

Leiomyoma of the extrapleural chest wall: an atypical location

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Abstract

Tumours of the thoracic wall constitute a wide histological spectrum. However, the literature includes very few reports of a leiomyoma occurring at the extrapleural chest wall. In this report we present our experience together with a review of the literature. Our patient was a 33-year-old woman who was diagnosed with leiomyoma of the chest wall and treated accordingly and successfully. To the best of our knowledge, this is the seventh report in the literature defining the clinical entity.

Key words: thorax, chest wall, pleura, leiomyoma, surgery.

Introduction

Leiomyomas are benign soft tissue tumours which take origin from the smooth muscles. They are classified in the mesenchymal tumour family [1]. The first definition of the tumour was made by Virchow [2] in 1854 and Kloepper *et al.* [3] described the hereditary form which presented with multiple leiomyomas [4, 5]. Although the multiple form has been defined in general, these neoplasms are regarded as benign curable tumours.

Leiomyomas may occur in any body parts where smooth muscles are present. According to the location of the tumour, there are three distinct types of leiomyomas: piloleiomyomas, angioleiomyomas, and genital leiomyomas [4-9]. On the other hand, when the name is announced, the recall indicates that they are located in the uterus or gastrointestinal tract, being more frequent in the small intestine and oesophagus [1]. Additionally, there are reports of leiomyomas defining the origin even from the arrector pili muscle of the pilosebaceous unit [9].

The thorax is a very atypical region for the tumour origin. Leiomyoma originating in the thoracic cavity is an extremely rare clinical entity and the literature includes only sporadic reports describing leiomyomas of the extrapleural chest wall [1, 10-16].

The purpose of this manuscript is to present our single case experience on the clinical entity and make a review of the literature about the pathology described up to the present moment.

Case report

A 33-year-old multiparous female patient without any previous history of systemic and familial disorders was referred to the clinic with chest

pain and heartburn for 2 years. She had multiple family doctor visits and was treated due to pyrosis. Increase in the symptoms and the pain of the patient led to her referral to our unit. She complained of an increasing pain at the left upper border of the sternum irrespective of exercise. Physical examination was completely normal with normal temperature (36.7°C), breath sounds (rate: 14/min), heart rate (68 beats/min) and blood pressure (110/60 mmHg). Laboratory findings did not reveal any abnormalities except hyperlipidaemia (total cholesterol: 253 mg/dl; N: 125-200 mg/dl). Chest X-ray showed a lesion at the left upper hemithorax (Figure 1). Pathology was confirmed with computed tomography and it revealed a well-circumscribed solitary lesion with a wide base located on the parietal pleura of the upper left chest wall which also led to a certain degree of destruction on the second rib (Figure 2). Bronchoscopy was normal and a bronchial lavage was reported to be benign.

An explorative diagnostic and therapeutic strategy was planned and after the consent of the patient was obtained, surgical treatment was performed through left anterior thoracotomy. There was an encapsulated mass lesion measuring 5.3 × 4.0 × 3.4 cm originating from the apex of the chest wall. On inspection there was low grade vascularization of the mass. The tumour was excised totally (Figure 3) by abrading from the rib. The second rib was healthy and was left in place. Frozen section study of the specimen indicated a benign lesion. Histopathological examination revealed strong smooth muscle actin positivity, probably caused by a leiomyoma (Figures 4-6). The postoperative course was uneventful and the patient was discharged from the hospital on the fourth postoperative day. She visits our outpatient clinic regularly and is symptom free 14 months postoperatively.

Discussion

Leiomyomas are soft tissue tumours originating from smooth muscles. They were first described by Virchow in 1854 [2]. They can develop wherever smooth muscles are present; however, they usually occur in the uterus, small intestine or oesophagus [1, 4-9]. Although theoretically the latter is correct, smooth muscle neoplasms originating from the serous membranes are extremely rare and reports of intrathoracic leiomyomas exist only in a few case report studies [10-16] rather than large series in the literature.

When they are located in other than well-known locations, i.e. oesophagus, small intestine or uterus, generally they are symptom free and detected by chance while performing routine check-up measures or diagnostic studies for other pathologies [4-9]. Among the 6 published cases [10-16],

5 were asymptomatic [10, 12, 13, 15, 16], one had chest pain [11], and one had chest pain and empyema [14]. In our case the patient complained of pain and heartburn. There appear atypical, unfamiliar lesions on radiographic films which cannot be differentiated from other soft tissue

