ejpmr, 2021,8(11), 9-12.

SJIF Impact Factor 6.222

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Study

ISSN 2394-3211

EJPMR

LYMPH NODE HISTIOCYTIC SARCOMA; A RARE DISEASE, ABOUT A CASE

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Article Received on 25/08/2021 **Article Revised on 15/09/2021** **Article Accepted on 05/10/2021**

ABSTRACT

Histiocytic Sarcoma (HS) presents as an extremely rare malignant tumor, which arises from the proliferation of phagocytic cells and histiocytes. However, the literature reports a few hundred cases. HS is diagnosed based on morphology and immunohistochemistry. It often presents at an advanced clinical stage, with limited response to chemotherapy and a high death rate. In this work we report the observation of a 33-year-old patient, occasional smoker, who presented with bilateral cervical lymphadenopathy. On clinical examination, there was an enormous left laterocervical lymph node magma; with left fluid effusion syndrome on pleuropulmonary examination. The anatomo-pathological study completed by immunolabeling was compatible with lymph node histiocytic sarcoma. The patient received COP-type palliative chemotherapy (cyclophosphamide, vincristine and prednisolone). There is no standard treatment for HS due to its rarity, and it usually has primary resistance to chemotherapy or relapses soon after treatment. It is important to establisha consensus of management of HS for improving the prognosis of this rare disease.

KEYWORDS: Histiocytic sarcoma, cervical lymphadenopathy, case report.

INTRODUCTION

Histiocytic Sarcoma (HS) presents as an extremely rare malignant tumor, which arises from the proliferation of phagocytic cells and histiocytes.

However, the literature reports a few hundred cases. HS is diagnosed based on morphology and immunohistochemistry.

It often presents at an advanced clinical stage, with limited response to chemotherapy and a high death rate.[1,2,3,4]

Observation

In this work, we report the observation of a 33-year-old patient, occasional smoker, who presented for 5 years with left cervical lymphadenopathy discovered incidentally on self-examination, gradually increasing in size. The course was marked 2 years rather by the appearance of another contralateral lymphadenopathy.

On clinical examination, we noted the presence of a huge left laterocervical lymph node magma "Fig.1" with centimetric occipital lymphadenopathy and left supraclavicular lymphadenopathy of about 50mm in diameter and several bilateral axillary lymphadenopathies of variable diameter; with left fluid effusion syndrome on pleuropulmonary examination.

Fig. 1: photos of left lateral cervical lymph node magma.

The cervical computed tomography (CT) examination showed the presence of a large left lateral cervical lymph node mass measuring 172 * 67mm in diameter, with a left supraclavicular lymphadenopathy of 48mm and axillary lymphadenopathy, the largest of which measures 36mm on the left and 4mm to the right.

The left cervical mass was biopsied with the pathological study: undifferentiated malignant tumor infiltrating the brought back fibrous tissue, without vascular emboli "Fig. 2".

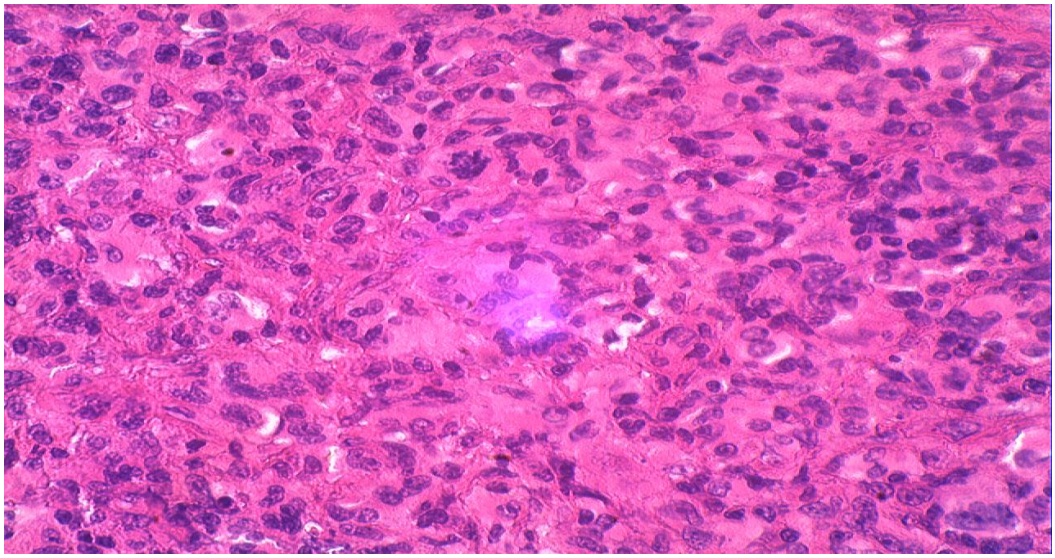

Fig. 2: (HE*200) tumour proliferation pleomorphic cells with moderately atypical, irregular and hyperchromatic nuclei with abnormal mitosis.

