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Surgery

Cervical Follicular Dendritic Sarcoma: A Rare and Challenging Diagnosis

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Abstract **Case Report**

Dendritic follicular sarcoma (DFS) is a relatively new tumor, very rare and often poorly understood. It occurs mainly in the peripheral lymph nodes at the expense of dendritic follicular cells. However, extra nodal locations have also been reported in the literature. Diagnosis is often difficult, depending mainly on immunohistochemistry and histopathology. There are currently no consensus treatment strategies proposed by the National Comprehensive Cancer Network. The aim of this work is to highlight the different aspects of follicular dendritic sarcoma. We report a rare case of cervical follicular dendritic sarcoma with a review of the literature.

Keywords: Dendritic Follicular Sarcoma, Diagnosis, Histopathology, Immunohistochemistry, Surgery.

INTRODUCTION

Dendritic follicular sarcoma (DFSS) is a very rare and often misunderstood neoplasm. It occurs mainly in peripheral lymph nodes at the expense of dendritic follicular cells.

However, extra nodal locations have also been reported in the literature [1]. Diagnosis is often difficult, relying mainly on histopathological and immunohistochemical data. There is currently no consensus treatment strategy proposed by the National Comprehensive Cancer Network. The aim of this paper is to focus on the different epidemiological, histopathological, immunohistochemical and therapeutic aspects of follicular dendritic sarcoma [2].

CASE REPORT

Mr S.D, 51 years old with no particular pathological history, presented to our department for a right laterocervical mass that had been evolving for 5 years without dyspnea, dysphagia, dysphonia or other associated signs, in a context of apyrexia and conservation of the general state. The clinical examination revealed a firm, fixed, painless right upper jugulocarotid mass with no inflammatory signs. Cervical MRI showed two contiguous right laterocervical tissue masses, well encapsulated, well limited and necrotic with a mass effect on the larynx without vascular involvement. The patient had undergone tumor reduction. Histology was consistent with a follicular dendritic sarcoma. A cervical CT scan was performed 2 months postoperatively which showed a mixed right laterocervical mass with a thickened wall and hypodense center pushing medially and posteriorly against the primary carotid artery and anteriorly against the internal jugular vein with no evidence of locoregional invasion (Figure 1). The patient then underwent total removal of the tumor mass with ligation of the internal jugular vein (Figure2). The anatomopathological study showed a tumor proliferation made of fusiform cells with irregularly enlarged nucleolus, nucleolated with moderate atypia without perineurial malignant peripheral nerve sheath or neoplastic vascular embolism. Immunohistochemistry showed positive anti-CD23, anti-CD35, anti-vimentin and anti-PS100 antibodies. The Ki-67 index was positive on 15% of the cells. This profile concluded in a follicular dendritic sarcoma. The postoperative course was simple. The patient was then referred to oncology for further radiotherapy. At 6 months, no signs of recurrence were noted.

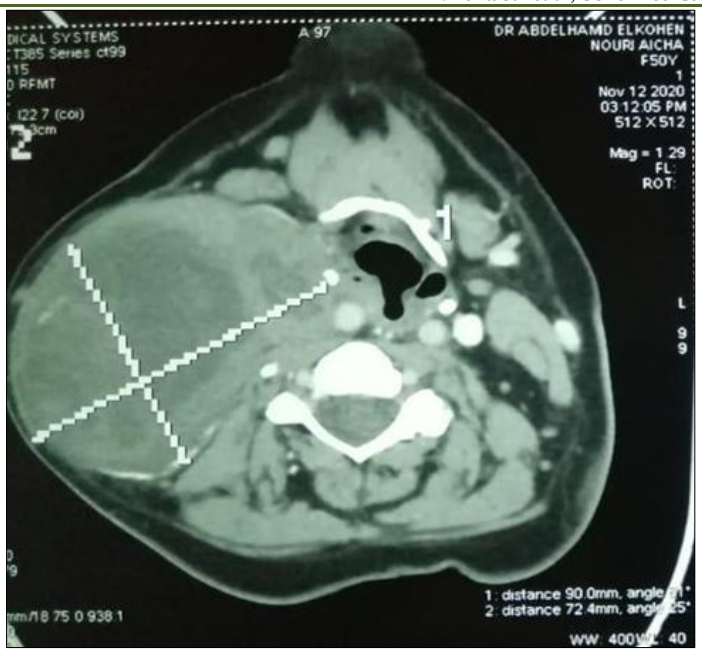

Figure 1: Cervical CT scan showing a mixed right laterocervical mass with hypodense centre compressing the vascular axis

