CASE OF THE MONTH

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FELLOW (ONCOPATHOLOGY)

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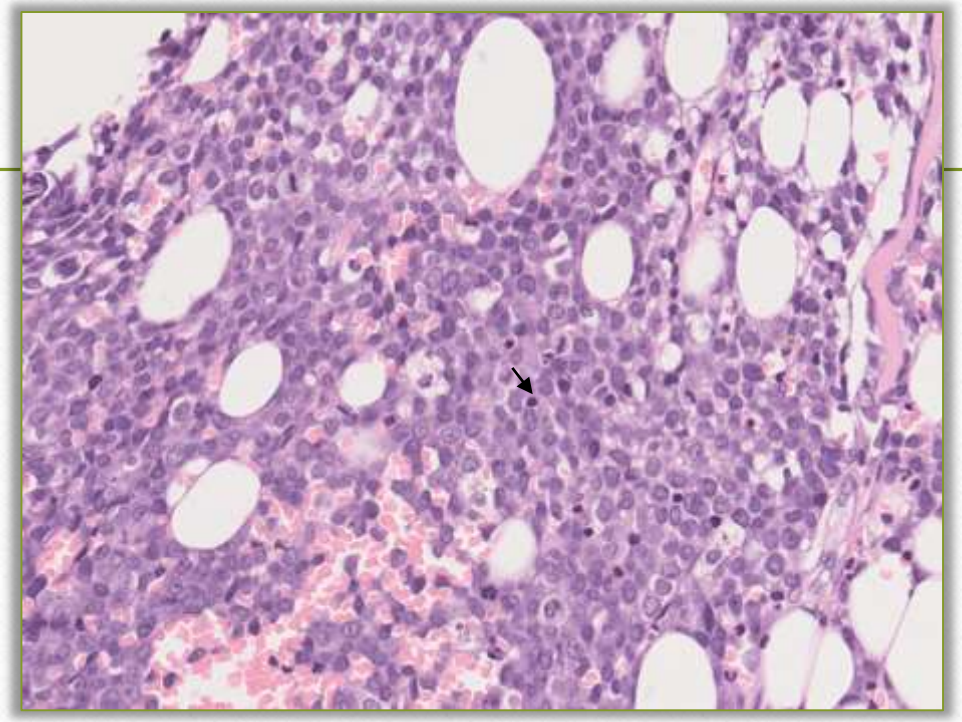
CASE

- A 29 year old male came with complaints of pain and weakness in right arm since 1.5 months.
- On examination, there was motor weakness in shoulder abduction, flexion, extension, elbow flexion and extension.
- There was no sensory loss.
- CBC and Peripheral Blood Smear was normal.

- On CT scan, soft tissue lesion at C4-5, C5-6 level was identified.
- On PET scan, multiple lytic sclerotic lesions were noted in right humerus, sternum, bilateral clavicles, bilateral scapulae, few ribs, multiple cervico dorsal lumber vertebrae, sacrum, bilateral pelvic bones, bilateral femori and right tibia. Few mildly avid subcentimeter sized right common iliac and internal iliac lymph nodes were also noted.
- On the basis of these findings, possibility of disseminated metastasis and lymphoma was suggested.

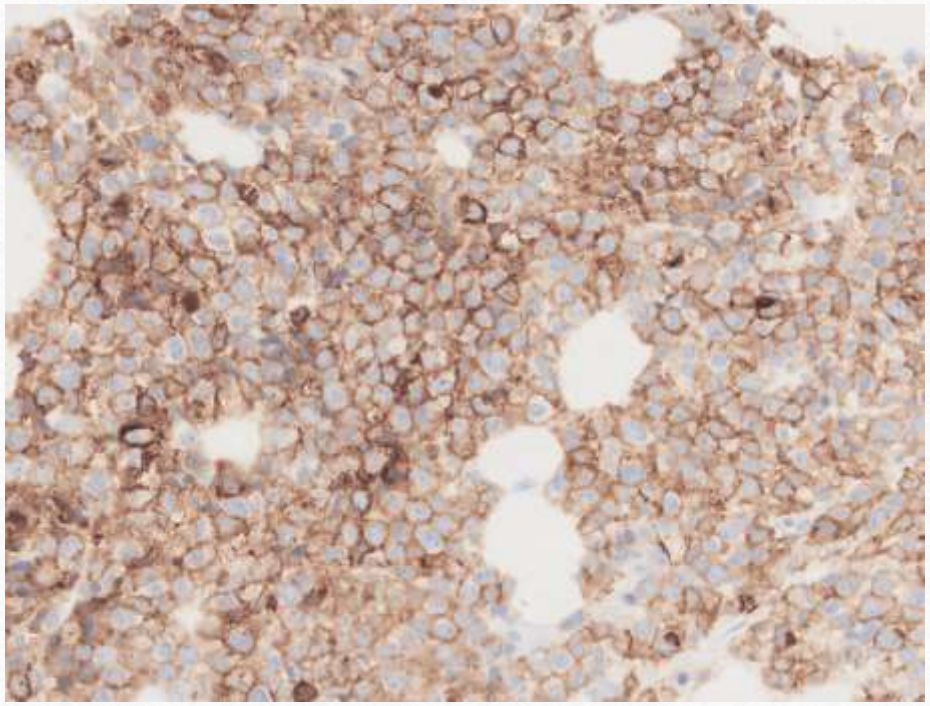
- Subsequently, bone marrow aspiration and biopsy was done which revealed cellular reactive marrow.
- No atypical cell or metastatic deposit.
- Next done was CT guided biopsy from left illiac bone lesion.

