ISSN No:-2456-2165

Follicular Dendritic Cell Sarcoma of the Tonssilar: About a Case

M.Belhouari, H.Zidani, Z.Qabala, S.Khalfi , M.Bourhafour , H.Jouhadi , , N.Tawfik , Z. Bouchbika, N.Benchakroune, S.Sahraoui, A.Benider
Oncology-Radiotherapy Department 1
Pathological Anatomy Laboratory 2
Faculty of Medicine and Pharmacy.
Hassan II University
University Hospital IBN Rochd Casablanca

Abstract:- Follicular dendritic cell sarcoma is a rare location and are generally indolent. whose diagnosis is essentially anatomopathological which remains delicate in view of the similarity with other types of soft tissue sarcomas, the therapeutic strategy is not clear because of the rarity of cases reported by the literature.

We describe the case of a 30-year-old patient followed for tonsil mass, pathologic study revealed of a dendritic follicular cell sarcoma of the tonsillar epithelioid type, the systemic treatment for his metastatic relapse. In this article on will raise the clinical, histology and therapeutic aspects of this location with a review of literature.

Keywords:- *Tonsillar, Follicular Dendritic Cell Sarcoma, Metastasis, Chemotherapy*

I. INTRODUCTION

Follicular dendritic cells (FDCs) are cells of the innate immune system found in the primary and secondary lymphatic follicles of the B-cell zones of lymphoid tissue (1). They play a role in antigen presentation, induction and maintenance of the humoral response. Follicular sarcomatous tumors are rare, initially described in 1982 based on 4 cases developed in the lymph nodes by Monda et al(2). These tumors most commonly arise in the lymph nodes, but also in several extranodal sites, including the oropharynx, soft tissues of the neck, parapharyngeal space, thyroid, liver, kidney and axial skeleton, among others(3). We report a case of this rare entity which developed in the tonsil of 36 years old man. Here we present our case with a review of the literature.

II. PATIENT AND OBSERVATION

This is a 30-year-old patient with no particular personal or family pathological history, consulting for a left tonsillar mass **(Figure 1),** for which He had a biopsy of the mass, the pathological examination had shown one of a process totally undifferentiated malignant tumor, then CF MRI showing a left tonsillar process 39.2 * 24 * 23.7 mm with minimal extension at the base of the skull **(Figure3).**

The patient was subsequently operated on and histological examination revealed a tonsillar mucosa subtended by tumor proliferation, sometimes epithelioid and sometimes fusiform, with the presence of abnormal mitoses, the stroma is richly vascularized. the immunohistochemical study had shown negativity of: cytokeratin, AE1AE3, CD45, CD3, CD20, P40, PS100, Sat6, PS100, CD34, desmine, MOP, EMA, CD30, CD99, WT, Melan A, HBB45 BLC2, on the other hand, it diffuse expression of CD33, in view of these morphological and immunohistochemical arguments, the diagnosis of a dendritic follicular cell sarcoma of the tonsillar epithelioid type arriving in places in contact with the limits of intermediate grade excision was retained (**Table1**).

The case was discussed in a multidisciplinary consultation meeting, the collegial decision of which was to provide for postoperative radiotherapy. Then the patient was sent to us for additional treatment. The CT chest /abdominal /pelvis carried out as part of the extension assessment included had shown an asymmetry of the tonsillar regions without anomaly of rise of contrast product without anomaly of PDC, a necrotic lymphadenopathy of the left IIA of 16 mm, a slight infiltration of the fat of the prestylar space. homolateral, at the pulmonary level: bilateral pulmonary micro and nodules (all sub pleural, the largest is left mediobasal measuring 10 mm, with mediastinal lymphadenopathy: left hilar and pre-vascular, the largest of which is necrotic pre-vascular, measuring 41 * 31 mm, on Conclusion: a pulmonary involvement associated with cervical mediastinal nodes in favor of seen the con of a secondary origin , a positron emission tomography (PET) - scanography was indicated but currently problem of availability .

