**Review Article**

**Leiomyomas of Chest Wall**

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**Abstract**

The chest wall is a region where the primary and metastatic neoplasms are detected rarely. Many histological types have been reported; however, smooth muscle neoplasms originating from chest wall are extremely uncommon and only a few primary tumors have been reported. The leiomyoma is a benign smooth muscle tumor which is generally determined in the urinary tract and occasionally in the gastrointestinal tract within the chest cavity. The chest wall leiomyoma develops from the extrapleural connective tissue where lots of small vessels exist. Clinically, they can be asymptomatic and are usually detected incidentally; or might present with chest pain. They cannot be differentiated from other tumors of the pleura or the chest wall, radiologically. Most of the cases present with solitary pleural-based mass and the definite diagnosis can only be made by the histological examination. Surgery is essential to confirm histopathological type and it is curative treatment option. Resection of tumor can be performed via video-thoracoscopic surgery if tumor’s radiological properties are suitable. But, the data from the literature show that generally thoracotomy has been used as the main procedure. Furthermore, some of the cases had needed chest wall resection and reconstruction. Despite its known to have benign histological features; smooth muscle tumors of the pleura have a potential for malignant transformation. Therefore, a complete resection of chest wall and strict follow-up for all patients is strongly advised.

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**INTRODUCTION**

Tumors originating from the chest wall or adjacent soft tissues are less common than the other tumors in any part of the body [1-3]. Primary chest wall tumors are characterized as either bone tumors or soft tissue tumors [3] and many histological types of chest wall neoplasms have been reported [4].

The leiomyoma is a benign tumor that derives from smooth muscle fibers which is mainly encountered in genitourinary [5-7], occasionally in the gastrointestinal [5,8-10] and rarely in the respiratory tract [5,11,12]. It can originate from the esophagus, bronchi or vascular structures at the thoracic level; therefore, it may be localized in mediastinum, pulmonary parenchyma, thoracic wall, diaphragm and pleural cavity [13-15]. Mediastinal leiomyoma is the most frequent one and its basis is excepted as the esophagus, aorta or vena cava [16].

Leiomyoma of chest wall is extremely rare [5,17-26]. The data about these cases concentrate on pleural or extrapleural masses. The focus of this review is extrapleural and parietal pleura based ones. Because of the sporadic case reports, the diagnostic criteria for smooth muscle tumors of chest wall have been controversial. Yet, only thirteen cases have been published up to now (Table 1). Three of these cases were defined by Moran and Proca. They have documented a metastatic site or less likely a needle tract seeding in a well circumscribed tumor which was histologically bland; emphasizing that these neoplasms have a low but definite malignant transformation potential. The rest of the cases were defined by authors as benign chest wall leiomyomas.

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**ETIOLOGY**

Etiology is not revealed yet; but ten of thirteen patients were women and mean age was 37,53 (21-55). It seems the tumor is more frequently seen in young to middle-aged females. Considering the statistical distribution of incidences indicating a female predominance, it might be pronounced that the estrogen and progesterone receptor theory has a role in pathogenesis of atypically located leiomyoma. But in our case there were no estrogen and progesterone receptors (25) like the case of Qui et al. [23].

Primary chest wall leiomyoma is differentiated from benign metastasizing leiomyoma (BML) with the absence of uterine operation in medical history. BML is a well-known infrequent disease that is usually detected years after hysterectomy or myomectomy. The metastatic lesions of such tumors are named as benign metastasizing leiomyoma and the most affected organs are generally the lungs [27,28]. Regions of metastasis may include lymph nodes, bones, central nervous system and heart. These locations are shown to be associated with the presence of estrogen and progesterone receptors; furthermore, hormonal therapy has been shown to be effective for the treatment of benign metastasizing leiomyoma [20,29]. However, we might speculate that hormonal therapy may not be effective on primary chest wall leiomyoma on the ground that no estrogen or progesterone receptors were detected in this case. In order to illuminate the curative statement of this therapy, further studies are needed.

