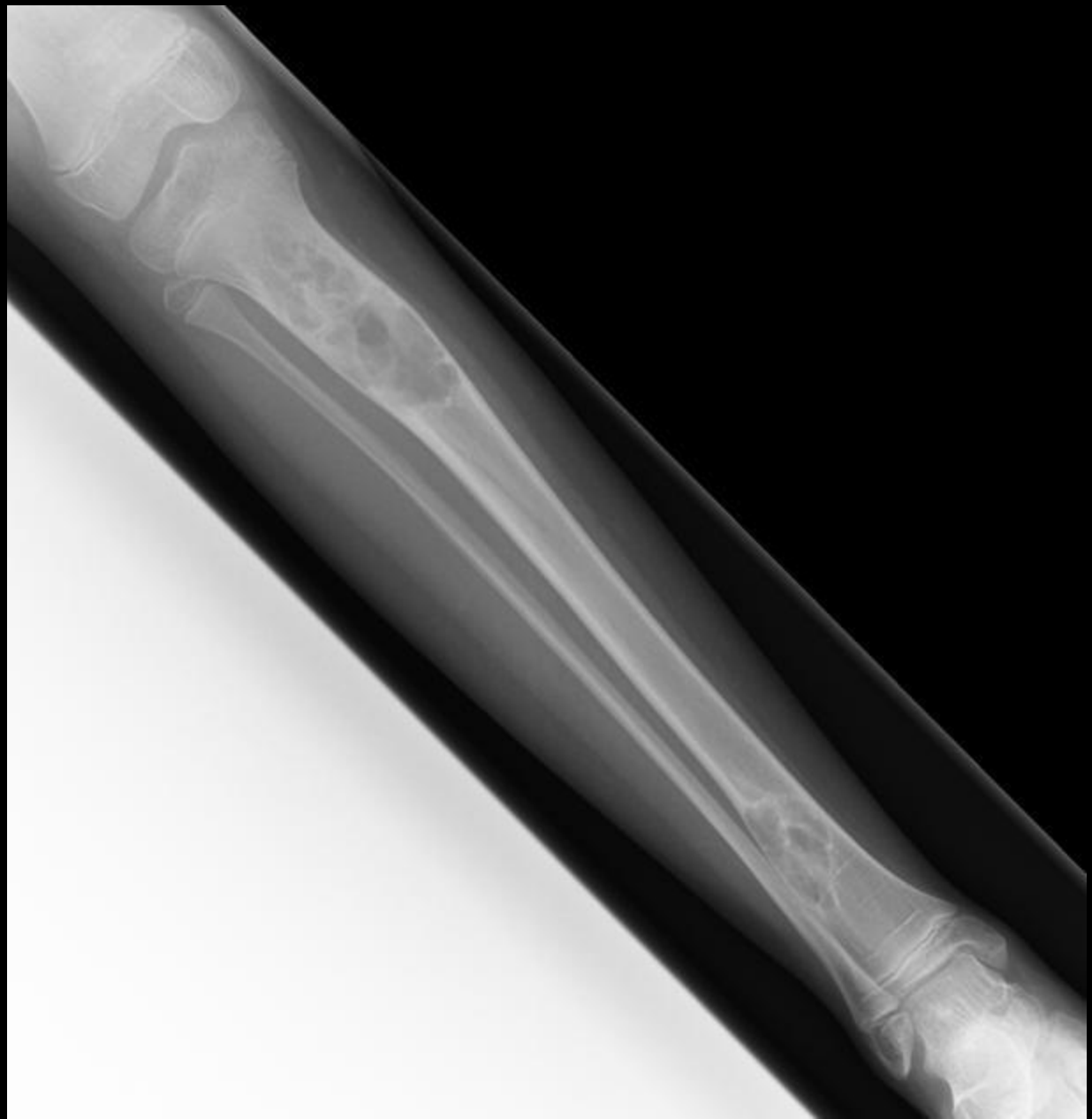
