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Case Study

Follicular Dendritic Cell Sarcoma About A Case With Literature Review

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ABSTRACT

Follicular dendritic cell sarcomas (FDSC) are classified as histiocytic and dendritic cell neoplasms in the WHO 2017 classification of hematopoietic and lymphoid tissue tumors [1]. It is an extremely rare malignant tumor with morphological and phenotypic characteristics of follicular dendritic cells (FDC) [2]. These are derived from the mesenchymal stromal cells of the lymphoid follicles. They are an indispensable link between innate and adaptive immune responses.

In this study, we raise through two observations the positive and differential diagnostic difficulties of this entity and we recall its morphological, immunohistochemical and evolutionary characteristics. Only 400 cases of FDSC are described in the literature. Our observation raises the problems of differential diagnosis frequently posed by follicular dendritic cell sarcoma. The main differential diagnosis is with metastasis of undifferentiated carcinoma. The positivity of the dendritic follicular markers (CXCL13, CD21, CD35, FDCSP and SRGN) makes it possible to affirm the diagnosis. Surgery is the treatment of reference. Dendritic follicular cell sarcomas are a nosological entity that is still under debate. Given their diagnostic difficulties, the demand for histological proof reading within the frame work of a specialized network is of great interest.

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INTRODUCTION

Follicular dendritic cell sarcomas (FDCS) are classified as histiocytic and dendritic cell neoplasms in the WHO 2017 classification of hematopoietic and lymphoid tissue tumors [1]. It is an extremely rare malignant tumor with morphological and phenotypic characteristics of follicular dendritic cells (FDC) [2]. These are derived from the mesenchymal stromal cells of the lymphoid follicles. They are an indispensable link between innate and adaptive immune responses [3]. The histogenesis of this disease is still ambiguous. Only 400 cases of reported FDCS are described in the literature [4], probably due to an under-diagnosis of this sarcoma.

Objective: to raise the difficulty of the diagnosis of this entity and to recall its morphological,

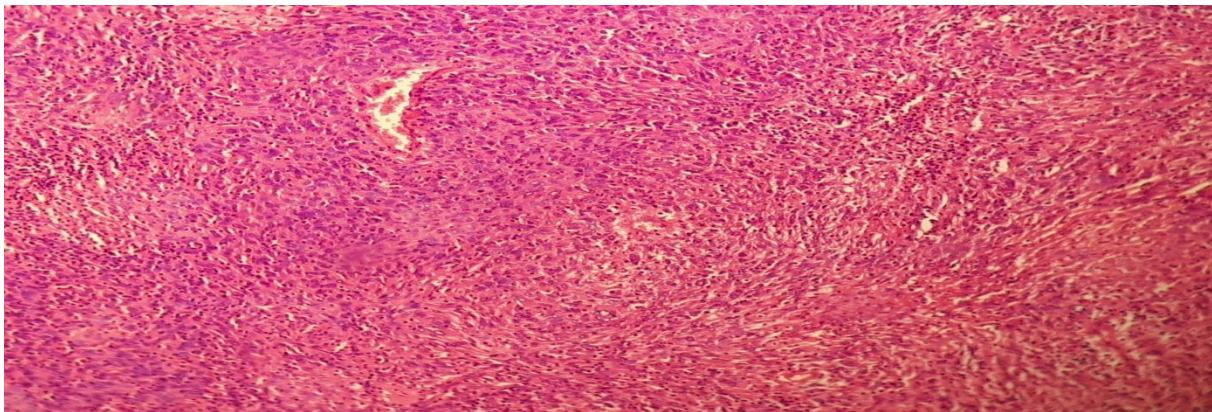
immunohistochemical and evolutionary characteristics.

OBSERVATION:

This is a 54-year-old patient with right lateral cervical polyadenopathy and sterno-cleido-mastoid mobile muscle with a hard consistency and no signs of ulceration, which has been evolving for 2 years, without fever or weight loss. His medical history was unspecific. No other lesions were found in his examination of the ENT sphere.