A biopsy of the prevascular mediastinal lymphadenopathy revealed a morphological appearance compatible with that of follicular cell sarcoma, the indication for adjuvant radiotherapy was canceled due to the disseminated nature of the disease, the patient received a the CHOP protocol (given the follicular subtype, it is treated like a lymphoma) with good overall tolerance and very good clinical response (Figure 2), then the patient referred to the hematology department for further treatment.

ISSN No:-2456-2165

III. DISCUSSION

We report a case of follicular dendritic cell sarcoma of the left tonsil, Follicular dendritic sarcoma is exceedingly rare. The incidence of the disease in both sexes is the same with a 1:1 ratio(5), age ranges from 14-77 years with a median of 45 years(4). a lymphoproliferative disease like Castleman's pathology is associated in approximately 20% of cases, EVB may be implicated as a viral risk factor specially on liver splenic location. the main localization is lymph node, however a localization in the head and the neck it is the predominant for the extraganglionic form, followed by hepatic, tonsil and intra-abdominal soft tissue [4,6]. Clinically the symptomatology is poor, with a risk of distant metastasis which remains questionable some authors suggest that this histological type is aggressive but other authors consider it of low metastatic risk having the character of low-grade soft tissue sarcoma [7,8,9].

The diagnosis of FDCS is based essentially on histology (morphology, immunohistochemistry and electron microscopy). Macroscopically, follicular cell sarcoma is generally well limited, solid in consistency, grey in color with in some cases, foci of necrosis and hemorrhage. Microscopically, the tumor proliferation is made of spindle-shaped, polygonal or ovoid cells of variable architecture (storiform, fascicular, sheet-likor nodular, sometimes with coils). The nucleus is elongated, with vesicular or finely granular chromatin and containing a nucleolus which is sometimes prominent. The mitotic index is 0 to 10 mitoses/ten fields. Residual lymphoid tissue is sometimes present as a residual germinal center or as a cluster of small perimetric of small lymphocytes in the perivascular area. Immunophenotypes of follicular dendritic cell sarcoma FDCS are positive for CD21, CD35, Ki-M4p, and KiFDC1p, and are variably positive for vimentin, S-100 protein, CD68, and muscle-specific actin. CD21 and CD35 are the most useful markers for diagnosis because of their specificity [1,10]. In total Follicular dendritic cell sarcomas (FDCS) is an extremely rare entity posing diagnostic difficulties, most often requiring rereading, this is the case of our patient who required a whole panel of markers, and rereading before making the final diagnosis.

The management of sarcoma requires in most situations for optimal results is a multimodal treatment combining surgery, chemotherapy, and radiotherapy in well-selected cases whose decision must be validated in a multidisciplinary meeting.

In a review of the literature on FDCS including twelve patients Fon seca et al reported Than the most used drugs were chemotherapy protocols for the treatment of non-Hodgkin lymphoma in which 2 patients had objective remission after the CHOP protocol [11], and in another series of 14 cases all patients received the standard most used lymphoma protocol almost in all the studies reported which is the CHOP protocol in the same article, they demonstrated that 3 patients in the series received molecules in the 3rd line used in particular in other types of sarcoma, for example gemcitabine, taxane with a response in one patient [12]. In another series of Perkins and Shinohara it was shown that the management of patients followed for FDS is the one adopted for soft tissue sarcoma, i.e. surgical resection followed or not by adjuvant [13].

The irradiation of an ENT sacoma must take into account a certain number of specificities related to histology,the anatomical location of the tumor and to the patient. These rare ENT cancers occur more readily in young subjects. For localized cancers and in a postoperative situation, radiotherapy is essential unless the surgery was planned and performed optimally from a cancer perspective. It is indicated for R1 resection and / or with massive invasion of the initially invaded soft tissues. the technique used is static, rotational or even helical RCMI.

The dose varies from 50 to 54 Gy in the case of complete resection and 66 to 74 Gy in the areas undergoing resection incomplete, with standard fractionation at the rate of 5 sessions of 1.8–2 Gy per week. The weak alpha – beta ratio of sarcomas does not allow hypofractionation to be considered outside of protocols specific hadrontherapy or stereotaxic re-irradiation protocols.

