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CASE REPORT

DIAGNOSTIC WORK-UP IN THE HISTOLOGICAL VERIFICATION OF A RARE SOLITARY FIBROUS TUMOR

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Abstract. *Solitary fibrous tumors are an extremely rare group of ubiquitous tumors of mesenchymal origin with an incidence of 1 per 1 million per year. The scarcity of published clinical cases describing their cytological findings and clinical features is the reason why the topic is of unique importance to both pathological and surgical studies. Originally thought to be exclusive to the pleural lining, it is now known that solitary fibrous tumors can occur anywhere in the body. In the past decade the diagnostic and therapeutic landscape of patients with verified solitary fibrous tumors has been refined. While fine-needle aspiration remains more accessible, the ultrasound- and computed tomography-guided biopsy paves the way to a narrower differential diagnosis. In the following case report, we present a case of a malignant solid fibrous tumor, located in the abdominal cavity, its clinical presentation, and the arduous diagnostic workup, which lead to its histological verification.*

Key words: *solitary fibrous tumor, ultrasound-guided needle biopsy, rare neoplasms*

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INTRODUCTION

First described in 1870 by Wagner, solitary fibrous tumors (SFT) are a rare group of mesenchymal tumors that are now known to be able to develop almost anywhere in the body. Their behavior is highly variable, ranging from low to highly aggressive, with the dedifferentiated SFT (DD-SFT) being the fastest-growing subtype, accountable for almost 40% of recurrent cases after curative surgery. In 2020 WHO found risk stratification models to be a superior tool of determining prognosis in SFT, as opposed to the classical typical versus malignant differentiation [3]. In 2022, a classification further divided SFT into three categories: benign (locally aggressive), NOS (rarely metastasizing), and malignant [1]. Metastatic spread is estimated to be as high as 45% and greater in series with a longer follow-up period. The 5-year metastasis free rate is reported to be 74%, dropping to 55% in 10 years. Recurrence beyond 10 years is said to be seen in up to 10% of patients and is as low as 18% in 20 years [5].

This pathology shows preference for adult patients, with a median age of 50 to 60 years old. The most common sites of occurrence were cited to be the pleura – 30%, meninges – 25%, abdominal cavity –

20%, trunk – 10%, extremities – 10%, head and neck – 5% [4].

There have been reports of less than 10% of patients suffering paraneoplastic syndromes, the most common of which was hypertrophic osteoarthropathy (HOA) in pleural SFT with overexpression of vascular endothelial growth factor. In fewer than 5% of cases, there was information of hypoglycemia as a result of non-islet cell tumor hypoglycemia due to overproduction of insulin-like growth factor.

Macroscopically, solid fibrous tumors present as a well-defined mass, which is most often silent in its clinical presentation. The average size as described in literature is around 13.4 cm, keeping in mind that larger SFT are more often malignant than benign. The cut surface is dense and pale in color with areas of fat, hemorrhage, necrosis and myxomatous changes.

Histologically, the tumor is comprised of randomly arranged cells with spindle or ovoid shape, without a collagenous stroma, intermixed with blood vessels with a characteristic staghorn shape [2].

Immunohistochemically, strong nuclear staining of transducer and activator of transcription 6 (STAT6) has been characteristic. Other unspecific markers used in the diagnosis of SFT are CD34, BCL-2, and CD-99. Strong and diffuse expression of CD34 is seen in more than 80% of SFT, but its expression can be lost in the most malignant variants. They show negative S-100 protein, actins, desmin and cytokeratins [6]. However, focal immunoreactivity for cytokeratins, actins, S-100 protein, epithelial membrane antigen can be seen.

Clinical features of this pathology consist of local mass effect symptoms and thus depend on the localization and size of the tumor.

The exact diagnosis depends on the correct imaging studies, with computed tomography (CT) and MRI being at the forefront. Precise ultrasound and CT-guided biopsy are used for further histological verification. Although fine-needle aspiration (FNA) is undoubtfully useful, it provides a non-specific material with a non-characteristic appearance.

