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Case Report:

Emperipolesis in a Case of Adult T Cell Lymphoblastic Lymphoma (Mediastinal type)-Detected at FNAC and Imprint Cytology

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Abstract: Emperipolesis is a condition in which viable hematopoetic cells are seen intact in the cytoplasm of host cell without damage. This phenomenon is seen in many physiologic and pathologic conditions, its presence in Rosai Dorfman disease (RDD) is characteristic of the disease. However emperipolesis is an uncommon finding in malignant lymphoma both Hodgkins and non-Hodgkin's lymphoma, wherein it has been described in bone marrow aspirate and tissue culture. In contrast there are only two case reports of emperipolesis phenomenon described in non-Hodgkin's lymphoma in tissue sections. We report a case of an adult T cell lymphoblastic lymphoma (mediastinal type) with features of emperipolesis demonstrated at fine needle aspiration cytology (FNAC) and imprint cytology of cervical lymph nodes. To our knowledge, this is the first case report of emperipolesis in a case of adult T cell lymphoblastic lymphoma (mediastinal type)-detected at FNAC and imprint cytology.

Key Words: Emperipolesis; Lymphoblastic lymphoma; Imprint cytology; Rosai Dorfman disease; Fine needle aspiration cytologyy

Introduction:

Emperipolesis is defined as the presence of intact hematopoetic cells like neutrophil, lymphocyte or plasma cell in the cytoplasm of host cell. This phenomenon is characteristic of Rosai Dorfmann disease (RDD).[1] However, it can be seen in association with other physiologic and pathologic conditions. Pathologic conditions include several benign and malignant disorders like autoimmune haemolytic anemia, idiopathic thrombocytic purpura, carcinoma, neuroblastoma, multiple myeloma, giant cell carcinoma of lung, leukaemia and malignant lymphoma.[2]

Finding of emperipolesis in malignant lymphoma has been described in bone marrow aspiration and tissue culture studies.[1] After extensive search of literature we came across only two cases reports of emporipolesis in Non-Hodgkin lymphoma as described in tissue sections.[2and3] Till date, there is no case report of emperipolesis in a case of adult T cell lymphoblastic lymphoma (mediastinal type)-detected at FNAC and imprint cytology.

RDD and lymphoma show considerable overlap of clinical picture. However, both the disorders have a contrasting management protocol and outcome. As a result it is of utmost important to be aware of the fact that emperipolesis does occur in a lymphoma.

Hence we report herein a case of 20 year old male presenting with bilateral massive cervical lymphadenopathy. At FNAC of cervical lymph node and imprint cytology of the biopsy from same site, extensive emperipolesis in a background of a T cell lymphoblastic lymphoma was noted. Findings were corroborated at biopsy and immunohistochemistry.

Case Report:

A 20 years old young male presented with history of swelling in both sides of the neck of one month duration. He also complained of anorexia and loss of weight since one month. On examination, he had bilateral massive cervical lymphadenopathy. Lymph nodes were firm and matted. Bilateral axillary lymph nodes were palpable. There was no evidence of hepatosplenomegaly. Laboratory investigations were within normal limits.

Fine Needle Aspiration Cytology (FNAC)

FNAC was initially done elsewhere and was reported as non-specific reactive lymphadenitis. We did FNAC from cervical lymph node in our laboratory. Smears showed mixed population of lymphoid cells comprising of mature lymphocytes, few plasma cells, eosinophils and histiocytes. Also seen were some atypical intermediate sized lymphoid cells with scant cytoplasm, high nucleocytoplasmic ratio, fine chromatin and inconspicuous nucleoli. Histiocytes showed emperipolesis.

The engulfed cells were surrounded by clear halo (Figure 1). Some of the histiocytes showed vacuolations in the cytoplasm (Figure 2). In view of clinical findings and presence of emperipolesis a possibility of RDD was considered. However the presence of atypical cells prompted us to the diagnosis of a lymphoproliferative disorder.