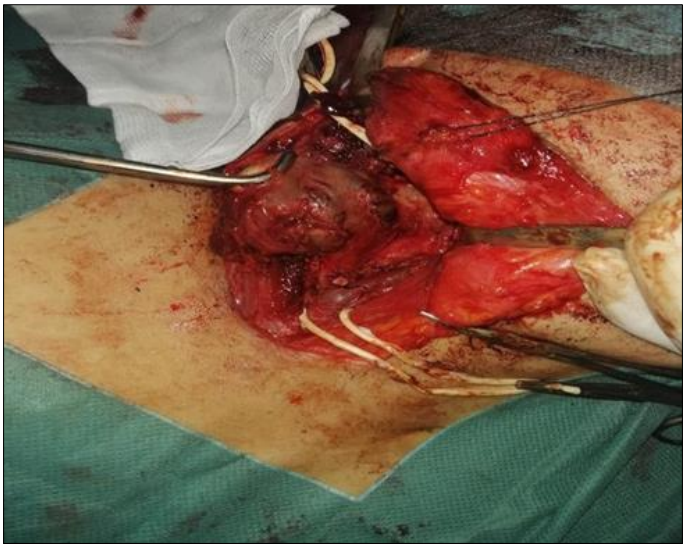

Figure 2: The intraoperative image of the tumor

Follicular dendritic sarcoma (FDS) is a relatively rare entity derived from follicular dendritic cells, which are antigen-presenting cells to B lymphocytes in the germinal centers of lymphoid follicles, thus playing a crucial role in humoral immunity. It mainly affects peripheral lymph nodes, particularly those in the head and neck. However, extranodal tissues are affected in about 30% of cases, particularly the pharynx, oral cavity, thyroid gland, pancreas, mediastinum and peritoneum [1]. SFD is a relatively new nosological entity. It was first described by Monda *et al*., in 1968 [2]. It is considered to be a tumor of intermediate malignancy because of its aggressive and metastatic potential [3]. The age of our patient was 50 years which is consistent with data from different series with a mean age of 45 years and a median age of 50 years. No gender or ethnic predominance was noted. However, a slight female predominance has been observed in the inflammatory pseudotumor like variant [4, 5]. Although the oncogenesis of SFD remains imperfectly known due to the small number of cases reported in the literature, some authors have described the possibility of incrimination of Castleman's disease, which is an Angio follicular lymphoid hyperplasia from residual germinal centers [6]. The role of Epstein Barr virus (EBV) has also been suspected due to the tumor expression of the CD-21 marker, which is a receptor for this virus. Numerous studies have also suggested an association between SFD and autoimmunity, with paraneoplastic pemphigus being the most frequently reported, followed by myasthenia gravis [7, 8]. The diagnosis of SFD is difficult and often poorly understood, due to its silent symptomatology, extremely varied morphology and aspecific imaging. The main

differential diagnoses are inflammatory myofibroblastic tumors, pseudotumourous tumors, interdigitating dendritic cell sarcomas, lymphomas and undifferentiated carcinomas. Diagnosis is based primarily on pathological examination with immunohistochemical complement. Macroscopically, follicular dendritic sarcoma is most often a well-limited, firm, white to greyish mass, varying in size from 1 to 20 cm long axis depending on the location (average 5 cm). In our case, the tumor was 6 cm. Areas of necrosis or hemorrhage may be seen in large tumors. Microscopically, the tumor proliferation is made up of spindle or ovoid cells with sparse eosinophilic cytoplasm. The nuclei are usually elongated with vesicular chromatin, sometimes granular, and a small nucleolus. The cells are arranged in bundles, fascicles, trabecular or diffuse sheets or whorls. The mitotic index is 0 to ten mitoses/ten fields [9]. A high mitotic rate (11 to 35 mitoses per 10 fields), the presence of necrosis and significant cellular atypia are indicative of high-grade tumors. As for the immunohistochemical profile, the tumor cells are positive for at least one of the follicular dendritic cell markers, namely CD23, CD21 or CD35. In our case, the CD23 marker was strongly positive. In addition, the tumor may variably express vimentin, CD68, S100 and EMA which may present diagnostic pitfalls. However, there is no immunoreactivity for CD3, CD1, CD30, CD34, and for HMB-45. In our case, the Ki-67 proliferation index was 15% which is in line with the literature (1-25%). [10] In terms of treatment, no prospective study comparing the different therapeutic modalities exists in the literature. Thus, no therapeutic consensus in the management of SFD has been established to date. Most clinicians rely on the recommendations for high-grade sarcoma, opting for radical surgery as the standard of care [2]. In the series by Perkins and Shinohara [12], one of the largest series in the literature - surgery was considered in 95% of cases. However, several studies have shown that the risk of recurrence and of metastasis, particularly to the lungs, is 40% and 28% respectively, suggesting that adjuvant treatment should be adopted [11]. Gounder *et al*., [13], showed no difference in 5- year disease-free survival between patients with follicular dendritic sarcoma treated by surgery alone and those who received adjuvant or neoadjuvant radiotherapy. In contrast, Soriano *et al*., [14], found a higher 5-year survival rate in patients who received surgical protocols combined with neoadjuvant radiotherapy. In the Li *et al*., series, only 10% of patients received radiation in ENT forms [15]. No adjuvant radiotherapy is then systematic according to the literature. The indications for chemotherapy are also controversial. In a series of 17 cases of SFD by Chan J.K *et al*., seven patients underwent CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) chemotherapy. Four cases of tumor recurrence were noted at 2 years. However, the two cases treated with neoadjuvant chemotherapy had a tumor response of more than 95% [16-18]. In the series by Perkin and Shinohara [12], no additional chemotherapy was adopted.

CONCLUSION

Follicular dendritic sarcoma is a tumor of intermediate malignancy. Although the diagnosis is strongly suspected histologically, additional immunostaining is routinely performed to avoid the risk of misdiagnosis. There is currently no consensus on treatment. Nevertheless, surgery remains the cornerstone of treatment for SFD.

DECLARATION

Author Contributions:

The first draft of the manuscript was written by AZ and AR all authors commented on previous versions of the manuscript. All authors read and approved the final manuscript.

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Declarations

Conflict of interest: The authors have no relevant financial or non- financial interests to disclose.

Ethics Approval and Consent to Participate Informed consent was obtained from all individual participants included in the study.

Consent to Publish: The authors affirm that human research participants provided informed consent for publication of the images in all Figures.

Informed Consent Written informed consent was obtained from all subjects (patients) in this study.

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