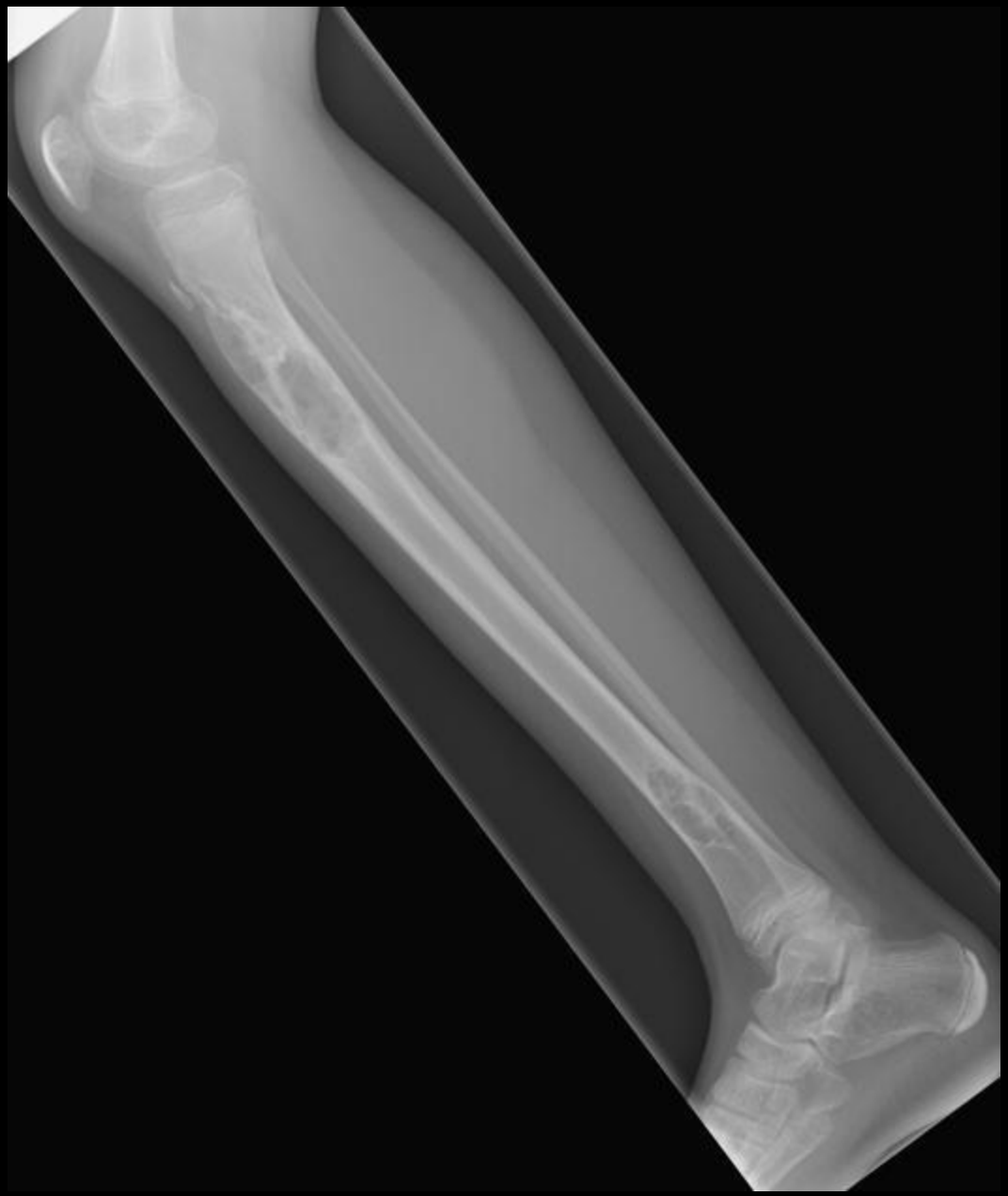
POSTERIOR