On immunohistochemistry (paraffin and freezing), cells were positive for anti-CD68 "Fig. 3", and anti-CD45 antibodies. The tumor expressed heterogeneously the PS100 antibodies. The Ki 67 was at 50%. Labeling was negative for EMA, PAX 5, CD1a, CD34, anti-Myeloperoxidase and anti-pancytokeratin: which was consistent with histiocytic sarcoma.

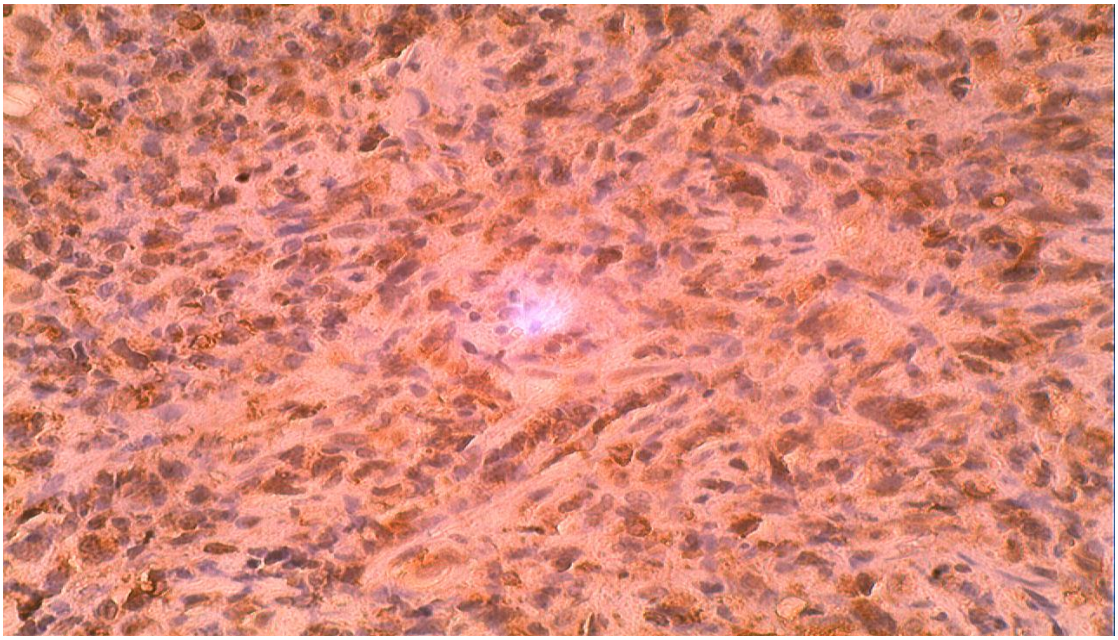

Fig. 3: Diffuse positivity of tumor cells for anti-CD68 antibodies (histocytes).

Thoraco-abdominal-pelvic CT scan performed as part of the extension workup demonstrated the presence of a large partitioned left pleural effusion "Fig. 4".