A cervical CT scan: a voluminous tissue mass of 20x8cm lateral right cervical, infiltrating deep spaces, compressive with probable thrombosis of the internal jugular vein.

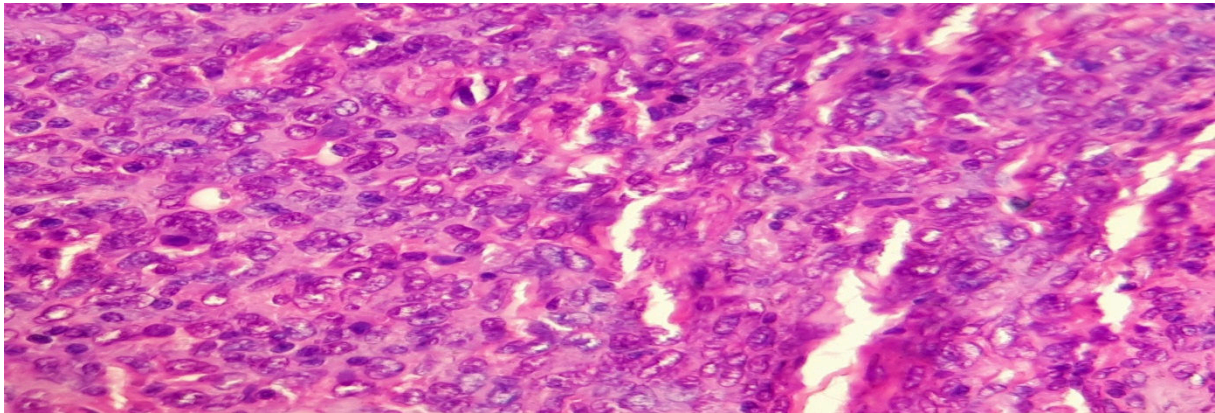
Figure1: FDCS, Follicular dendritic cell sarcoma: storiform appearance pattern. H&E.



A biopsy of the cervical lymphadenopathy concluded an undifferentiated malignant tumor infiltrating the fibro-muscular tissue. A review was performed in our training. Histological examination shows striated fibrous and muscle tissue dissociated by malignant tumor proliferation. This is arranged in fascicular, storiform, whorled and diffuse pattern. The cells are joined. The nuclei are hyperchromic multinucleate in places with abnormal mitoses. The cytoplasm is scarce basophilic. The stroma is fibro-inflammatory (Figure 1).

The immunohistochemical study (Figure 2) showed the expression of CD23 (diffuse), PS100 (focal), EMA (focal), CK (focal) and the absence of expression of CD68 and CD1a, allowed us to diagnose a poorly differentiated malignant tumor proliferation expressing the Cytokeratin and anti-CD23 antibodies compatible with follicular dendritic cell sarcoma.

Figure2: Polygonal cells with abundant cytoplasm, eosinophils and well rounded or oval nuclei with fine chromatin, moderate atypia with some atypical mitoses, numerous lymphocytes and neutrophils isolated or grouped into clusters.



The immunohistochemical study (Figure 3,4) showed the expression of CD23 (diffuse), PS100 (focal), EMA (focal), CK (focal) and the absence of expression of CD68 and CD1a, allowed us to diagnose a poorly differentiated

malignant tumor proliferation expressing the Cytokeratin and anti-CD23 antibodies compatible with follicular dendritic cell sarcoma.

Figure3: positive immunostains in a case of spindle cell FDSC for CD23.