Differential Diagnosis

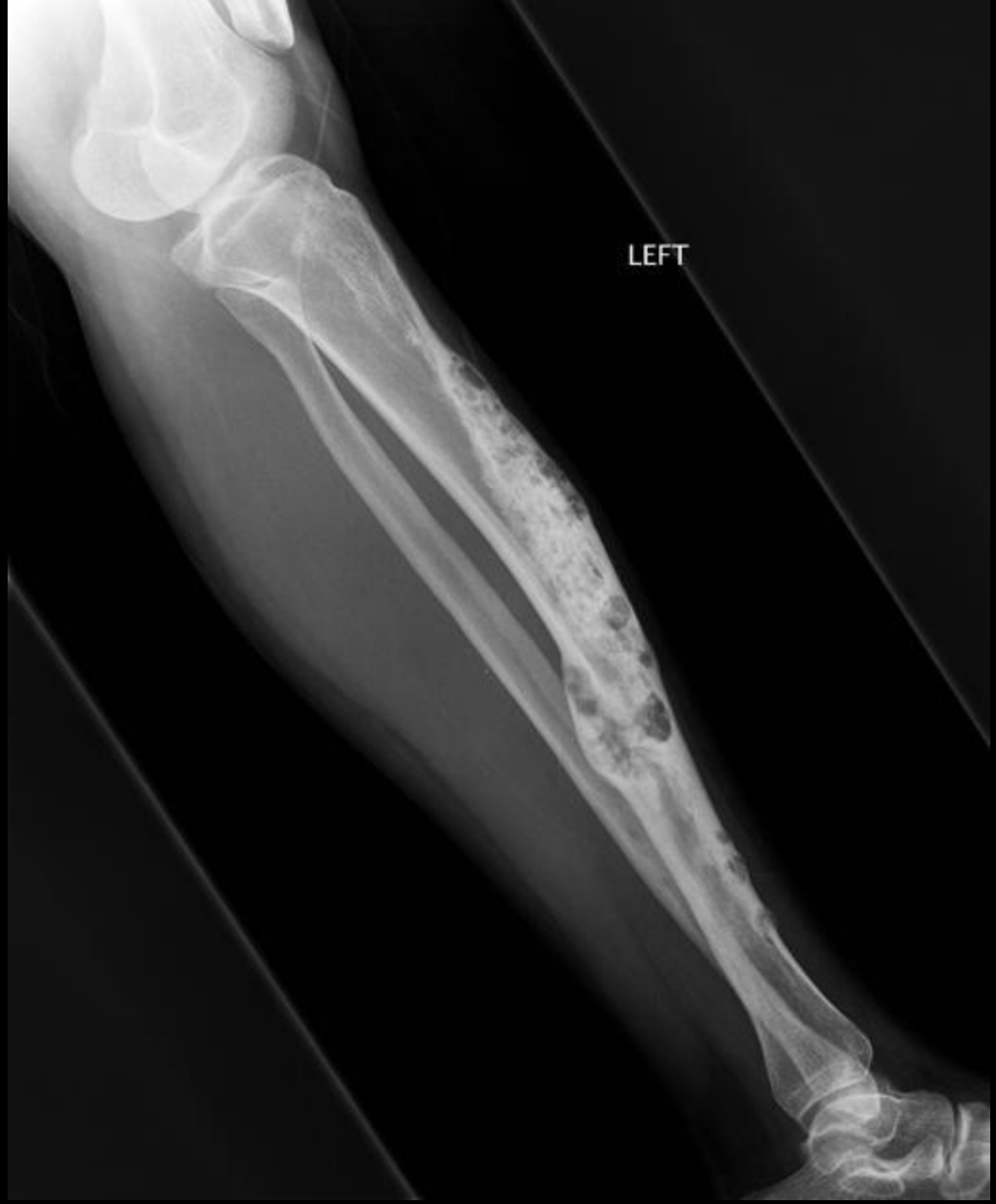
- Multiple nonossifying fibromas
- Polyostotic Fibrous Dysplasia
- Aneurysmal bone cyst
- Eosinophilic Granuloma
- Mets – neuroblastoma/leukemia
- Adamantinoma
- Infection – multifocal osteomyelitis

Polyostotic Fibrous Dysplasia





Adamantinoma



Synchronous lesions in fibula seen in 5-10%.