MICROSCOPIC FINDINGS



Interstitial infiltration by neoplastic round cells with convoluted nuclei. Brisk mitosis seen(arrow)

LCA: POSITIVE



- On IHC, tumor cells were positive for LCA, while negative for CK.
- So, we narrowed it down to hematolymphoid malignancy.

- Further, CD79a, PAX5, CD20, CD3, CD5, TdT were done. All were negative.

- B cell and T cell lymphoma including lymphoblastic lymphoma were largely rule out.

- So , now we left with the differentials of-

  ALK negative ALCL

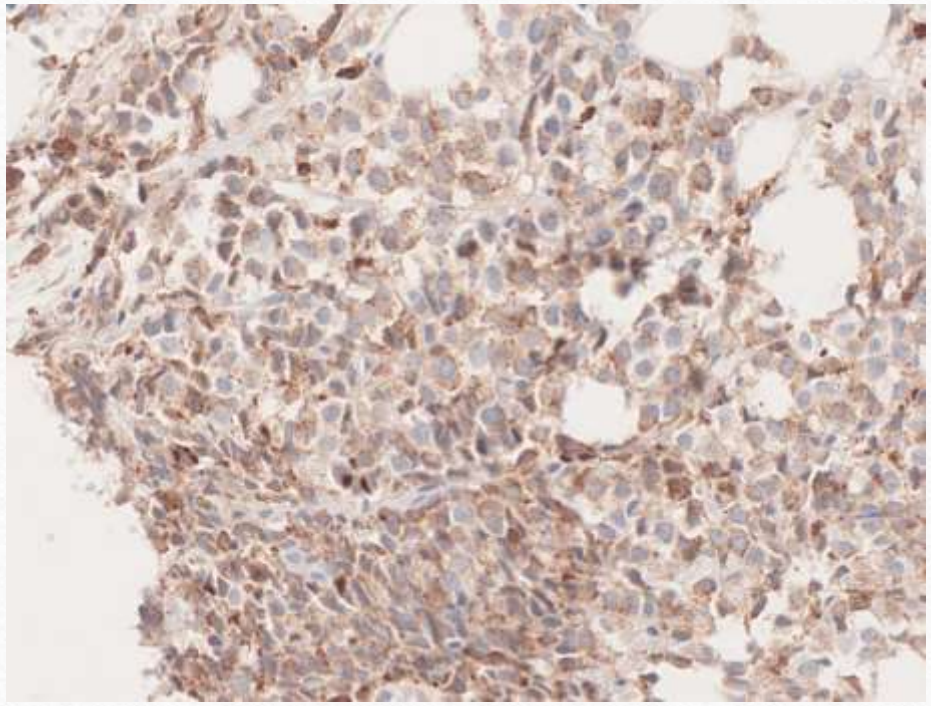
  Myeloid sarcoma

  Histiocytic sarcoma

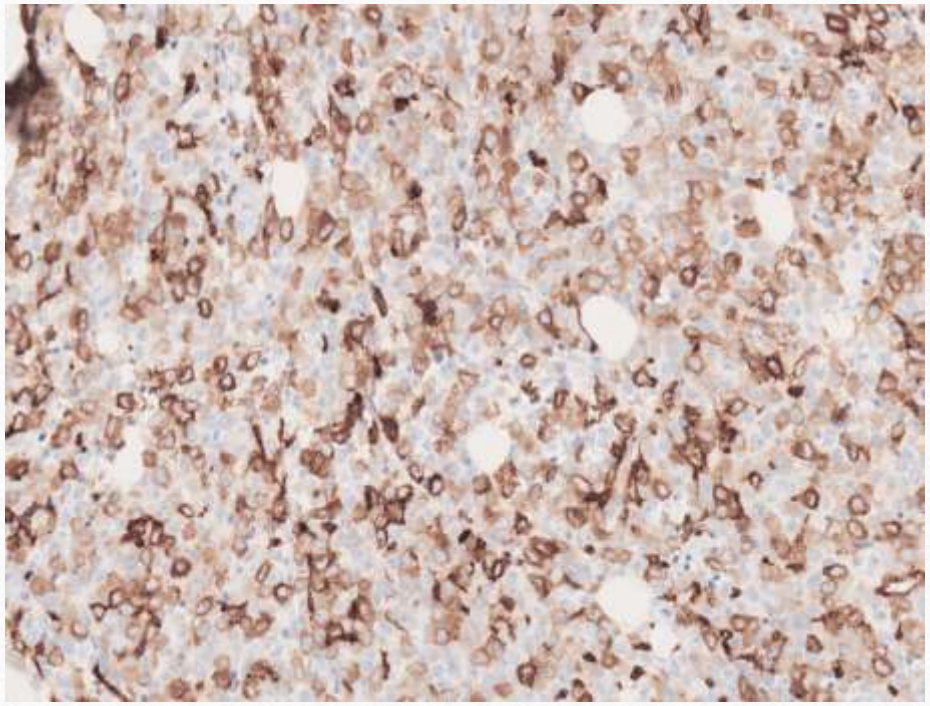
- Subsequently, ALK, CD30, EMA, c-kit, MPO, CD34 were done, and all were negative.

