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Case Study

Rare Localization Of An Extrapleural Solitary Fibrous Tumor: A Case With Literature Review

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ABSTRACT

The solitary fibrous tumor (TFS) is a rare mesenchymal tumor, described initially in the pleura, but can sit in any anatomical site. Subcutaneous localization in the lumbar region is rarely described.

We report through this work the main super pleural localizations currently described, as well as the most characteristic elements of the diagnosis in terms of histological and immunohistochemical analysis likely to improve their recognition.

This is a 36-year-old woman who was seen for subcutaneous parietal mass in the lumbar region. The histological aspect and the immunohistochemical profile made it possible to evoke the diagnosis of solitary fibrous tumor.

The TFS has the same anatomo-clinical characteristics regardless of its seat. It occurs in adults between 26 and 82 years, with no predominance of sex. It is most often asymptomatic. The radiological aspect is not specific. This unusual superficial presentation in the lumbar region may pose a diagnostic challenge, given the importance of the morphological and immunophenotypic spectrum and the overlap of SFT with other neoplasms. A fusion of the two NAB2-STAT6 genes by intrachromosomal inversion on chromosome 12 (q13q13) is found in 70 to 90% of cases. Postoperative monitoring is necessary given the risk of local recurrence in the absence of predictive criteria for the evolution.

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INTRODUCTION

The fibrous solitary tumor (FST) is a fibroblastic tumor originally described in the pleura [1]. The pleural location presents 32% of cases, followed by meningeal TFS. Extra pleural and extra menigated localizations include retroperitoneum, lower limbs, orbit, bone and skin [2].

Objective of the work: to study the main super pleural sites currently described, as well as the most characteristic elements of the diagnosis in terms of histological and immunohistochemical analysis likely to improve their recognition.

OBSERVATION

The study of super pleural sites is described in the following pages: 36 year old woman right lumbar region. Ultrasonography showed a pattern of subcutaneous mass at subcutaneous tissue t 1,34x 1,48x1 cm, hyperechoic, with poorly circumscribed and speculated contours (fig1). Abdominal computed tomography with injection of contrast objectified a tissue mass at the level of the subcutaneous fat, polylobed contours, spontaneously heterogeneous density, measuring 1.52 x 1.59cm, enhanced after injection of the contrast product and not adherent to the deep planes. The patient was operated on. There was no extension to neighboring organs facilitating its resection. Histological examination had shown tumor tissue that had variable cellularity. The tumor cells were round or fusiform. They had monotonous nuclei showing no atypia. Mitoses were rare. Immunolabeling was negative for epithelial markers (cytokeratin and EMA) for desmin, c-kit and S100 but positively positive for STAT6 (Fig 2). The postoperative course was simple.

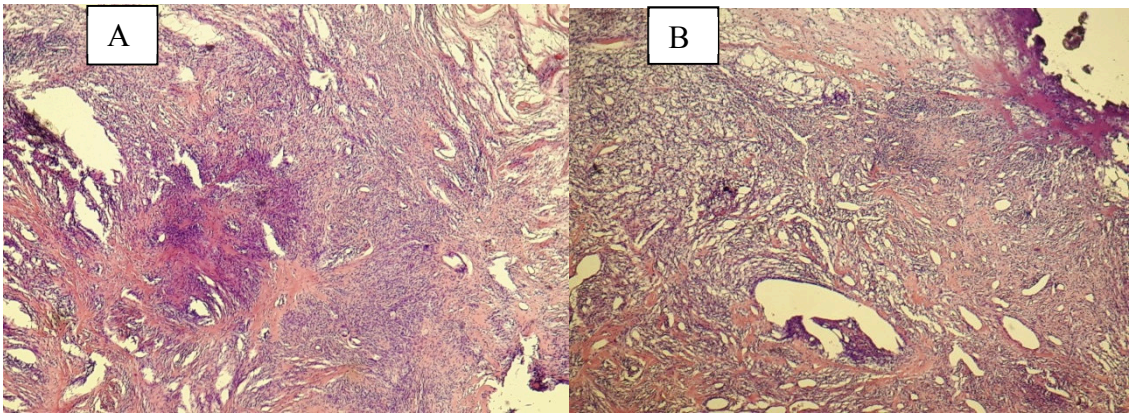

Figure 1 : A et B, monomorphic proliferation of hypocellular fusiform cells, atypical, with no particular pattern less pattern, abundant vascularization, of tree architecture, made of vessels of varied caliber.