T1- or T2-weighted MRI with gadolinium injection should be merged with the CT scan diagnostic. A post-injection CT scan is required for delineation and is fused with a CT scan without injection used for dosimetry. The place of positron emission tomography (PET) is not validated in sarcomas. The macroscopic tumor volume is defined with MRI (T1 contrast enhancement) and computed tomography. The margins from macroscopic tumor volume to anatomoclinical target volume are 1 to 2 cm and are suitable for specific natural history. Principle bilateral irradiation is not carried out. With some exceptions, there is no prophylactic irradiation of the cervical lymph node areas, the irradiation being limited to known macroscopic invasions. Margins from anatomoclinical target volume to target volume forecast are dependent on the centers and depending on the image guidance technique used from 3 to 5 mm [14].

The evaluation of the prognosis remains delicate because of the rarity of this histological entity, Lan Li, et al report in their article that among the criteria that determine recurrence and mortality are: the histological grade associated with a large tumor size seems to be closely related to the mortality rate as well as a large tumor size is in favor of a risk high recurrence [15].

IV. CONCLUSION

Follicular dendritic cells are rare and generally The tumor is clinically silent, painless, slow-evolution, the tonsillar localization is extremely rare, the diagnosis of which constitutes a challenge requiring the use of a panel of markers, the treatment has not yet been codified because of the rarity of this histological entity.

Declartion of Interest: The Submitters did not submit a Declaration of Conflict of Interest.

ISSN No:-2456-2165

Consent for publication : The patient agreed that doctors could use and publish her case, including the accompanying pictures. A copy of the writing consents is available for review by the editor of this journal.