- Further, CD4, CD163, CD68 were done

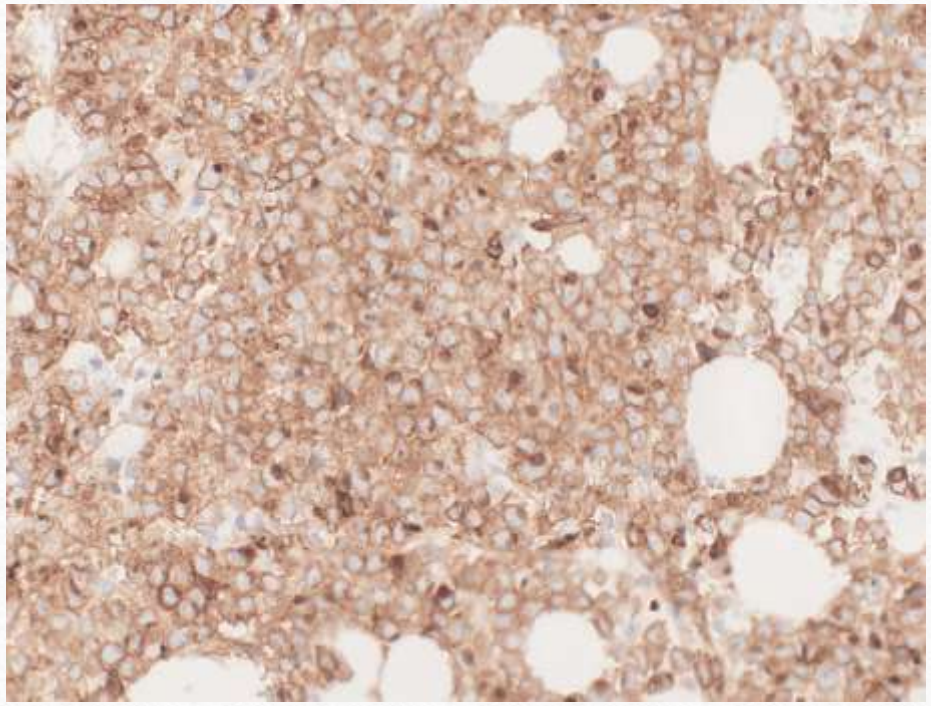
CD68: Strong positive



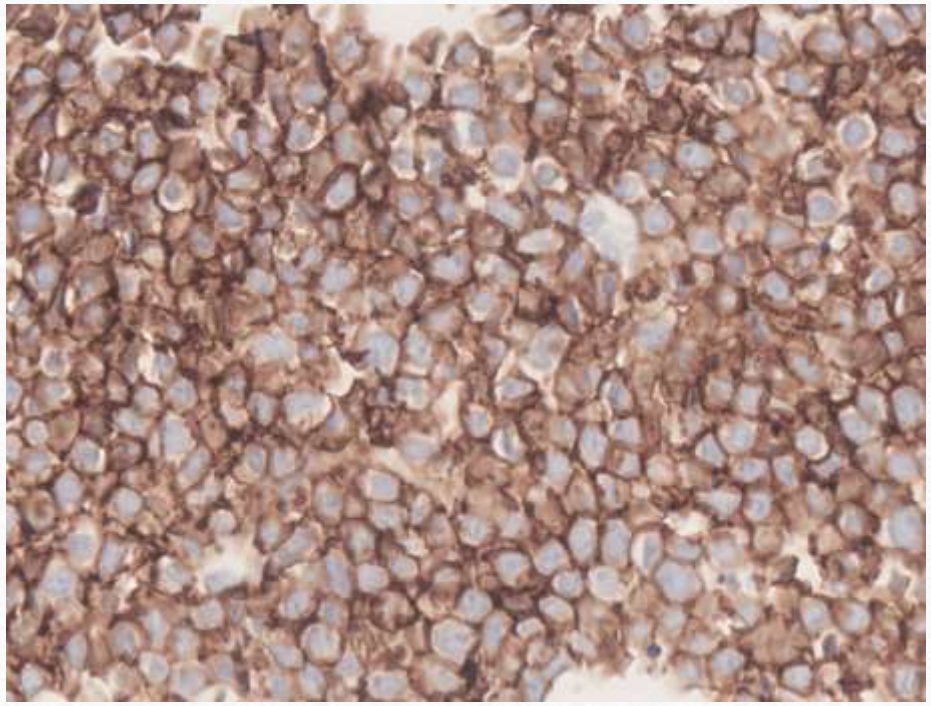
CD163: Strong positive



CD4: strong positive



CD43: Strong positive



FINAL DIAGNOSIS

- Case was discussed in multi speciality tumor board.
- In view of morphology and immuno-histchemistry, diagnosis of myeloid sarcoma (likely monocytic lineage) was suggested.

DISCUSSION

- Myeloid sarcoma, also known as, granulocytic sarcoma, chloroma, extramedullary myeloid tumor, is a tumor mass consisting of myeloid blasts, with or without maturation, occuring at anatomical sites other than the bone marrow.

- The tumor can involve any part of the body, but commonly involved sites include skin, lymph nodes, gastrointestinal tract, bone, soft tissue and testes.

- The clinical manifestations of MS are not specific and consist of local pain and mass effect.
- The patient had weakness of upper limb probably due to mass effect on the spinal cord.

- Histopathology has a central role in diagnosing MS, and this is further strengthened by the use of IHC.
- Diagnosis is comparatively easier when it arises in a setting of AML/MPS or MDS. However, in the absence of known hematological disorder, arriving at the diagnosis may be challenging.
- Routine histological examination of the tumors shows pleomorphic infiltrate of primitive cells of varying sizes and nuclear configuration with mononucleate and granulocytic cells of variable maturity along with scattered eosinophilic myelocytes.
- Eosinophilic myelocytes are a useful clue to the diagnosis; however, they may not always be present.

- MS has to be differentiated from other morphologic mimickers which include

1. Non-Hodgkin lymphoma (NHL) (lymphoblastic lymphoma, diffuse large B-cell lymphoma, Burkitt lymphoma)