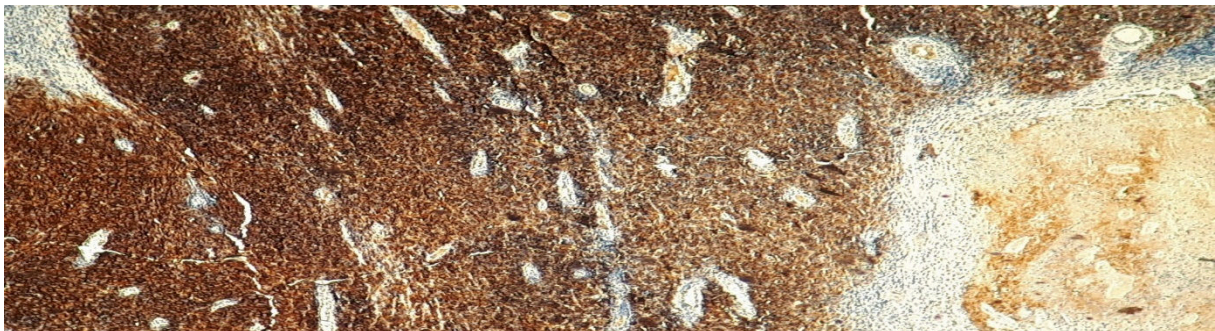
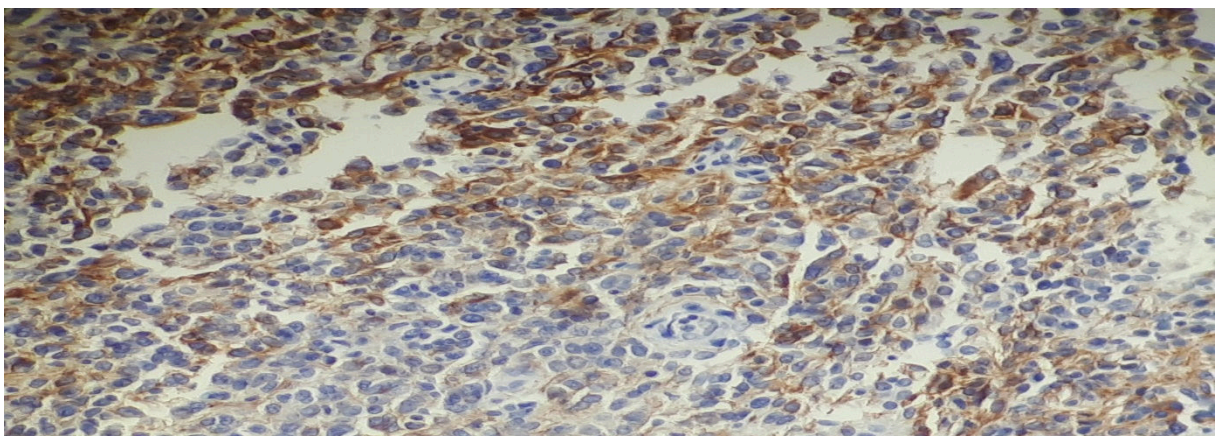


Figure 4: focal positivity immunostains in a case of spindle cell FDSC for CK



DISCUSSION

Follicular dendritic cells: origin and phenotype

Follicular dendritic cells (FDCs), also known as dendritic reticulum cells, are essential for humoral response by providing antigen presentation and maturation of B-cell response. FDCs are located primarily in the germinal centers of lymphoid follicles primary and secondary nodal and extranodal sites [1]. FDCs have pale fibrillar cytoplasm, indistinct cell membranes with long dendritic processes, round nuclei with fine chromatin, and small nucleoli. FDCs are often bi or multinucleate and the nuclei overlap or show a molding (so-called "kissing" model). [2] In electron microscopy, the FDC extensions form a complex network connected via desmosomes and are covered with an electron-dense amorphous material corresponding to the immunocomplexes, [2]. They express one or more dendritic follicular markers: CD21, CD23, CD35 and CXCL13. They express more rarely EMA, PS100, CD68, CD45 and CD20 antigens.

History of FDSCS

In 1986, Monda and Rosai reported for the first time four cases of FDC-derived tumors, all occurring in the cervical lymph nodes [3], now recognized as follicular dendritic cell sarcoma [4]. Follicular dendritic cell sarcoma (FDSCS): categorized as histiocytic and dendritic cell neoplasms in the WHO 2017 classification of hematopoietic and lymphoid tissue tumors. DCS are unusual tumors. Only 400 cases are described in the literature, probably due to an under-diagnosis of this sarcoma [2].