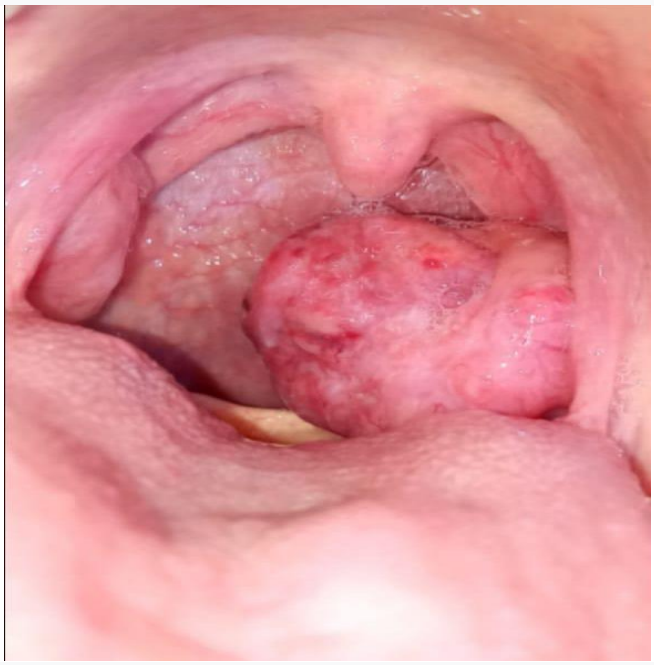

Figure1: clinical aspect of the left tonsil before chemotherapy

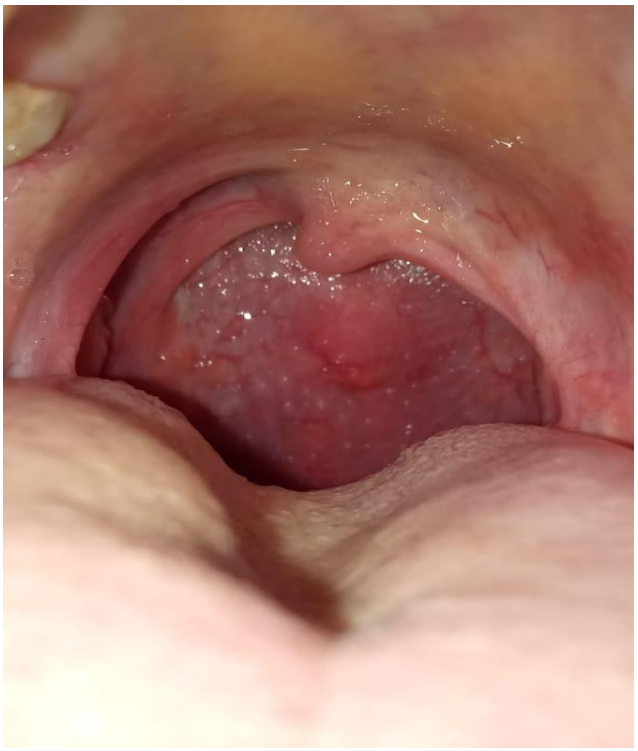

Figure 2: almost complete disappearance of the mass after the first cycle of chemotherapy

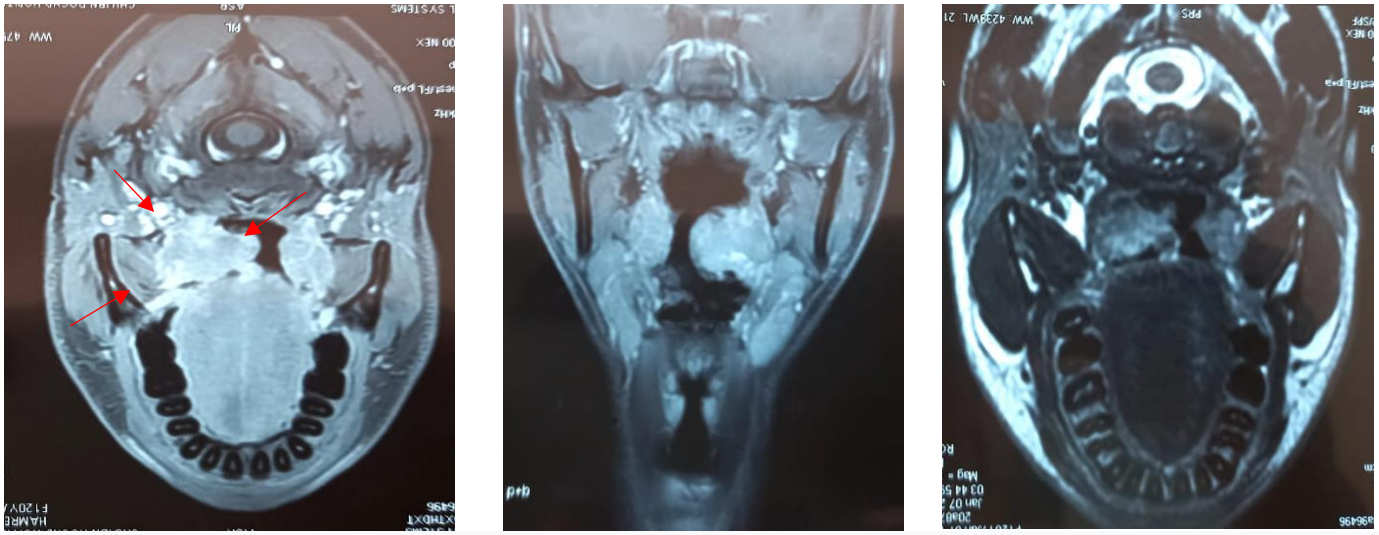

Figure3 : the CT MRI shows a process of the locally advanced left tonsillar region filling the oropharyngeal cavity with polylobed contours in T1 isosignal, heterogeneous hypersignal T2, heterogeneously enhanced after the injection of gadolinium 43x39mm by 41mm

ISSN No:-2456-2165

Table 1 : immunohistochemical results

Antigen negatif	Antigen positif
Cytokeratine	CD23 Diffusely positive
AE1AE3	
CD45	
CD3	
CD20	
P40	
PS100	
Stat6	
CD34	
AML	
Desmine	
MPO	
EMA	
CD30	
CD99	
WT	
HMB45	
MelanA	
BCL2	

REFERENCES

[1]. Chan JK, Fletcher CD, Nayler SJ, Cooper K. Follicular dendritic cell sarcoma. Clinicopathologic analysis of 17 cases suggesting a malignant potential higher than currently recognized. Cancer. 15 janv 1997;79(2):294-313.