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Regarding the growth of angioleiomyoma, other probable theories might be aligned as include estrogen theory, traumatic theory, and congestive theory [30].

FEATURES OF TUMORS

Most of the cases have presented as solitary masses; and only one case had multiple chest wall tumors with two separate lesions in the same hemithorax (Figure 1a/b) [25].

Macroscopic appearances of the masses were different from each other. Mostly prescribed as solid tumor with smooth margins and shape covered with intact pleura but some of tumors can be cystic or had irregular margins and cut-surface [25].

They were described as pleural or extrapleural except one case which was placed in intraosseus-intracartilagenous area [25]. Dimensions were documented in 13 tumors of 12 patients, and mean size was 6.35 cm (1.5-18 cm).

RADIOLOGY

Chest radiography, computed tomography (CT), magnetic resonance imaging (MRI), and positron emission tomography (PET) can all been used to determine the features of chest wall tumors. When the cases are evaluated in terms of radiological features and the necessity of resection; the idea that they originate form intercostal muscles rather than pleura comes to mind. Chest radiography reveals calcification, ossification, or bone destruction as well as location and size but the information obtained is limited in detail. CT scan can reveal the characteristics such as the vascularity of a tumor when a radiocontrast is used; as well as a more detailed appraisal of extent, location, and composition of the mass [31].

Most of them were neoplasms with smooth, round margins, either isodense or hypodense to chest wall muscle. Many soft-tissue neoplasms share similar characteristics at cross-sectional imaging. Also they were solitary and well-circumscribed with benign appearance suggesting a diagnosis of solitary fibrous tumor or metastatic carcinoma rather than mesothelioma. Tumors can be detected as solid or cystic structures. On the other hand, irregular calcifications with sclerosis and remodeling of the underlying ribs [5] or multiple lesions [25] can be determined by thorax CT.

MRI is an important tool of assessment because of its superior tissue-resolving features and multiplanar image acquisition that allows accurate tissue characterization [31]. Only one leiomyoma was seen as intraosseus soft tissue mass on chest wall and MRI showed a lesion that destructed seventh rib and expanded to the subcutaneous tissue (Figure 2).

Three-dimensional imaging of MRI sections allowed evaluation of the extensiveness of inflammation caused by concomitant osteomyelitis (Figure 3) [25].

Traditionally, bone tumors that demonstrate extensive cortical destruction and extraosseous soft tissue mass formation have been categorized as malignant.

When a malignant bone tumor does not have any specific imaging features and demonstrates only the typical features of a malignant lesion, limiting the differential diagnosis becomes difficult.
Because of the rarity of these lesions, PET CT has not been formally established as a diagnostic tool in the evaluation of these lesions [31]. However, elevated standardized uptake value (SUV) level has been detected at intraosseous leiomyoma, the cause of which could be concomitant osteomyelitis [25].

**HISTOPATHOLOGY**

The leiomyoma is classified as piloleiomyoma and anjoleiomyoma. Piloleiomyoma refers to the origin of the tumor - arrector pili muscle of the pilosebaceous unit-. Genital leiomyoma shows a wide range of origin such as muscle of the scrotum, the labia majora, and erectile muscle of the nipple. Examination of routine hemotoxylin-eosin sections of a piloleiomyoma has revealed pink staining bundles of fibers interwoven in many directions, so that some fibers were cut crosswise, some longitudinally. Basophilic nuclei were noted in the cross-sectioned areas to be within the fibers. Small numbers of round cells, lymphocytes and histiocytes, were present in trabeculae traversing the tumor area. With Masson’s trichrome stain, the tumor tissue stained pink, confirming the muscular nature, in contrast to the blue stain of collagen (33).

Angioleiomyoma originates from smooth muscle-the tunica media layer within the arterial and/or venous wall based on its origin [29,32]. Histological examination demonstrates well circumscribed fascicles of mature smooth muscle cells surrounding vascular lumina, lined by normal appearing endothelium but with no elastic lamina present.