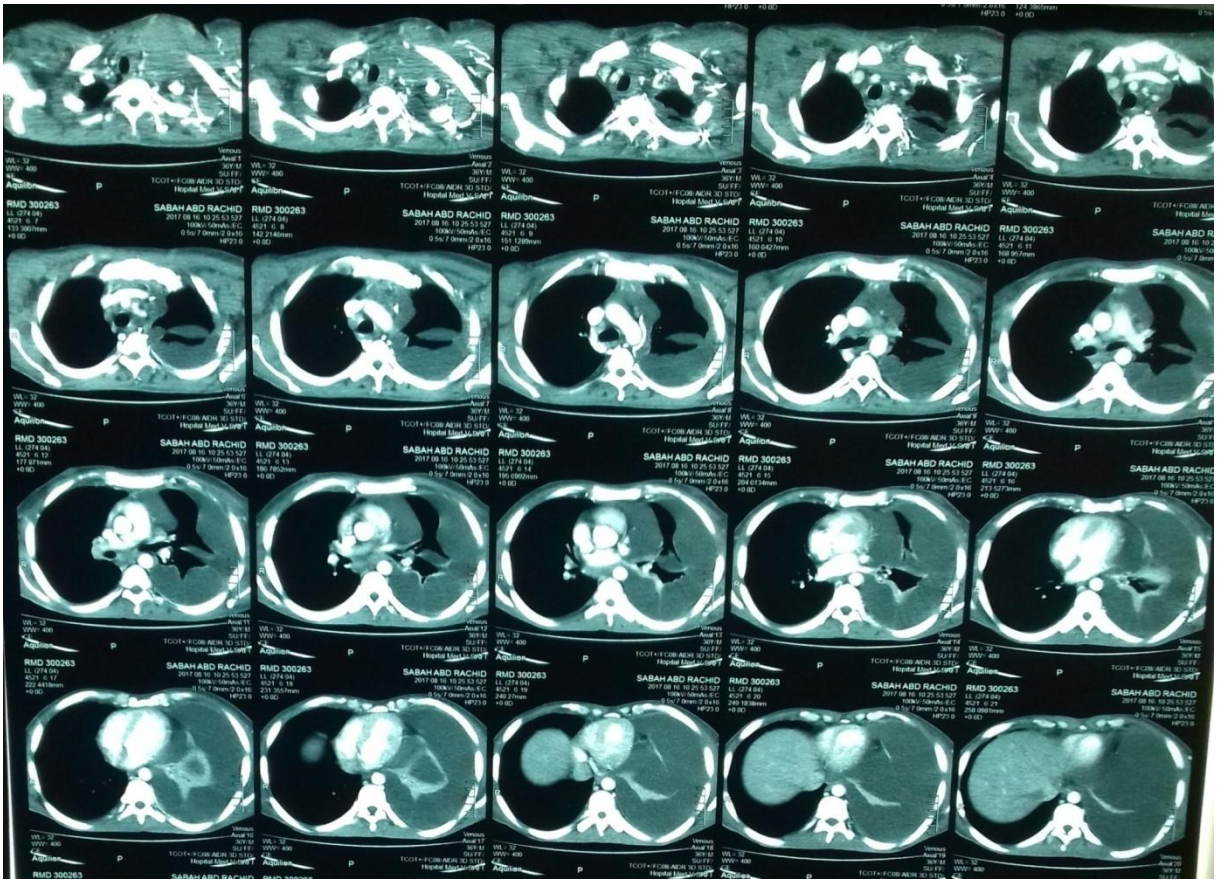

Fig. 4: left lateral cervical lymph node mass with left susclavicular lymphadenopathy and axillary lymph nodes with left pleural effusion partitioned of great abundance.

The pre-treatment work-up included a cardiac ultrasound, and a standard laboratory work-up was found to be normal. The patient received COP-type palliative chemotherapy (cyclophosphamide, vincristine and prednisolone).

The patient on day 8 of the protocol had presented respiratory distress following which he died.

DISCUSSION

HS is a hematopoietic tumor of extreme rarity and aggressiveness. Epidemiologically speaking, it represents less than 1% of all hematolymphoid cancers, although its rarity suggests its true incidence. It can affect all age groups, from infants to the elderly (6 months to 89 years, the median age being 46 years), with a male predominance (sex ratio of 3/1).[2,5,6]

Analysis of the literature reveals few cases of histiocytic sarcoma which can be localized or disseminated; most of them being of lymph node location, although various extra-nodal sites can be affected (including the gastrointestinal tract, spleen, soft tissues and skin ...).[2]

Clinically, systemic symptoms such as fever, fatigue, night sweats, weight loss and asthenia are relatively common. Lymphadenopathy is also often seen. Skin manifestations, intestinal obstruction, hepatosplenomegaly with associated pancytopenia, and lytic bone lesions may also occur.[7]

HS is diagnosed by histopathological examination, including morphology and immunohistochemical studies.[3]

Histologically, the tumor presents as a diffuse infiltration of large, round to ovoid pleomorphic cells. During the immunohistochemical study, tumor cells express several histiocytic markers, including CD163, CD68 (KP1 and PGM1) and the detection of lysozyme is an important element underlining their membership in the histiocyte lineage. With typical absence of B cell, T cell, Langerhans cell (CD1a, langerin / CD207), follicular dendritic cells (CD21, CD23, CD35, CAN.42), epithelial (pancytokeratin, EMA), melanocytic cells (HMB) -45, Melan A) and myeloid cells (CD13, CD33, myeloperoxidase). Tumor cells are negative for CD30. Some cases may be positive for the S-100 protein, but the staining is often weak and patchy rather than uniform. The Ki-67 index is variable.[4.2]

However, it should be emphasized that none of the antibodies are specific for histiocytic differentiation. Therefore, study with a panel of antibodies in the context of morphology is paramount. There is no standard treatment for HS due to its rarity.[3,4]

Patients with advanced nodal or extranodal histiocytic sarcoma usually receive multidrug chemotherapy used for non-Hodgkin lymphoma of high malignancy, with a second line of multidrug therapy if initial treatment fails. However, there are no prospective trials of the best protocol, and treatment data are limited to small case series. Commonly used treatment regimens are ICE (Ifosfamide, Carboplatin, Etoposide with mesna) as well as CHOP (Cyclophosphamide, Hydroxyadriamycin, Vincristine, Prednisone). HS usually has primary resistance to chemotherapy, or relapses soon after treatment.[3,4]

CONCLUSION

This observation confirms the need to establish a consensus of diagnosis and management of HS in order to prevent diagnostic and therapeutic delays and improve the prognosis of this rare disease.

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