[2]. Monda L, Warnke R, Rosai J. A primary lymph node malignancy with features suggestive of dendritic reticulum cell differentiation. A report of 4 cases. Am J Pathol. mars 1986;122(3):562-72.

[3]. Saygin C, Uzunaslan D, Ozguroglu M, Senocak M, Tuzuner N. Dendritic cell sarcoma: a pooled analysis including 462 cases with presentation of our case series. Crit Rev Oncol Hematol. nov 2013;88(2):253-71.

[4]. Eun YG, Kim SW, Kwon KH. Follicular Dendritic Cell Sarcoma of the Tonsil. Yonsei Med J. 2010;51(4):602.

[5]. Idrees MT, Brandwein-Gensler M, Strauchen JA, Gil J, Wang BY. Extranodal follicular dendritic cell tumor of the tonsil: report of a diagnostic pitfall and literature review. Arch Otolaryngol Head Neck Surg. sept 2004;130(9):1109-13.

[6]. Youens KE, Waugh MS. Extranodal follicular dendritic cell sarcoma. Arch Pathol Lab Med 2008; 132: 1683-1687.

[7]. Peeters C, Falini B, Gascoyne RD, Gaulard P, Gatter KC, Isaacson PG, Jaffe ES, Kluin P, Knowles DM, Mason DY, Mori S, Müller-Hermelink HK, Piris MA, Ralfkiaer E, Stein H, Su IJ, Warnke RA, Weiss LM. Tumours of histiocytes and accessory dendritic cells: an immunohistochemical approach to classification from the International Lymphoma Study Group based on 61 cases. Histopathology 2002; 41: 1-29

[8]. Chan JKC, Pileri SA, Delsol G, Fletcher CDM, Weiss LM, Grogg KL. Follicular dendritic cell sarcoma. In: Swerdlow SH, Campo E, Harris NL, Jaffe ES, Pileri SA, Stein H, Thiele J, Vardiman JW, editors. WHO classification of tumors of haematopoietic and lymphoid tissues. 4th ed. Lyon: International Agency for Research on Cancer, 2008: 364-365

[9]. Perez-Ordonez B, Erlandson RA, Rosai J. Follicular dendritic cell tumor: report of 13 additional cases of a distinctive entity. Am J Surg Pathol 1996; 20: 944.

[10]. Khalid T, Folman R. Symptoms in cancer patients and an unusual tumor: Case 3. Follicular dendritic cell sarcoma. J Clin Oncol Off J Am Soc Clin Oncol. 20 déc 2005;23(36):9425-6.

[11]. Fonseca R, Yamakawa M, Nakamura S, et al. Follicular dendritic cell sarcoma and interdigitating reticulum cell sarcoma: A review. Am J Hematol 1998;59: 161–167

[12]. Andres O. Soriano,1 Michael A. Thompson,1 Joan H. Admirand,2 Luis E. Fayad,1 Alma M. Rodriguez,1 J. E et al Follicular dendritic cell sarcoma: A report of 14 cases and a review of the literature Am. J. Hematol. 82:725–728,

[13]. Perkins, Stephanie M. MD; Shinohara, T E Interdigitating and Follicular Dendritic Cell Sarcoma A SEER Analysis American Journal of Clinical Oncolog 36: 395-8.

[14]. LePéchoux et al . Radiotherapy of adult soft tissue sarcoma . Cancer/Radiothérapie 20S (2016) S235–S243

[15]. Li L, Shi YH, Guo ZJ, Qiu T, Guo L, Yang HY, Zhang X, Zhao XM, Su Q et al . Clinicopathological features and prognosis assessment of extranodal follicular dendritic cell sarcoma. *World J Gastroenterol* 2010; 16(20): 2504-2519