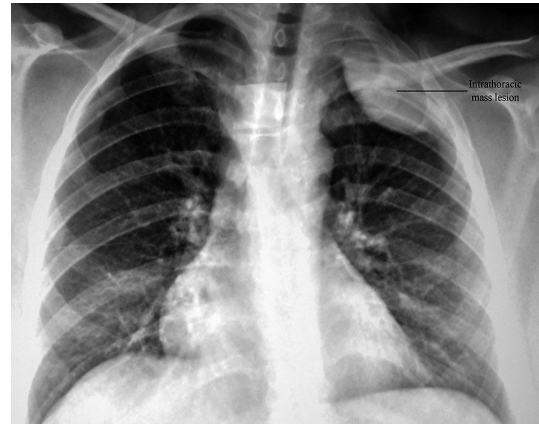


Figure 1. Chest X-ray indicating the lesion at the left upper hemithorax

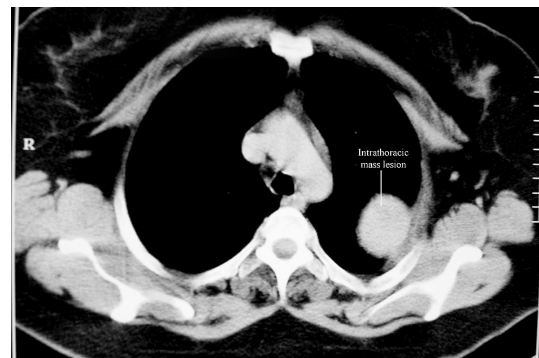


Figure 2. Computed tomography indicating a well-circumscribed solitary lesion with a wide base located on the parietal pleura of the upper left chest wall and leading to a certain degree of destruction on the second rib



Figure 3. The excised mass lesion measuring 5.3 × 4.0 × 3.4 cm

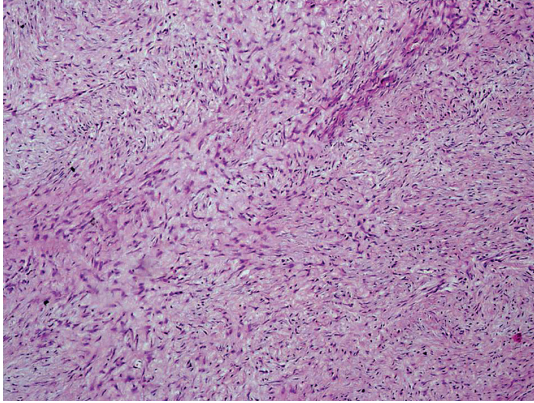


Figure 4. Haematoxylin and eosin staining indicating proliferating spindle cells with cigar-shaped nuclei and multiple vascular channels (×100, Olympus DP2BSW)

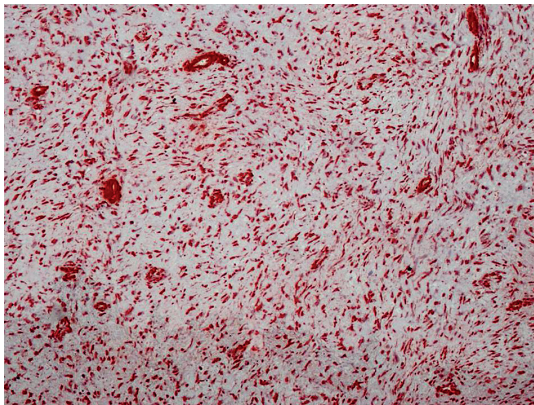


Figure 5. Strong smooth muscle actin positivity detected by anti-SMA immunostaining (×100, Olympus DP2BSW)

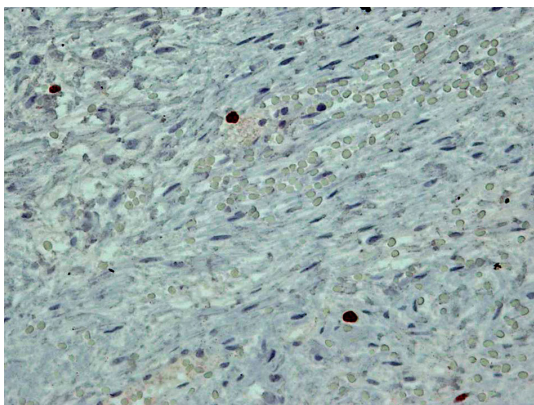


Figure 6. Immunohistochemical staining for Ki-67 indicated that the tumour had a low score, which indicated a benign lesion (×400, Olympus DP2BSW)

tumours of the chest [1, 10, 11]. Histopathological examination is required for the definitive diagnosis.