CASE PRESENTATION

A male patient in his 70s was admitted to the clinic of thoracic surgery after being treated for pneumonia for several days prior, when a performed CT scan revealed a basal nodule in the left lung. He reported a known abdominal mass, which was biopsied via explorative laparotomy 5 years beforehand, but remained histologically unverified. Both tumors were otherwise asymptomatic. The patient noted a preceding episode of pulmonary embolism 2 years ago and was currently treated with clopidogrel and antihypertensive medication for elevated blood pressure. He had no history of occupational exposure to hazardous materials and disclosed no substance or tobacco abuse. Upon physical examination no peculiarities were noted.

A thoracoabdominal CT scan was carried out in order to evaluate the state of the two known lesions. A 2.3/1.8 cm mass was seen in the posterior basal (10) segment of the left lung against the background of massive fibrosis (Fig. 4, 5, 6). On the abdominal phase a 12.1/11.5 cm mass, arising from the psoas muscle lying intimately adjacent to the common iliac vessels, was revealed (Fig. 1, 2, 3).

Upon discussion, a decision for performance of ultrasound-guided transthoracic cutting needle biopsy to verify the two lesions was made. The two procedures were carried out with no complications in the postoperative period. Histological examination of the lung mass did not achieve diagnosis as it was assumed to be organized fibrosis; however, the biopsy taken from the abdominal tumor underwent further immunohistochemical examination and revealed a poorly differentiated carcinoma. Due to the positive expression of CD34, STAT6, and after excluding any malignant epithelioid neoplasia, the diagnosis of a malignant type of solitary fibrous tumor was made.

The patient was discharged on the first postoperative day and was deemed a viable candidate for curative surgery after operative verification of the lung mass and accurate preoperative evaluation and staging. He, however, declined any further testing and refused treatment due to undisclosed personal reasons.