2. Poorly differentiated carcinoma,
3. Plastic plasmacytoid dendritic cell neoplasm (BPDCN)
4. Small round blue cell tumors such as RMS and neuroblastoma.

- Tumors with more immature myeloid profile express CD33, CD34, CD68 and CD117. Staining for MPO, Tdt, and CD45 is inconsistent.
- Promyelocytic cases lack CD34 and Tdt but express MPO and CD15.
- Myelomonocytic tumors are positive for CD68, with MPO and CD68 confined to populations which are CD34 negative.
- The monoblastic variant express CD68 and CD163 but lack MPO and CD34, as evident in our case.

- Owing to their rarity, there is no well-documented optimal treatment for MS.
- However, the consensus opinion suggests cytarabine-containing remission inducing systemic chemotherapy for isolated MS.
- The correct diagnosis of MS is important for adequate therapy, which is often delayed because of a high misdiagnosis rate.

- MSs are very rare tumors with an aggressive clinical course.
- Morphologically, they can mimic a variety of tumors which fall broadly under the category of small round cell tumors.
- This problem is further complicated by the lack of clinical suspicion in patients with no known hematological disorder.
- The presence of scattered eosinophilic myelocytes in tissue biopsy specimens is a very useful clue to the diagnosis. However, accurate diagnosis requires appropriate use of IHC markers, MPO being the most frequently expressed marker. The possibility of MS should be considered when dealing with unusual lymphoma-like neoplasms that cannot be categorized as any of the NHL subtypes. A diligent search for eosinophil precursors can be rewarding. A delay in the diagnosis may result in unwarranted fatality particularly so in pediatric patients.

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THANK YOU