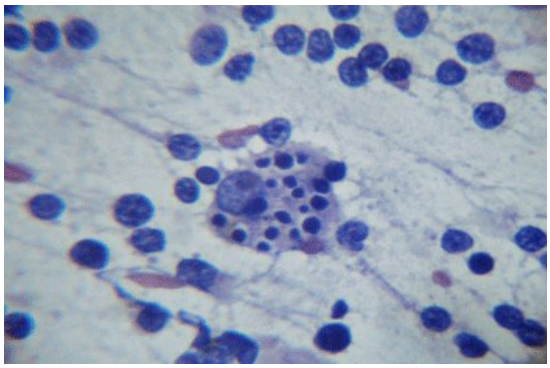

Figure 1: Engulfed cells surrounded by clear halo

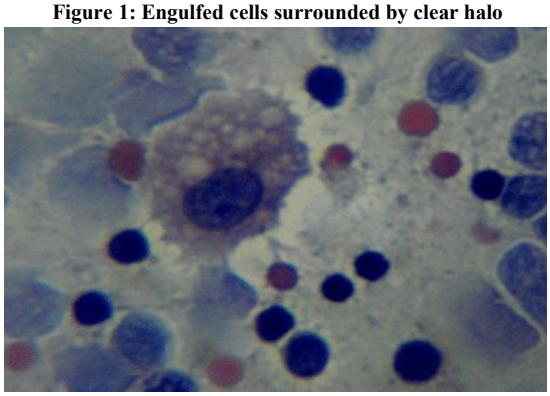

Figure 2: Histiocytes showing vacuolations in the cytoplasm

X ray chest was done which showed a mediastinal widening. CT scan confirmed the mediastinal mass as lymph nodes.

Biopsy of cervical lymph node was done.

Imprint Cytology

Imprint smears were made, fixed in ethanol and stained with haematoxylin and eosin, Giemsa and Papanicolaou stain. Smears revealed the same pattern as in FNAC, i.e., atypical lymphoid cells and emperpolesis phenomenon.

Histopathology

Histopathology showed complete effacement of lymph node architecture by monotonous population of intermediate sized lymphoma cells with scant cytoplasm, high nucleocytoplasmic ratio, fine chromatin and inconspicuous nucleoli. Crush artefact was noted extensively. However we could not demonstrate emperipolesis inspite of complete sampling of the tissue.

Immunohistochemistry

Immunohistochemistry showed positivity for T cell markers, i.e., CD3, CD 7and CD 43. B cell markers –CD 19 and CD20 were negative.

Hence a final diagnosis of adult T lymphoblastic lymphoma – mediastinal type was made.

Discussion:

Emperipolesis is a condition in which viable haematopoietic cells are seen in the cytoplasm of host cells. These host cells may be megakaryocyte, monocyte, endothelial cells, fibroblast or malignant cells.[2-6] First described by Humble et al in 1956, emperipolesis is a characteristic finding in RDD.[4] However it can be seen in other physiological and pathological conditions. Erythroblast emperipolesis by megakaryocyte in fetal liver is one such physiologic condition.[7] Pathologic disorders associated with emperipolesis include benign and malignant disorders like autoimmune haemolytic anemias, myelosclerosis, myeloproliferative disorders, idiopathic thrombocytopenic pupura, neuroblastoma, multiple myeloma, leukaemia and malignant lymphoma.[2]

In emperipolesis, the haematopoietic cell is enclosed in a membrane bound vacuole in the cytoplasm of host cell.[5] The only difference between emperipolesis and phagocytosis is the absence of destruction of the engulfed cell and host cell in the former.[1]

Association of emperipolesis and malignant lymphoma is rare, with very few case reports in literature. Emperipolesis has been noted more commonly in non-Hodgkin's lymphoma than in Hodgkin lymphoma. In most of these cases emperipolesis was demonstrated at bone marrow aspirate or tissue culture studies.[2,3,5,8,9] Dzieciol J et al observed megakaryocytic emperipolesis in six out of 30 cases of non-Hodgkin's lymphoma in the bone marrow aspiration.[10] There are only two case reports of non-Hodgkin's lymphoma in which emperipolesis was demonstrated in tissue sections. In one case it was a diffuse large B cell lymphoma, in another case the subtype was not specified.[1,3]

There is only one case of emperipolesis being detected as a key feature in imprint cytology of thymus. In this case, thymus was removed mistakenly at thyroid surgery. Imprint revealed emperipolesis of thymocytes in epithelial cells, the significance of which was not known. Emperipolesis was not detected at histopathology.[11]

Aetiology of emperipolesis phenomenon occurring in a lymphoma is eluding, however, some authors suggest the role of cytokines liberated by lymphoma cells in its occurrence. Some investigators believe that emperipolesis is the result of active adherence of lymphocytes to tumor cells or macrophages with a subsequent inclusion in vacuoles inside the cytoplasm of these cells.[4,5] However further studies are required to resolve the issue.

Presence of emperipolesis in a lymph node with mixed inflammatory infiltrate prompts the cytopathologist to think in terms of RDD. RDD is a benign proliferative disorder of histiocytes. It has an indolent clinical course with most of the patient undergoing complete resolution without any treatment.

RDD shares common clinical scenario of young age, fever, non-tender massive lymphadenopathy and weight loss with adult T cell lymphoblastic lymphoma.

Lack of awareness of occurrence of emperipolesis phenomenon in a lymphoma leads to miss diagnosis and delay in management and treatment.

A careful attention to the accompanying cells is a key for differentiating a benign condition from an aggressive lymphoma at cytology.

This is the first case report of emperipolesis being documented at FNAC and imprint cytology in a case of adult T cell lymphoblastic lymphoma.

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