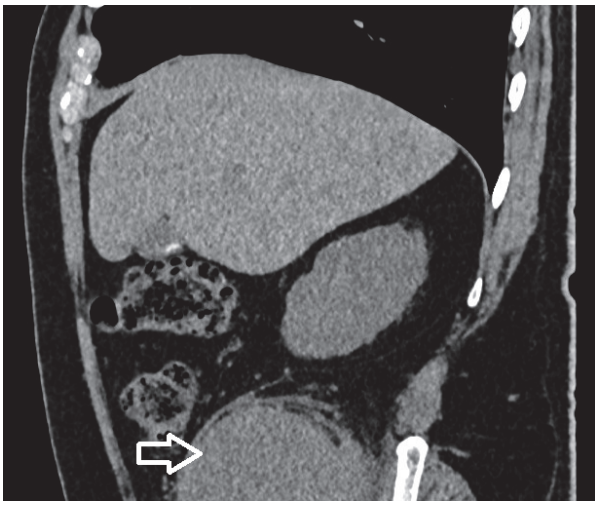

Fig. 1. Showing the abdominal mass, later verified as a SFT

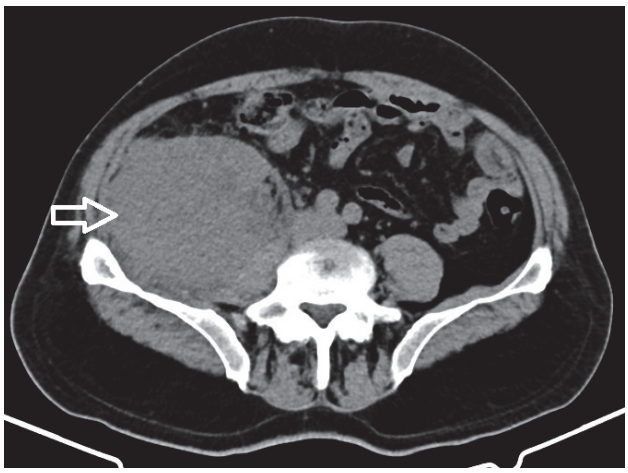

Fig. 2. Showing the abdominal mass, later verified as a SFT

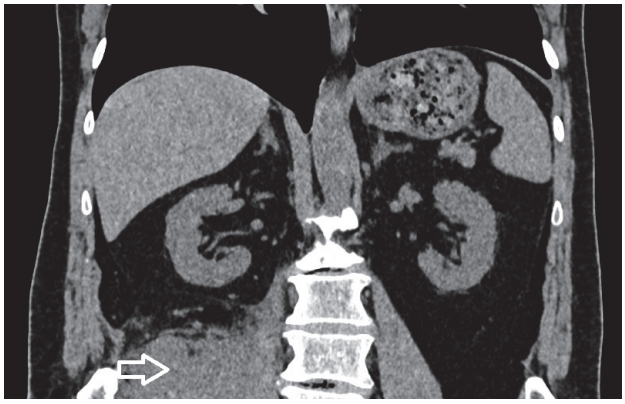

Fig. 3. Showing the abdominal mass, later verified as a SFT

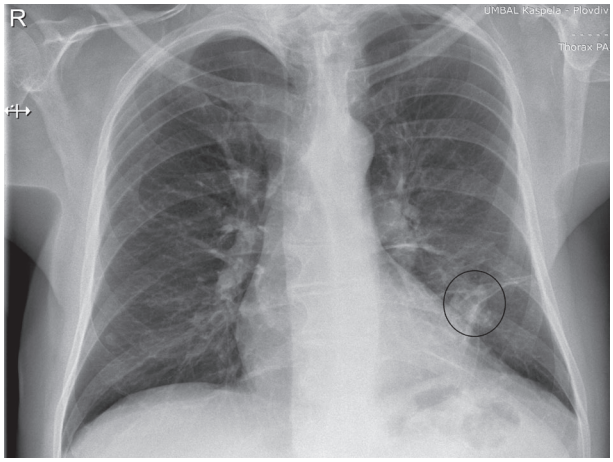

Fig. 4. Preoperative radiographic image

DISCUSSION

Surgical resection of localized or oligometastatic disease remains the gold standard when dealing with solitary fibrous tumors [1]. The aim is to achieve wide resection lines utilizing the sarcoma surgical approach. The technical aspects of surgery vary across different sites of localization. SFT arising from the ab-

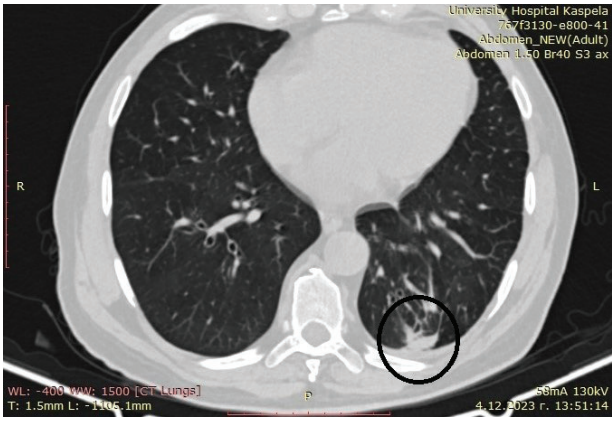

Fig. 5. Image revealing the newly-discovered lesion in the left lung

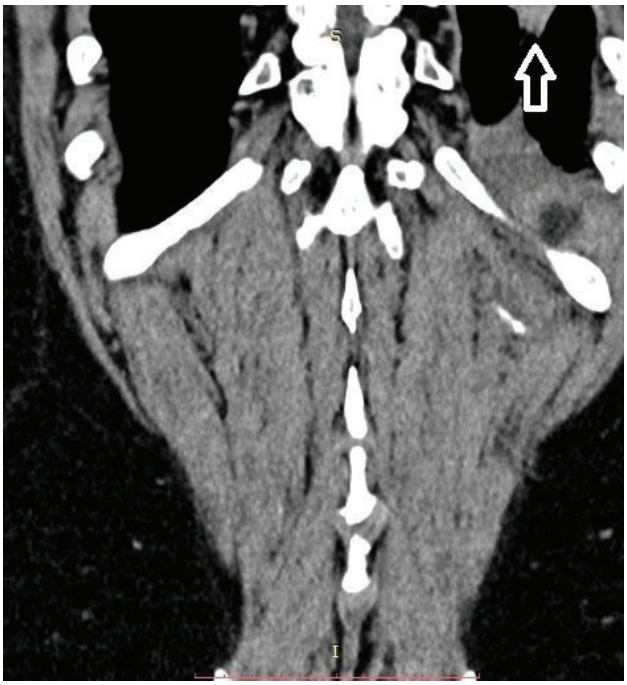

Fig. 6. The image shows the lesion in the left lung as seen in a coronal plane

dominal cavity are predominantly located in the retroperitoneum. They involve the pelvic space relatively frequently, however adjacent organs can be spared as long as they are not encased or invaded.