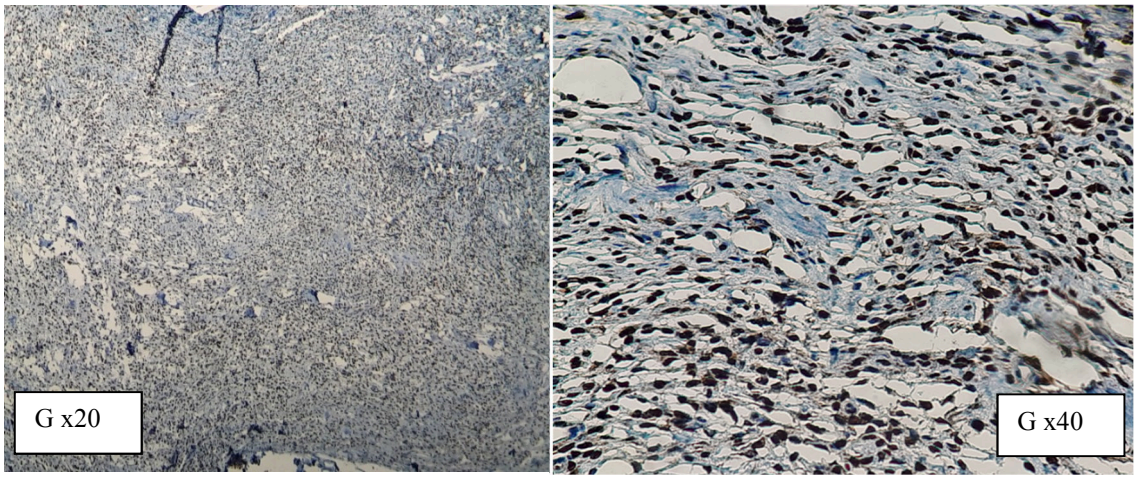

Figure 2: STAT6: Diffuse and intense nuclear staining.

DISCUSSION

TFS are previously classified as: hemangiopericytoma, localized fibrous mesothelioma and solitary fibrous mesothelioma [3]. The WHO (2016) classification of tumors of the central nervous system distinguishes hemangiopericytomas solitary fibrous tumors of the meninges. That of the soft tissues WHO (2013) brought them together since [4].

In 2013, discovery of a common oncogenesis by fusion of NAB2-STAT6 genes. Different types of fusion have been reported which correlate with the different histological variants [5]. Solitary fibrous tumor (SFT) can occur at any age, more common between the sixth and seventh decade. Sex ratio H / F: 1. Its etiology remains largely unknown [2]. The extrapleural localizations reported in the literature are very varied; ovaries, cervix, vagina, scrotum, prostate, seminal vesicle, kidney, mammary gland and pericardium. Superficial SFTs are much less widespread than deep ones. Reported cases were more frequent at the head level with a discreet female predominance. The clinical signs are variable depending on the location, often related to compression effects on adjacent structures, hypoglycemia may be associated (secretion of insulin-like growth factor) [2, 6, 7]. Ultrasound shows an echogenic solid mass with significant Doppler vascularization. Macroscopically, extra pleural SFT are similar to pleural ones. They are well circumscribed, often encapsulated, lobulated in appearance and firm in consistency. Their size varies from 1 to 20 cm. Microscopically, it is a fusocellular proliferation without real architecture with alternating hypocellular collagenic zones and hypercellular zones. The cells are not very atypical with little mitosis. The vascularization is arborized of hemangiopericy type. The stroma reaction is sclerosing, hyalinizing, sometimes myxoid or oedematous [8, 9]. Two variants are reported; the lipomatous TFS with mature adipocyte contingent that can mimic a spindle cell lipoma and SFT with multinucleated giant cells, often reported at the level of the orbit [2]. Most TFS are often benign. Malignant TFS are often hyper-cellular with numerous mitoses (> 4/10 large fields) [9]. Immunohistochemistry plays a determining role in the histopathological diagnosis. The most sensitive marker with a 100% specificity is STAT6 with an intense and diffuse nuclear positivity. It presents a new diagnostic criterion for TFS. Other less specific positive markers are CD34, CD99 and Bcl2 [9]. Joint oncogenesis by fusion of the two NAB2-STAT6 genes reported in 70-90% of TFS by intrachromosomal inversion on chromosome 12 (q13q13). Different types of fusion have been demonstrated which correlate with the different histological variants [5].

