PHOTO QUIZ

What is your diagnosis?

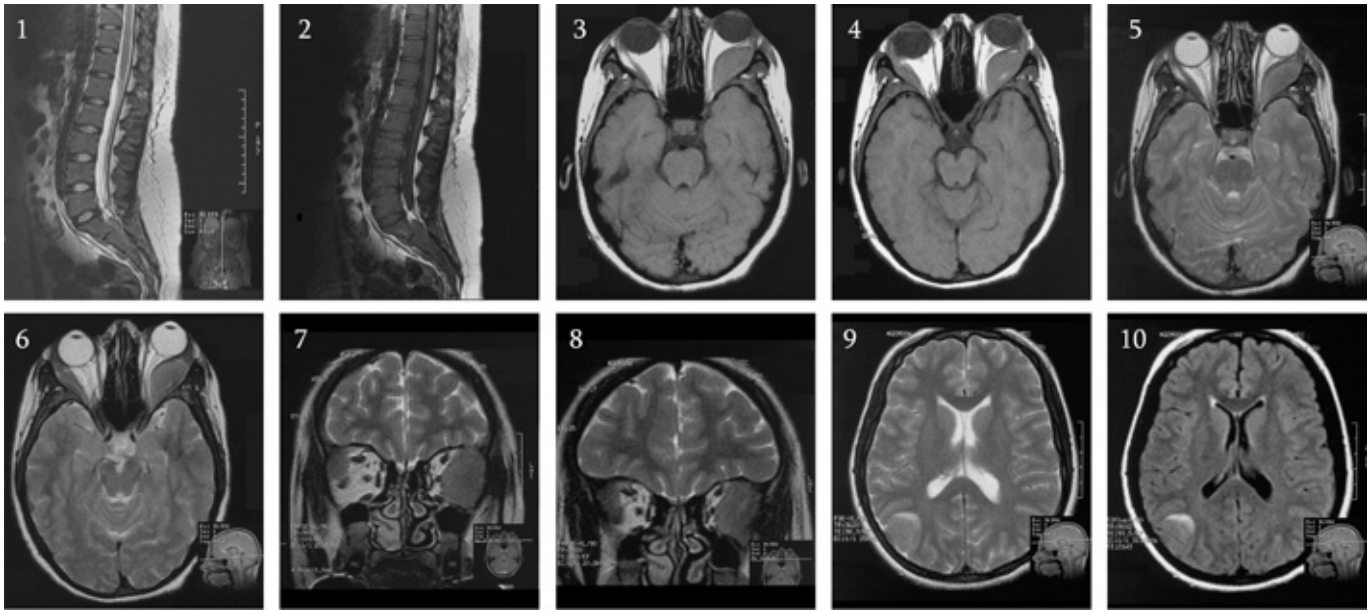

Fig 1. sagittal T2/ W: extradural soft tissue mass in sacral canal and in presacral region.
Fig 2. sagittal T1/W: Generalized decreased bone marrow signal intensity in lumbosacral spine associated with soft tissue mass in sacral canal.
Figs 3-8. axial T1(3,4) &T2(5,6) and coronal T2: extraconal soft tissue mass in lateral portion of both orbits with medial displacement of lateral rectus muscles.
Figs 9 (axial T2), 10(axial flair). extra-axial isosignal soft tissue mass in right parieto- occipital region with peripheral edema

A 21- years- old girl presented with diarrhea, left sided proptosis, and eyelid swelling.

At first, the patient was treated as a case of allergy, but she had recurrence of symptoms plus malaise, weakness, and generalized pain. The patient was admitted, and by primary lab test results (raised WBC and ESR, low PLT), the physicians suspected to connective tissue disease, so prednisolon was started.

In the course of the disease, the patient got abdominal pain, melena, and hematuria, so she was admitted again.

Abdominal sonography was normal, however; as the patient developed headache, proptosis and low back pain, brain and lumbosacral MRI was done in our department.

The MRI findings included a massive extracranial mass lesion in the left orbit causing proptosis and medial displacement of left lateral rectus muscle. There was also similar small lesion in the right orbit, and finally an intracranial extra-axial mass lesion in the right posterior parietal region.

Lumbosacral spine image showed soft tissue mass within lower spinal canal associated with destruction of S2 and S3 segments, and subtle enhancement in the lesion in post injection images.

What is your diagnosis?

Diagnosis: Chloroma (Granulocytic Sarcoma)

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Leukemia is the most frequent malignancy of childhood. In children, most cases are from acute lymphoblastic leukemia Bone pain and demonstrable skeletal lesions are common[1]. But in adults, leukemic bone lesions are much less frequent, occurring in less than 10% of cases, and the most common findings are diffuse osteoporosis or focal areas of osteolysis. On X-ray, study particular affected areas are the spine, ribs, skull, and pelvis.[2]

Chloromas are extramedullary soft tissue tumors composed of early myeloid precursors, which are well-known complications of myelogenous leukemias.[3,4] These locally invasive masses are referred to as **granulocytic sarcomas**; the distinction is based on demonstration of green pigmentation on pathologic study. Chloromas are somewhat more frequent in the acute myelogenous leukemias, although these are encountered in less than 10% of patients.[5] They can occure before, during or after the onset of systemic leukemia,or even when the patient is in clinical remission.[5,6] Younger patients are affected more commonly than older ones.

Chloromas may occur anywhere (most commenly in bone and nerve tissue), more often in the spine and make symptoms of nerve root or cauda equina compression.[4,5,7] They are usually extradural but may occasionally either intradural or both intradural and extradural(dumbbell-type).[7] They may destruct adjacent bone and make pathologic fracture.[8]

Multiple spinal tumors may be encountered, and either nerurofibromatosis or lymphoma may be suspected initially.[5]

CT-scan is superior to X-ray in demonstrating both the extent of bone destruction and associated soft tissue mass. CT with intratechal contrast is useful to demonstrate of abnormally thickend nerve roots due to leukemic infiltration.[5,7]

On MRI, the leukemic infiltrate within the marrow is isointense or mildly hyperintense on T2–weighted images, and hypointense on T1-weighted images. An inflammatory or necrotic process involving the marrow appears as hypointense signal on T1- weighted, and hyperintense on T2-weighted images. Lukemic infiltration of nerve roots and adjacent intraspinal and paraspinal soft tissue can be better demonstrated on MRI. In addition, the multiplanar capabilities of MRI allow visualization of larger segments of the spine in coronal and sagittal palnes. MRI is useful in diagnosis of recurrent disease, confirmation of remission, and as a screening test in detecting focal lesions, providing a guide for direct needle biopsy of the marrow site.[8,9]

In the brain, graulocytic sarcomas project inward from the meninges or within the brain tissue near the ventricular wall ependyma in the cases with systemic leukemia, typically AML or CML.[10] On CT-scan, the lesion appears hyperdense or isodense relative to brain parenchyma with unsharp margins. On MRI, it appears as heterogenous isointensity or hypointensity relative to gray matter on T1-weighted images, and iso- to hyperintensity on T2-weighted images. Homogenous contrast enhancement of the tumor is the rule.[10]

Recognition of Chloromas is important because they are extremely sensitive to radiation, which may be preffered over or combined with chemotherapy. Surgery is usefully reserved for symptomatic spinal cord compression.

In our case, BMA and BMB were taken and showed normocellular and myeloid hyperplasia with Teardrop cells. Immunotyping revealed AML (compatible with M2) and cytogenetic study conclusion was specific for AML–M2.

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