Case Report

Histiocytic Sarcoma: An Unusual Presentation of a Rare Entity

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Abstract

Histiocytic sarcoma (HS) is a rare non-Langerhans histiocytic disorder accounting for less than 1% of all hematolymphoid disorders, with an aggressive clinical course with limited treatment options. It has a male predominance with extranodal sites as the commonest site of involvement although it can affect nodal sites, can also arise de novo from these sites or certain hematolymphoid disorders such as B-cell lymphomas, chronic lymphocytic leukemia/small lymphocytic lymphoma.

Keywords: Histiocytic sarcoma, hematolymphoid, mesenteric lymph node

INTRODUCTION

Histiocytic sarcoma is a rare and aggressive non-Langerhans histiocytic disorder. It is classified under tumours of histiocytic/dendritic cell neoplasm under World Health Organisation (WHO, 2022) Classification of Haematoymphoid tumours. [1] Although any age can be affected, it predominantly affects adulthood, with site of involvement been either nodal or extranodal.[2]

Histiocytic sarcoma can occur as a secondary event following some hematolymphoid neoplasms such as acute lymphoblastic lymphoma/chronic lymphocytic leukemia.[3] Follicular lymphoma, Diffuse large B cell lymphoma, hairy cell leukemia, mantle cell and Malt type lymphoma, chronic myelomonocytic leukemia and mediastinal germ cell tumours.[4-6] Diagnosis of this tumour is challenging because of its rarity and its close resemblance to other histiocytic/dendritic cell neoplasms, carcinomas and metastasis.[7]

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We report a case of HS sarcoma involving the mesenteric lymph nodes in a 22-year-female patient who presented with abdominal pain and distension.

CASE PRESENTATION

A 22-year-old woman presented with 3 weeks history of fever and two months history of

abdominal pain and distension. The pain was colicky, extending from the right lumbar to the epigastric region. No peripheral lymphadenopathy and her past medical history were unremarkable. An Ultrasound scan revealed multiple mesenteric lymph nodes measuring 4x4cm. All the other base line investigations including full blood count, electrolyte urea and creatinine were within the normal reference limit. A diagnosis of non-Hodgkin lymphoma was made by the clinician and a biopsy of the mesenteric lymph node was taken to the Pathology Department for histology.

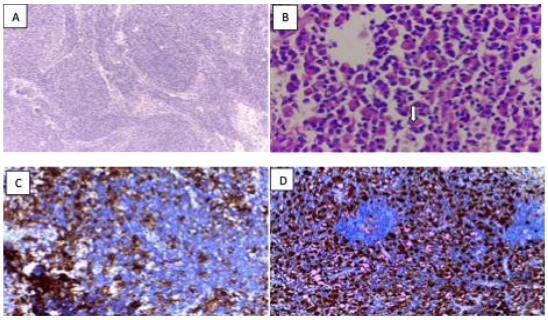

Plate 1- A- Photomicrograph of H&E stain slide showing partial effacement of nodal architecture by proliferating neoplastic cells. x 40

B-Photomicrograph of H&E stain slide showing pleomorphic round to oval cells with epitheloid features and hemophagocytosis (white arrow).x200

C-Photomicrograph of an ABC-stained slide with CD163 antibody showing strong and diffuse membranous and cytoplasmic staining.x400

D-Photomicrograph of an ABC-stained slide with CD 68 antibody showing strong diffuse cytoplasmic staining.x400

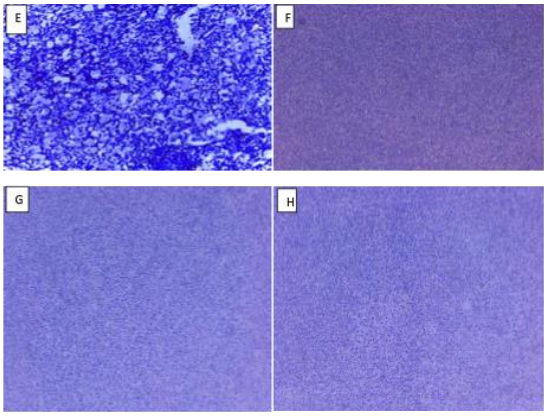

Plate 2- E- Photomicrograph of ABC-stained slide with CD1a antibody showing negative stain.x400

F- Photomicrograph of ABC-stained slide with CD5 antibody showing negative stain.x40

G- Photomicrograph of ABC-stained slide with CD23 antibody showing negative stain.x40

H- Photomicrograph of ABC-stained slide with CD 15 antibody showing negative stain.x40

On histopathological examination of the lymph node, there was total effacement of nodal architecture by diffuse sheets of round to oval cells with prominent nucleoli and abundant eosinophilic cytoplasm with some of the cells displaying epitheloid morphology (Plate 1A&1B).

Haemophagocytosis and necrotic areas were also seen. Immunohistochemistry showed CD163 and CD68 strong membranous and cytoplasmic immunopositivity in the neoplastic cells (Plates 1C&D) while, CD 5, CD15, CD 30, CD 23, S-100 and Pan-cytokeratin were all negative (Plate 2E-H). A diagnosis of HS was made and patient was placed on a chemotherapy regimen of cyclophosphamide, doxorubicin, vincristine and prednisolone (CHOP). Was discharged after a remarkable improvement and was lost to follow- up.

DISCUSSION

Histiocytic sarcoma is an uncommon non-Langerhans histiocytic disorder classified under tumours of histiocytic/dendritic cell neoplasm under World Health Organisation (WHO, 2022) Classification of Haematoymphoid tumours with an aggressive clinical course and can occur in association with other hematological malignancies (Secondary HS). The etiology of HS is unknown and there is no standardized treatment. It is mainly seen in adults as in this index patient with male predominance, although pediatric cases have been reported. [8]

Most cases present as a solitary mass at extranodal sites, which is not the case in our patient that presented with multiple mesenteric masses. Systemic (fever, fatigue, weight loss) (B) symptoms can be present in some patients as in this index case.[9]

Histiocytic sarcoma usually shows a diffuse architecture with neoplastic cells been large and oval with abundant eosinophilic cytoplasm and well-defined cell borders. The nuclei can be vesicular with prominent nucleoli while the cytoplasm can have epitheloid morphology as seen in this patient. Emperipolesis or Haemophagocytosis by neoplastic cells can be seen with variable number of necrotic areas (Figures 1).

Immunophenotyping is very necessary in establishing the diagnosis of HS because it has a wide range of differential diagnosis. HS is mostly positive for CD68, CD163, CD45, lysozyme and negative for CD1a, S-100 and CD30. Positive expression of CD163, CD68 and negative expression of CD1a, CD5, CD15, CD30, CD23, S-100 and Pan- cytokeratin allowed us to rule out most B and T lymphomas, Langerhans cell histiocytosis, Rosai Dorfmans disease, Metastatic carcinoma and melanoma in this index patient. CD163 which is very specific for identifying machrophages/histiocytes as well as non-neoplastic monocytes is necessary for the diagnosis of HS.[10]

Molecular studies which were not done in this patient have also shown frequent mutations in RAS/MAPK pathway suggesting the possibility of targeted therapy in the future for patients with this disorder.[11]

CONCLUSION

Histiocytic sarcoma presenting as solitary nodal masses is very rare with challenges in excluding a wide range of differential diagnosis.
However, the careful use of immunohistochemistry can help reduce these differentials thereby establishing the correct diagnosis.

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