The two main differential diagnoses of subcutaneous TFS are spindle cell lipoma and dermatofibrosarcoma protuberans. These last two do not express the STAT6 [9]. Evolution remains unpredictable. 10 to 15% of TFS are aggressive with pulmonary, hepatic and bone metastases. Superficial forms are usually indolent tumors. Most known cases show no documented recurrence or metastasis [2]. The authors agree on the absolute necessity of performing the widest possible excision surgery in order to avoid local recurrences [9,10].

CONCLUSION

This unusual subcutaneous localization of solitary fibrous tumors poses a diagnostic challenge, given the importance of their morphological and immunohistochemical overlap with other neoplasms of the same localization. The anti-STAT6 antibody is useful for confirming the diagnosis of TFS [9]. In the absence of histo-prognostic criteria, any solitary fibrous tumor must be considered as having a malignant potential. Postoperative monitoring is necessary given the risk of local recurrence in the absence of predictive criteria for the evolution.

REFERENCES

1) Sepideh Gholami, MD1, Michael R. Cassidy, MD1, Amanda Kirane, MD2, Deborah Kuk, ScM3. Size and Location are

the Most Important Risk Factors for Malignant Behavior in Resected Solitary Fibrous Tumors. Society of Surgical Oncology 2017

2) Patrick Feasel, MD,* Alyaa Al-Ibraheemi, MD,† Karen Fritchie, MD. Superficial Solitary Fibrous Tumor: A Series of 26 Cases. Am J Surg Pathol 2018;00:000–000).
3) Fletcher C, Bridge J, Hogendoorn P, Mertens F. WHO classification of tumours of soft tissue and bone. IARC Press, Lyon. 2013;80-2.
4) Savary caroline.Tumeurs fibreuses solitaires ethemangiopericytomes des meninges:immunophenotype et histopronostic de 17 cas. Thèse soutenu le 24/12/2016. Université Angers.
5) Hui-Chun Tai1,9, I-Chieh Chuang2,9, Tse-Ching Chen3. NAB2–STAT6 fusion types account for clinicopathological variations in solitaryfibrous tumors. Modern Pathology (2015) 28, 1324–1335 USCAP.
6) Falconieri g., lamovec j., mirra m., pizzolitto S. : Solitary fibrous tumor of the mammary gland : A potential pitfall in breast pathology. Ann. Diag. pathol., 2004 ; 8 : 121-125.
7) Okamura JM, Barr RJ, Battifora H. Solitary fibrous tumor of the skin. Am J Dermatopathol. 1997;19:515–518.
8) Andrea Ronchi, Immacolata Cozzolino, Federica Zito Marino. Extrapleural solitary fibrous tumor: A distinct entity from pleural solitary fibrous tumor. An update on clinical, molecular and diagnostic features. DOI: doi:10.1016/j.anndiagpath.2018.01.004.
9) Bugel h., gobet f., baron m., pfister ch., sibert l., grise p. Tumeur fibreuse solitaire du rein et autres localisations à l'appareil urogénital: caracteristiques morphologiques et immunohistochimiques. Prog.Urol., 2003 ; 13 : 1397-1401.
10) Abdelahad Ounaies (1), Akram Ouni (1), Adnène Moussa (2). La tumeur fibreuse solitaire rétropéritonéale : à propos de deux observations. Progrès en Urologie (2005), 15, 1128-1131
11) Nicholas DeVito1, Evita Henderson1,2, Gang Han3, Clinical Characteristics and Outcomes for Solitary Fibrous Tumor (SFT): A Single Center Experience. DOI:10.1371/journal.pone.0140362 October 15, 2015 1 /
12) Durand x., deligne e., camparo p., riviere p., best o., houlgatte A. : Tumeur fibreuse solitaire de la loge rénale. Prog. Uro., 2003 ;13 : 491-494.

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