Jaffe-Campanacci syndrome

- 1942, Jaffe and Lichtenstein described nonosteogenic fibroma (now known as nonossifying fibroma) of bone

- 1958, Jaffe described a clinical entity in which multiple nonossifying fibromas occurred in association with *café-au-lait spots and axillary freckling,* but without accompanying neurofibroma.

Discussion

- diagnosis of this rare disease is usually made in the peripuberty years (ten to fifteen years of age), although the age of presentation may range from four years to more than eighteen years

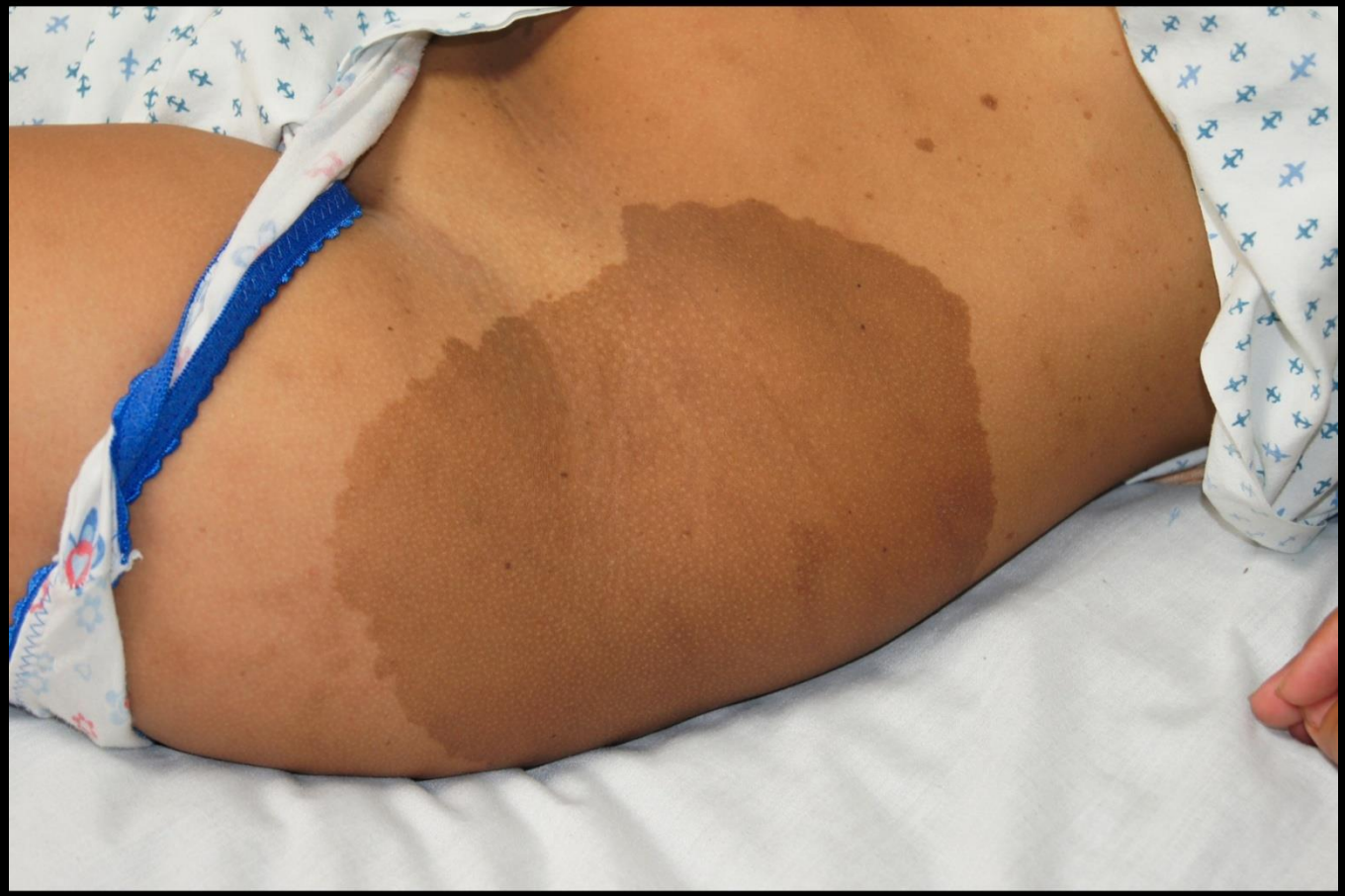
- Males and females seem to be affected equally