In a large retrospective study, which includes 549 SFT patients [7], 428 (78%) underwent surgery and 121 (22%) – surgery plus postoperative radiotherapy (RT). A significant benefit (p=0.012) was reported, favoring RT after propensity score matching.

To further back this up, a retrospective series of 40 patients treated with definitive RT (60G) reported an overall response rate (ORR) of 67% with 5-year local control of 81.3% and 5-year overall survival of 87.5%. This leads to the conclusion that the cases, in which

a marginal resection is foreseen, are suitable candidates for neoadjuvant RT.

For metastatic and advanced SFT, chemotherapy remains the last line of defense. However, standard cytotoxic drugs are backed up by a limited amount of prospective evidence. Moreover, no specific clinical trials have been carried out in favor of this type of treatment.

Cytological features of SFT are relatively nonspecific. In the scarce number of series describing cytological features of malignant SFT, malignancy is characterized by hypercellularity, pleomorphism and epithelioid or round cell features, necrosis, and mitoses. In order to attain improved prognostication, stratification models were developed. Among them is the Demicco model (D-score or MDACC score), which is the most commonly used. Its main variables are the age of presentation, tumor size and mitotic count (some authors include tumor necrosis, as well). They are used to classify SFT with low, moderate and high risk of developing metastatic recurrence.

CONCLUSION

In conclusion the pathology of SFT is in the hands of a multidisciplinary team of professionals, working in tandem to figure out the correct treatment plan. Low risk solitary fibrous tumors smaller than 10 cm have a better prognosis with surgical resection. On the other hand, high risk tumors larger than 10 cm with histological signs of malignancy have a poorer prognosis, especially when R0 resection is not achieved. Newer treatment for advanced cases includes antiangiogenic therapy. Its usefulness, however, is yet to be studied.

Disclosure summary: *The authors have nothing to disclose.*

REFERENCES

1. De Bernardi A, Dufresne A, Mishellany F, et al. Novel Therapeutic Options for Solitary Fibrous Tumor: Antiangiogenic Therapy and Beyond. Cancers (Basel). 2022 Feb 20; 14(4):1064. doi: 10.3390/cancers14041064.
2. Khanchel F, Driss M, Mrad K, Romdhane KB. Malignant solitary fibrous tumor in the extremity: Cytopathologic findings. J Cytol. 2012 Apr; 29(2):139-41. doi: 10.4103/0970-9371.97160.
3. Martin-Broto J, Mondaza-Hernandez JL, Moura DS, Hindi N. A Comprehensive Review on Solitary Fibrous Tumor: New Insights for New Horizons. Cancers (Basel). 2021 Jun 10; 13(12):2913. doi: 10.3390/cancers13122913.
4. Niknejad M, Campos A, Machang'a K, et al. Solitary fibrous tumor. Reference article, Radiopaedia.org (Accessed on 21 Jan 2024) https://doi.org/10.53347/rID-21592.
5. Milano MT, Singh DP, Zhang H. Thoracic malignant solitary fibrous tumors: A population-based study of survival. J Thorac Dis 2011; 3(2):99-104. doi:10.3978/j.issn.2072-1439.2011.01.04.
6. Dermawan JK, Fritchie K. Solitary fibrous tumor. PathologyOutlines.com website. https://www.pathologyoutlines.com/topic/softtissuesft.html (Accessed January 21st, 2024).
7. Haas RL, Walraven I, Lecointe-Artzner E, et al. Extrameningeal solitary fibrous tumors-surgery alone or surgery plus perioperative radiotherapy: A retrospective study from the global solitary fibrous tumor initiative in collaboration with the Sarcoma Patients EuroNet. Cancer. 2020 Jul 1; 126(13):3002-3012. doi: 10.1002/cncr.32911.