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Solitary fibrous tumor: A center's experience and an overview of the symptomatology, the diagnostic and therapeutic procedures of this rare tumor

Wolfgang Hohenforst-Schmidt [a], Konstantinos Grapatsas [b], Manfred Dahm [b], Paul Zarogoulidis, M.D, Ph. D [c, *], Vasileios Leivaditis [b], Christophoros Kotoulas [d], Periclis Tomos [e], Efstratios Koletsis [f], Zoi Tsilogianni [g], Naim Benhassen [h], Haidong Huang [i], Christoforos Kosmidis [j], Bora Kosan [a]

[a] Sana Clinic Group Franken, Department of Cardiology/Pulmonology/Intensive Care/Nephrology, "Hof" Clinics, University of Erlangen, Hof, Germany
[b] Department of Cardiothoracic and Vascular Surgery, Westpfalz Klinikum, Academic Educational Hospital, Heidelberg University and Mainz University, Kaiserslautern, Germany
[c] Pulmonary Department-Oncology Unit, "G. Papanikolaou" General Hospital, Aristotle University of Thessaloniki, Thessaloniki, Greece
[d] Department of Cardiac Surgery, 401 General Military Hospital of Athens, Athens, Greece
[e] Department of Thoracic Surgery, "Attikon" University Hospital of Athens, Athens University Medical School, Athens, Greece
[f] Department of Cardiothoracic Surgery, Medical School, University of Patras, Patras, Greece
[g] First Department of Internal Medicine, 401 General Military Hospital of Athens, Athens, Greece
[h] Medical Clinic I, "Fuerth" Hospital, University of Erlangen, Fuerth, Germany
[i] Department of Respiratory and Critical Care Medicine, Changhai Hospital, The Second Military Medical University, Shanghai, China
[j] Surgery Department, Interbalkan European Medical Center, Thessaloniki, Greece

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ABSTRACT

Solitary Fibrous Tumor of the Pleura (SFTP) is a rare tumor of the pleura. Worldwide about 800 patients diagnosed with this oncological entity have been described in the existing literature. We report our center's 13 year experience. During this time three patients suffering from this rare disease have been treated in our department. All patients were asymptomatic and their diagnosis was initially triggered by a random finding in a routine chest x-ray. The diagnosis was set preoperatively through a needle biopsy under computer tomography (CT) guidance. The tumors were resected surgically though video-assisted thoracoscopic surgery (VATS) or thoracotomy. Because of the lack of specific guidelines due to the rarity of the disease a long-term, systematic follow-up was recommended and performed. Parallel an overview of the diagnostic and therapeutic procedures of the rare tumor is made.

1. Introduction

The tumors of the pleura are an important nosological entity of the thoracic cavity. The most known tumor of the pleura is the mesothelioma. However, other tumors of the pleura have also been described. A less known and less common tumor is the solitary fibrous tumor of the pleura (SFTP). SFTP is a rare localized mesenchymal tumor which was initially thought to be a mesothelial pleural lesion [1]. Solitary fibrous tumors can arise from visceral organs or mesothelial tissues [1,2]. Solitary fibrous tumors have also been described in other localizations such as the pelvis, abdomen, retroperitoneum, buccal space, maxillary sinus, liver, pancreas, suprarenal region, and kidneys. It is believed that these tumors originate from extrapleural sites of these anatomical cavities and organs [3]. As far as the pleural solitary fibrous tumors are concerned, about 800 cases of SFTP have been described in the literature. Historically several terms have been used to describe this tumor, such as benign mesothelioma, localized mesothelioma, localized fibrous mesothelioma, localized fibrous tumor of the pleura, sub-pleural fibroma, pleural fibroma, localized benign fibroma, and sub-mesothelial fibroma [1,4,5]. The first description of the tumor is chronologically debatable. The first description of this entity is contributed to Lieutaud in 1767, while in other reports

* Corresponding author.
E-mail address: pzarog@hotmail.com (P. Zarogoulidis).

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suggest that it was first described by Wagner in 1870. The first official description of the tumor's pathology was however made by Klemperer and Rabin. The majority of tumors are benign, but 10–20% of the tumors are malignant [1,4,6].

The tumor often presents no symptomatology and is usually randomly discovered during a routine chest x-ray. During the period of the last six years (2010–2016) three patients with SFTP were treated in the Department of thoracic and cardiovascular surgery in Kaiserslautern. The preoperative diagnosis was made through needle biopsy under computed tomography guidance. The patients underwent surgical excision and a subsequent long term follow-up. In this article we attempt to describe our experience in this field as well as to present a general overview of the existing literature regarding the diagnostic and therapeutic procedures of this rare tumor.