- Most patients have no family history of familial disease or neurofibromatosis

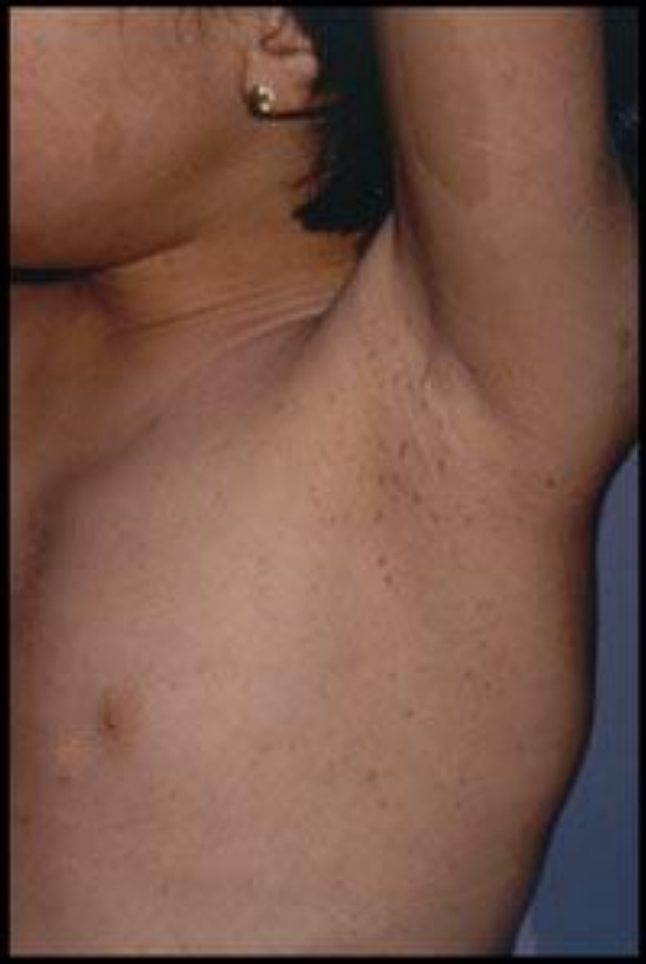
- The patients often present with a pathologic fracture through a nonossifying fibroma in the lower extremity.

- The clinical finding of smooth-bordered “coast of California” *café-au-lait spots and axillary freckling in* association with multiple nonossifying fibromas without accompanying skin, subcutaneous, or deeply placed neurofibromas is considered to be characteristic of the Jaffe-Campanacci syndrome.

Coast of California



Axillary Freckling “Crowe’s Sign”



Refrences

- Moser,R. Multiple skeletal fibroxanthomas: radiologic-pathologic correlation of 72 cases. *Skeletal Radiology*. 1987. 16:353-359.
- Anwar Hau,M. Jaffe-Campanacci Syndrome a case report and review of the litetrature. *Journal of bone and joint surgery.* 84: 634-638.