It is believed that tumors in each classification have unique clinical and histopathological characteristics [33,34]. Chest wall leiomyomas have been considered as angioliemiomyoma.

A differential diagnosis for pleural spindle cell neoplasms includes solitary fibrous tumor, leiomyoid variant of mesothelioma, sarcomatoid variant of mesothelioma, spindle cell carcinoma, thymoma and metastatic tumors [35-39].

Although the clinical course of leiomyoma of the chest wall is not always concordant with histological findings, histological examination is necessary for definitive diagnosis. Despite its benign histological appearance, leiomyoma has a low but definite malignant transformation potential [5,19,20,25].

Histological precise diagnosis can be obtained by x-ray guided transthoracic biopsy in the preoperative phase but the tumor may metastasize or disseminate through the needle tract years after a transthoracic fine-needle biopsy [5].

Oncologists may prefer computed tomography-guided transthoracic fine-needle aspiration (FNA) of chest wall tumors in order to obtain an accurate preoperative tissue diagnosis by histopathological examination [40,41]. In our opinion, if tumor is unresectable or the patient refuses surgery, FNA should be performed in order to allow pathologic diagnosis for further medical treatment. Otherwise, if the tumor is resectable, FNA should not be performed to avoid needle seeding [23,25].

A definitive diagnosis of leiomyoma is always histological and requires confirmation of the presence of smooth muscle fibers without a sign of malignancy (pleomorphism, mitotic figures, and poor differentiation) by hematoxylin and eosin (H&E) staining.

The histological features of H&E-stained tissue sections plus positive staining for SMA and desmin provide unambiguous evidence for the diagnosis of leiomyoma. Immunohistochemical staining should be positive for smooth muscle actin (SMA), vimentin, desmin, and HH35 soft muscle protein [5,41].

Electronic microscope technique may help to provide a precise diagnosis [41] and shows spindle cells with elongated nuclei displaying marginalization of chromat and indentation of the nuclear membrane. Parallel arrays of intracytoplasmic actin microfilaments with interspersed fusiform dense bodies, discontinuous external lamina, and occasional cell junctions were also seen. No microvilli, tonofilaments, or desmosomes were present [5].

**CLINICAL FEATURES**

Earlier publications stated that most patients were asymptomatic, but with increased number of case reports, chest pain dominancy was observed. Symptoms are non-specific, and are frequently related to the tumor size or its location, manifesting as pleuritic pain, cough or dyspnea. A physical examination usually reveals normal findings or there might be auscultatory hypophonosis if the size is considerable. Routine blood biochemistry does not provide anything specific for the diagnosis. Leiomyoma cannot be differentiated from other soft tissue tumors of chest wall, radiologically [20,21].

Sporadic cases were reported as intra-osseous, intracartilaginous localized leiomyoma (of mostly upper extremities) [42,43]. However, only one case has been reported in chest wall bone. As far as we have learned from this case; leiomyoma can be seen in intra-osseous area and mimic the symptoms of bone tumors. If tumor growth causes inflammation and osteomyelitis; symptoms of osteomyelitis may be seen such as local fever, tenderness, discoloration and endurance [25].

Chest wall leiomyoma may increase in dimension with local invasion to the mediastinum and it may not be possible to resect the mass completely [18].

Soft tissue leiomyoma of chest wall shows itself as atypical, unfamiliar lesions on X-Ray images which cannot be differentiated from other soft tissue tumors of the chest [4,19,20].

