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Case Report

An unusual case of mediastinal mass with pleural and pericardial effusion

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ABSTRACT

Background: Myeloid sarcoma is a tumor mass consisting of myeloid blasts, with or without maturation, occurring in an anatomical site other than bone marrow. More than 2000 case reports so far, only few comprehensive studies have been done, which reflects the rarity and difficulties in treatment of this neoplasm.

Case Report: A 17-year-old female presented with complaints of neck swelling since 2 months, breathlessness since 5 days. PET CT- Bilateral pleural and pericardial effusion Large mediastinal mass, multiple enlarged lymph nodes and appendicular skeleton showing increased FDG uptake. CT-Guided biopsy of the mediastinal mass: Uniform blue cells in sheets.

IHC: CD45-Weak positive CD20, CD3, TdT-Negative CD99-Diffuse strong positive Parallelly, blood and bone marrow examination was done. Peripheral smear-80% blasts. Bone marrow-Monomorphous population of myeloblasts.

Discussion: About 21% of Myeloid Sarcomas (MS) are reported to occur in the mediastinum. Clinical presentation is dependent on tumour location, with symptoms due to tumour mass effect or local organ dysfunction. Recent studies show a misdiagnosis of 25-47%, with Hodgkin's lymphoma, lymphoblastic lymphoma, DLBCL, Ewing's sarcoma, thymoma, round blue cell tumours, or poorly differentiated carcinomas, mostly due to inadequate immunophenotyping. It was not corrected until a diagnosis of acute leukaemia was later established by bone marrow biopsy or peripheral blood examination.

Conclusion: Recognition of MS with/without AML is essential for prognosis. Correlating radiological, hematological, bone marrow and flowcytometry features with histomorphology and immunohistochemistry of the tumor is essential. A rare possibility of MS should be kept in mind for mediastinal masses for timely diagnosis and treatment.

1. Introduction

Myeloid sarcoma is a tumor mass consisting of myeloid blasts, with or without maturation, occurring in an anatomical site other than bone marrow.[1]

Myeloid Sarcoma (MS) is only called so if there is a mass formation with complete effacement of tissue architecture.

More than 2000 cases have been reported so far, with about 21% cases occurring in the mediastinum.[2]

Clinical features depend on the location, with symptoms due to tumor mass effect or local organ dysfunction. It may sometimes precede/ coincide with AML or constitute blastic transformation of MDS/ MPN.[1]

2. Case Report

A 17-year-old female came with the complaints of swelling in the neck since 1 month, breathlessness for the past 5 days.

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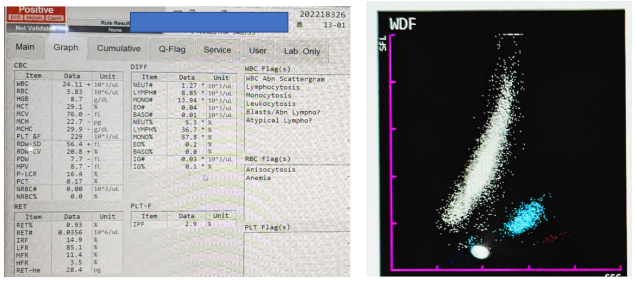

Fig. 1: CBC

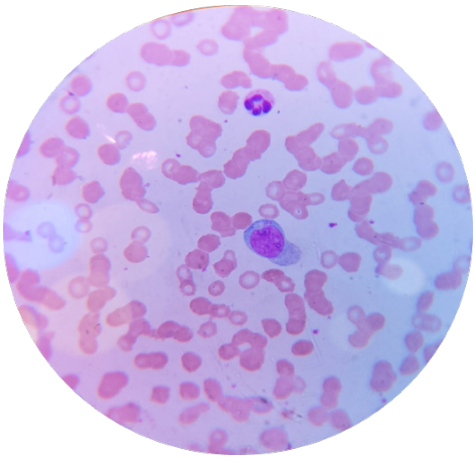

Fig. 2: Peripheral smear: Abnormal cells: 80% blasts; Impression: Acute leukemia

2.1. *Pet-CT*

Metabolically active disease in the large soft tissue mass occupying the entire mediastinum, encasing the mediastinal vessels, trachea and esophagus.

Metabolically active disease in the large ill-defined soft tissue mass in bilateral supraclavicular region.

Mild pleural effusion and pericardial effusion.

Metabolic activity in the cardiophrenic nodes.

Diffuse hypermetabolism in the marrow of all bones of axial and proximal appendicular skeleton.

2.2. *Pericardiocentesis*

Appearance: Turbid

Color: Red

Coagulum: Present

Cell count: 74,000 cells/cumm

Cell type: Mononuclear atypical cells: 90%

Neutrophils: 10%

Protein: 6.8 g/dL

Glucose: 10 mg/dL

Myelopoiesis: Increased in number, shows predominant population of blasts (86%), having high N:C ratio, fine chromatin, 1-2 nucleoli, scanty cytoplasm. Few blasts are seen to have nuclear convolutions.