Leiomyomas are classified according to the origin of the tumour. Accordingly, there are three distinct

types of leiomyomas: pileleiomyomas, indicating the origin of the tumour from the arrector pili muscle of the pilosebaceous unit; angioleiomyomas, originating from smooth muscle – the tunica media layer within the arterial and/or venous wall; and genital leiomyomas, showing a wide range of region of origin such as muscle of the scrotum, the labia majora, and erectile muscle of the nipple [17]. It is believed that tumours in each classification have unique clinical and histopathological characteristics [2, 3].

The literature contains 6 published cases of leiomyomas originating from the chest wall [2-8]. Nose *et al.* [10], Turhan *et al.* [11], Proca *et al.* [12], and Tanaka *et al.* [13] presented their single case experiences, whereas Moran *et al.* [14, 15] presented two sporadic cases of leiomyomas of the chest wall. Tumours including our case originated from the vascular smooth muscle in four cases [11, 14, 15] and from the microvascular wall in two cases [10, 13], whereas Proca *et al.* [12] did not define the origin of the tumour exactly in their manuscript. Usually the tumours were solitary and could be resected completely [10-12]; however, due to the extent of the disease, Moran *et al.* [14,15] reported that complete resection was not possible. Being completely or partially resected, the longest follow-up has been provided by Turhan *et al.* [11] with up to 53 months. Like the other authors [10-16], they indicated cure with complete resection [10-12]. On the other hand, for the two cases of Moran *et al.* [14, 15], volume reduction surgery has also been effective and their 4-6 months follow-up results with partial resection did not indicate malignant transformation, metastasis or enlargement in the tumour size.

Metastasis of uterine leiomyomas years after hysterectomy or myomectomy is a well-known issue and the metastatic lesions of such tumours are named as benign metastasizing leiomyomas [18]. Regions of metastasis may include lungs, lymph nodes, bones, central nervous system and heart. These locations are shown to be associated with the presence of oestrogen and progesterone receptors and hormonal therapy has been shown to be effective for the treatment of benign metastasizing leiomyomas [11, 17]. Like the other solitary leiomyomas reported in the literature [10-16], our case did not have a history of uterine leiomyoma.

Although literally gender preference of the tumour has not been documented for extra-uterine leiomyomas, when we look at the literature for intrathoracic leiomyomas, up to the present moment, among the presented seven cases including our patient, six of them were females [10, 11, 13-16], which may indicate a female predominance and additionally support the oestrogen

Table I. Characteristics of intrathoracic leiomyomas reported to date

Case	Author	Age/gender	Tumour origin	Tumour size [cm]	Procedure	Follow-up course
1	Turhan <i>et al.</i>	50 years/F	Vascular smooth muscle	4 × 4	Complete resection	Alive at 53 months w/o recurrence
2	Nose <i>et al.</i>	55 years/F	Microvascular wall	1.5 × 1.5	Complete resection	Alive at 26 months w/o recurrence
3	Tanaka <i>et al.</i>	40 years/F	Microvascular wall	3.5 × 3	Complete resection	Alive at 17 months w/o recurrence
4	Proca <i>et al.</i>	32 years/M	No details	First diagnosis: 3 × 5.2 Second diagnosis: 4.3 × 7 (intrathoracic portion) + 4 × 6 (extrathoracic portion)	Detected with needle biopsy and followed due to benign behaviour. Resected completely with part of the anterior chest wall and a segment of the third rib 4 years later	Alive at 12 months, w/o recurrence
5	Moran <i>et al.</i>	23 years/F	Vascular smooth muscles	10 × 9 × 5.5	Resected partially	Alive at 6 months w/o enlargement or metastasis of the tumour
6	Moran <i>et al.</i>	21 years/F	Vascular smooth muscles	Unresectably huge	Resected partially	Alive at 6 months w/o enlargement or metastasis of the tumour
7	Our case	33 years/F	Vascular smooth muscle	5.3 × 4 × 3.4	Resected completely	Alive at 14 months w/o recurrence

F – female, M – male, cm – centimetres, w/o – without

and progesterone receptor theory for atypically located leiomyomas.

Leiomyomas are classified as benign tumours although histopathologically they may include proliferating spindle cells with cigar-shaped nuclei. They usually do not contain mitotic figures or cellular dysplasia [10]. However, as in the cases of Proca *et al.* [12] and Moran *et al.* [14, 15], the tumour may exhibit local invasion characteristics which may complicate complete resection. Even after partial resection the course of the disease is still benign.

In conclusion, a leiomyoma originating from the extrapleural chest wall is an extremely rare clinical presentation and to the best of our knowledge, this is the seventh report in the literature presenting

a chest wall leiomyoma. According to the review of the literature with this modest extent, when located in atypical regions, these tumours are generally benign, and usually completely resectable. Complete resection generally results in cure, and partial resection for unresectably huge tumours significantly decreases the symptoms without long-term evidence of size enlargement of the remaining lesion.

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