**TREATMENT**

Main goal of surgery is the complete resection of tumor. Leiomyoma can reach to large dimensions; it may cause serious symptoms or signs and transform to a malignant tumor [21,24]. Most tumors can be completely resected if the contrast-enhanced chest computed tomography scan shows a well-capsulated pleural-based mass with minimal invasion of the adjacent organs; and the patient does not have signs of invasion, such as very severe pain. Generally, most primary chest wall leiomyomas can be easily and completely resected. If the tumor is a small localised one and its origin is pleura, complete resection can be achieved by a minimally invasive surgical technique like video-assisted thoracic surgery [19]. In addition, the prognosis is fairly good in patients whose tumoral mass has completely been resected.

Tumor excision was made in six cases without chest wall resection as operational procedure and one of these was made...
via video-thoracoscopic resection in literature [19]. Two cases were reported as partially resectable because of adjacent skeletal muscle growing or involvement of diaphragm [18]. One case had been followed-up after the confirmation with benign needle biopsy findings. But the lesion disseminated in the adjacent tissue and chest skeletal muscles through the needle line after 4 year follow-up period; and needed en-bloc chest wall, skeletal muscle and adjacent lung tissue resection [5]. We resected two separate lesions in one case with surrounding pleura en-bloc (Figure 4). But local recurrences occurred at the same areas after 1 year follow-up period and chest wall resection was needed [25].

Tumor excision with surrounding pleura might be the adequate option if the source is pleural microvascular wall. However; if the tumor is originated from vessels of intercostal muscles, a wider resection including the chest wall is necessary. In my opinion, local recurrence may be seen in cases where chest wall resection has not been performed. Radiology can be useful in estimating the origin. Tumors which are widely based on chest wall might be originating from intercostal muscles [25].

Because of their aggressive behavior, chest wall resection should be added to the surgical procedure of these benign tumors to achieve complete resection. Also, to avoid local recurrence, chest wall resection may be more appropriate than the tumor excision only (Figure 5a/b/c) [25].

Each patient warrants individual evaluation for chest wall resection. Operative selection is based on the potential benefits, operative feasibility, patient health, and anticipated tumor biology. The choice of reconstruction techniques is based on the tumor’s location, the size of the remaining defect, and the availability of autogenous graft materials. Skeletal and soft tissue reconstruction can be performed safely in a single stage [5].

Chest wall reconstruction should be considered if more than two ribs are resected (Figure 6).

Typically, defects less than 4 to 5 cm or posterior defects covered by the scapula do not require reconstruction [30].

Diverse reconstruction techniques such as diaphragm reconstruction is needed after resection of left arcus costarium at intraosseous leiomyoma and we stitched it to chest wall.

Initially, we sutured the exposed rib ends to each other and to upper intact rib with steel wire stiches to provide the stabilization following arcus resection. We also sutured diaphragm to above of this new reconstructed arcus and used double layer prolene mesh to cover the arcus [25]. Same operation can be carried out by titanium plate and a mesh.

The ideal prosthetic graft material should be easily available, durable, easily usable, adaptable, rigid, resistant to infection, translucent to radiographs, and of low cost. Combination of a metal material and a mesh is an appropriate prosthesis, because of its durability, ease of use, adaptability, rigidity, and translucency to radiography [44].

Various prostheses have been used, with sufficient rigidity, such as sandwiched polypropylene mesh and stainless steel mesh [45], methyl methacrylate sandwiched between polypropylene mesh [46], titanium plate-supported methyl methacrylate resection.
sandwich [47], titanium plate with Gore-Tex® dual mesh [48], and Composix Mesh™ [49].

After surgery, strict follow-up of patients is needed since chest wall leiomyoma have a low but definite malignant transformation potential and recurrence risk (Figure 7). [18,20,25].

CONCLUSION

Despite its benign histological features; leiomyoma of chest wall have a low but definite malignant transformation potential. Therefore, tumor resection with surrounding tissue should be the first choice. On the other hand, it should be considered that tumor recurrence may occur in procedures which did not include chest wall resection. These tumors can grow into the ribs and reveal with the symptoms of bone malignancy. A close and strict follow-up of the patients is essential because of their malignant potential and recurrence risk.

REFERENCES