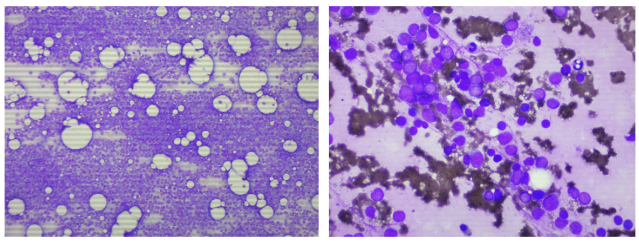

Fig. 3: Bone marrow aspiration; Posterior superior iliac spine

Erythropoiesis:
Megakaryopoiesis: } Markedly diminished

Impression: Acute Leukemia, immunophenotyping indicated for further subtyping.

2.3. *Flowcytometry*

1. Low side scatter in CD45 and CD13
2. Moderate expression of CD34, CD33, CD117, HLA-DR, CD7. Rest of the markers were negative.

2.4. *CT-guided biopsy of the mediastinal mass*

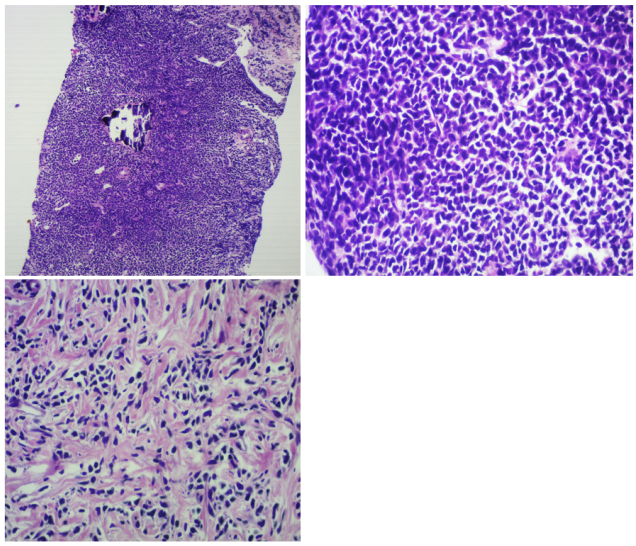

Fig. 4: Tumor cells arranged in sheets, composed of monomorphous population of large to medium sized cells having high N:C ratio, hyperchromatic nuclei, scant cytoplasm. Microcalcification seen. Adjacent areas show fibrous tissue infiltrated by tumor cells

3. Discussion

Myeloid sarcoma is a rare neoplasm with a greater chance of it being misdiagnosed as malignant lymphoproliferative

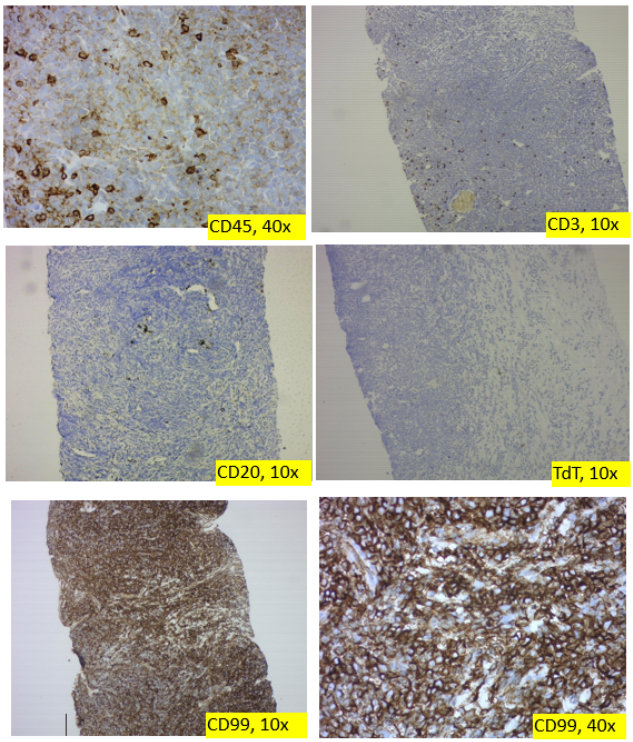

Fig. 5: Immunohistochemistry

disorders including Hodgkin's lymphoma, MALT lymphoma, DLBCL, Ewing's sarcoma, thymoma, round blue cell tumors or poorly differentiated carcinoma.[3] The misdiagnosis was commonly due to inadequate immunophenotyping of the MS lesion and was not corrected until a diagnosis of acute leukemia was later established by bone marrow biopsy or peripheral blood examination.[4]

In a study by J E Takasugi et.al, mediastinum was the commonest intrathoracic site for MS and 6 out of 9 cases presented with pleural effusion.[5]

Very few cases of MS with pleural and pericardial effusion have been reported.

4. Conclusion

Recognition of MS with or without AML is necessary for prognosis.

Correlating radiological, hematological, bone marrow and flowcytometry features with histomorphology and immunohistochemistry of the tumor is essential.

A rare possibility of MS should be kept in mind for mediastinal masses for timely diagnosis and treatment.

5. Source of Funding

None.

6. Conflict of Interest

None.

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