The histogenesis of this disease is still ambiguous, a proportion of cases appears in the context of a Castleman vasculo-hyaline disease, with a pre-sarcomatous phase in the form of hyperplasia of CFDs outside the follicles [5]. Castleman's disease may be associated with CFD sarcoma or may precede it for several years [5]. The location of the tumor is most often extra-nodal in 60% of cases but can be ganglionic in 30% of cases. The lymph node

involvement is most often cervical. Extrganglionic lesions affect the tonsils, the digestive tract, the soft tissues, the mediastinum, the retroperitoneum [6, 7]. It is difficult to analyze the clinical and evolutionary features characteristic of follicular dendritic cell sarcoma (FDSCS) since it is so rare a diagnosis. There are no major series in the literature. Most patients have presentation-like disease as a slow-growing, isolated tumor with no systemic syndrome. Pulmonary and hepatic metastases are possible [5].

Macroscopic appearance: It is a well-circumscribed tumor, with an average size of 7 cm, tanned solid with fickle foci of necrosis and haemorrhagic reshaping.

Microscopic appearance: The FDSCS presents a wide architectural range, tumor cells are fusiform or more rarely ovoid forming sometimes storiform bundles and vortices or sometimes vaguely nodular windings. The cytoplasmic boundaries are indistinct and the nuclei are oval or elongated with fine chromatin, sometimes small nucleoli. Cytological atypia may be very discrete or more pronounced in some cases, as may the mitotic index [2]. Spindle-shaped cells are closely intertwined with a contingent of small lymphocytes more or less abundant.

Histochemical phenotype: The diagnosis of FDSCS requires the support of immunohistochemistry, the use of a large panel of antibodies, given the frequency of loss of sarcomatous follicular dendritic cell specific antigens and the possibility of focal reactivity for epithelial markers [8,9].

Dendritic follicular cell sarcoma is positive for the usual markers of CFDs such as CD21, CD35 and CD23. Frequent expression of podoplanin (D2-40) is observed but not specific. Cluster is also often positive. Other less specific markers may be positive such as desmoplakine, vimentin, fascine, EGFR, as well as PDL1 and more rarely EMA, PS100, CD68 [10]. Ki67 is often low (average of 13%). Follicular tumor dendritic cells express CXCL13 in a diffuse and intense manner,

which differentiates them from other subtypes of dendritic cell sarcomas. Recent markers like FDCSP and serglycin seem very specific. According to Luisa Lorenzi and al [10, 11], in the FDC-S diagnosis, the FDCSP and the SRGN showed both a good sensitivity and a very good specificity for CD23 and Claudin 4, evaluated on 214 controls including carcinomas, soft tissue tumors, melanomas, thymomas and interdigitating dendritic cell sarcomas. SRGN, as well as CD21 and CD35, was negative in all non-FDC-S tumors. The combination of CXCL13, CD21, CD35, FDCSP and SRGN achieved a very good discriminatory power in distinguishing FDC-S from other soft tissue tumors. In particular, CXCL13, all properly classified FDC-S cases were positive for at least two markers among CXCL13, CD21, CD35, FDCSP and SRGN [10, 12, 13]. This information could be easily applicable in the routine diagnosis of this tumor.

Differential diagnosis: The main differential diagnosis is metastasis of undifferentiated carcinoma. Nevertheless, the absence of expression of cytokeratin AE1 / AE3 by the tumor cells eliminates this diagnosis. Another differential diagnosis of FDCS is interdigital cell sarcoma. The latter, with a clinical and morphological presentation comparable to that of dendritic follicular cell sarcoma, is distinguished from the latter by the absence of expression of markers of dendritic follicular cells. and the expression of intense and diffuse PS100 by tumor cells [14]. The recommended treatment is surgical resection sometimes associated with adjuvant chemotherapy. The evolution of SCFD can be complicated by local recurrence in 50% of cases and by metastases in 25% of cases [6].

CONCLUSION

Follicular dendritic cell sarcoma remains a nosological entity in the course of dismemberment which poses diagnostic difficulties. The demand for a histological proofreading within a framework of a specialized network finds all its interest as well

as an expert CPR for the therapeutic management.

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