2. Cases presentation

2.1. 1st case

A-56-year-old female was presented to our outpatient clinic with a mass in the area of the lower lobe of the left lung that was incidentally revealed by a chest x-ray performed due to influenza symptoms. No other symptoms or physical sings implying malignancy existed. Patient's past medical history revealed COPD, hypothyroidism, type 2 diabetes mellitus and heavy smoking of thirty pack years (py). The chest computer tomography (CT) revealed a pleural mass having a size of $4 \times 7 \times 5$ cm. A needle biopsy under CT guidance was performed and histology showed a SFTP. A further staging with bronchoscopy and Positron Emission Tomography - Computed Tomography (PET-CT) revealed no further pathological findings. A left posterolateral thoracotomy was performed. A tumor arising from the lower lobe with no infiltration of the thorax wall was found. A complete tumor resection with atypical lung parenchyma wedge resection was performed. The histopathological examination of the mass revealed a large SFPT of 11 cm diameter with circumscribed subcapsular necrosis, sometimes moderately gradiger nuclear pleomorphism and with 3 mitoses to 10 HPF (high-power field). The examination findings partially fulfilled the England's criteria for the characterization of a malignant SFTP (described below) [7]. For this reason, the tumor was characterized semi-malignant. In addition the histological examination showed tumor free margins of the resected tissue. In immunohistochemical analysis cells were positive for CD34 and negative for CD117. According to Demicco et al. the stratification risk for the patient was 4. According to the literature a metastasis free disease and a disease-specific survival is expected in a percentage of 64% and 93% respectively expected in ten years [8].

The patient was discharged on the 9th postoperative day. The hospital stay was prolonged due to a postoperative pneumonia that was conservatively treated. During the postoperative follow-up in our outpatient clinic, no complications were observed. The patient did not undergo chemotherapy or radiation. After a systematic (on a 6-month basis) two year follow-up the patient appeared with peripheral, rounded nodules of variable size, scattered throughout both lungs. The patient underwent a new full staging examination. All tests which were carried out including abdominal ultrasound, abdominal CT, colonoscopy, gastroscopy and tests for gynaecological malignancy revealed no pathological findings. A diagnostic video-assisted thoracoscopic surgery (VATS) of the right pleural cavity was performed and tissue biopsies where received. Histologically the biopsy showed no signs of metastases. The findings were attributed to interstitial pneumonia. A further long-term follow-up was suggested.

2.2. 2nd case

A-50-year-old female appeared in our outpatient clinic with a detected mass in the area of the upper lobe of the left lung. This patient's tumor was also revealed incidentally during a chest x-ray examination. She presented no other symptoms. The patient's past medical history revealed hypertension, thrombocytosis and thyroidectomy because of multinodular goiter and smoking. A CT scan revealed the mass (diameter: 3cm) and the final diagnosis of SFTP was also set through a CT needle biopsy. Because of a patient's denial for surgical treatment a follow-up was alternatively suggested. After a three years follow-up time and because of a significant increase of the mass diameter in the last CT (5,3cm) a VATS was finally performed. Intraoperatively the tumor presented no infiltration of the chest wall. A tumor excision was performed through a wedge lung parenchyma resection. The finding of a benign SFTP with a diameter of 6cm was showed histologically. In immunohistochemical assay cells were positive for CD34, positive for less than 2% of cells for Ki-67 and negative for CD117, D2-40 and TTF1. The patient was discharged on the 4th postoperative day without having any complications. A systematic follow-up was in this case also recommended.

2.3. 3rd case

A 77-years-old male patient aged was incidentally diagnosed with a mass in the left hemithorax and a lung nodule in right hemithorax. Patient's past medical history revealed only arterial hypertension. The CT scanning showed a mass of 9cm in diameter in the left hemithorax. The staging procedures with bone scintigraphy and CT showed no sign of metastases. A diagnostic CT guided needle biopsy showed a SFTP. A thoracoscopic wedge resection of the middle lobe showed an old tuberculoma. Because of the size of the tumor an excision through thoracotomy was finally conducted. Intraoperatively the SFTP presented adhesions to the visceral pleura of the lower lobe. The patient's postoperative course was uncomplicated. The patient was discharged on 5th postoperative day after the second surgery. Histology revealed a SFTP sized $9 \times 5.5 \times 4$ cm. In immunohistochemical analysis cells was clearly positive for CD34 and negative for CD117. A long-term follow-up was also recommended to this patient as well.

3. Discussion

SFTP is a very rare tumor [1,4], which has gained appropriate recognition in the last two decades as a discrete pathologic entity [9]. It represents 5% of the tumors of the pleura. Only 800 such cases have been described in the literature between 1931 and 2002 [1,4,6,10]. However, Cardillo et al. reported that the number of the SFTP can possibly be about 960. The patients with SFPT are 2.8 per 100,000 hospitalized patients. The number of SFTP seems to increase, but it is significantly smaller than the commoner tumor of mesothelioma [6]. The age of the tumor diagnosis varies from 5 to 87. However, the most common age of diagnosis is the sixth and seventh decade of life [1,5,6]. The tumor also occurs with the same frequency in men and women [1]. Sook et al. however described a light deviation of frequency to the side of the females [11]. There is no evidence of heredity. A case with a tumor in mother and daughter has however been described in the literature [10]. No association with asbestos exposition, nicotine effusion or exposure to another environmental factor has been reported [1,5,6,9,11]. In our three treated cases two of the patients were females and no hereditary among relatives was reported.

SFTPs often present no special clinical signs and no special symptomatology that could lead the clinical physician to a secure