ACTA SCIENTIFIC CLINICAL CASE REPORTS

Volume 5 Issue 2 February 2024

Case Report

Rare Intra-Thoracic Tumors with Aggressive Potential: Case Series and Literature Review

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DOI: 10.31080/ASCR.2024.05.0506

Received: November 08, 2023

Published: January 02, 2024

Abstract

Background: Intrathoracic tumors of aggressive potential are rare, especially when talking about extra axial intrathoracic chordomas and lipomas. Usually, chordomas more commonly present as axial skeleton tumors and lipomas more commonly involve the extremities and trunk. Due to their low incidence, only a few cases of intrathoracic tumors are reported to be either chordomas or lipomas.

Cases Presentation: In this case series, 2 cases were reported as rare mediastinal tumors. The first case, we report a 78-year-old female patient presenting with a large right superior mediastinal mass resulting in dyspnea and cough, found to be a chordoma on histopathology after surgical excision. The second case, a 76-year-old male patient presenting with dyspnea and pleuritic chest pain, found to have a large left pleural-based fat-containing tumor on imaging resulting in contralateral mediastinal deviation to the right with left pulmonary parenchymal volume loss, which turned out to be a myxolipoma on histopathology following complete surgical resection. Finally, a literature review is performed to showcase the prevalence of these tumors, epidemiology, radiological and histopathological findings as well as the appropriate management.

Conclusions: Intrathoracic tumors with aggressive potential are rare, especially chordomas and lipomas. Total excision is the treatment of choice, aiding in both diagnosis and symptom relief.

Keywords: Chordomas; Myxolipoma; Liposarcoma; Sternotomy

Abbreviations

CSU: Cardiac Surgery Unit; CT: Computed Tomography; ER: Emergency Room; MRI: Magnetic Resonance Imaging

Background

Intrathoracic tumors of aggressive potential are rare, especially when talking about extra axial intrathoracic chordomas and lipomas. Chordomas are rare low-grade slow-growing yet locally aggressive tumors of notochordal origin affecting around 1 per 1 000 000 people yearly, and extra-axial chordomas arising outside the axial skeleton are extremely rare. While lipomas are also slow-growing benign soft tissue tumors more commonly found subcutaneously. Thus intrathoracic lipomas are rarely identified. However these deep seated intrathoracic tumors couldhave a malignant potential, particularly the myxoid subtype. In contrast to other lipoma myxoid lipoma, subtypes, can transform into a liposarcoma.

In fact, such tumors are usually found incidentally on routine examination or present with symptoms depending on their exact intrathoracic location and their size. They most commonly cause local pain and symptoms related to invasion and/or compression of adjacent anatomic structures. Symptoms might include dyspnea, chest pain, cough or even hemoptisis yet only fewcases are identified so far hence the lack of deep clinical knowledge about the entity.

Furthermore, in order to identify and differentiate between different types imaging and histo-pathological studies are needed.

Only a few cases of these mediastinal tumors are reported in the literature. We report the first case of a 78-year-old female patient presenting with a right superior mediastinal mass resulting in dyspnea and cough, found to be a chordoma on histopathology after surgical excision. And thesecond case of a 76-year-old male patient presenting with dyspnea and pleuritic chest pain, foundto have a large left pleural-based fat-containing tumor on imaging, responsible for contralateral mediastinal deviation to the right, with left pulmonary parenchymal volume loss, which turnedout to be a myxolipoma on histopathology after complete surgical resection.

Finally, a literature review is performed to showcase the prevalence of these tumors, epidemiology, radiological and histopathological findings as well as their management.

Case Presentation 1

We present the case of a 78-year-old female patient known to have hypertension, dyslipidemia and hypothyroidism, presenting to the outpatient clinic with dyspnea and cough. Her history goes back to few months prior to her presentation when she started to develop shortness of breath and chest pain. She denied any palpitation, numbness or weakness in her upper limbs or dysphagia.

A magnetic resonance imaging of the chest performed showed a large well-defined right superiormediastinal mass seen extending superiorly through the thoracic inlet into the right lower neck, grossly measuring 7.4 x 6 cm in maximal axial dimension and spanning a craniocaudal length of 10.5 cm. It appears predominantly hyperintense on T2-weighted images showing internal T2 hypointense septa, some demonstrating nodular thickening, and predominantly hypointense on T1-weighted images showing few T1 hyperintense foci likely representing internal bleed. It is heterogeneously enhancing and showing areas of diffusion restriction. It is seen anteriorly displacing the superior vena cava, right common carotid artery and jugular vein and slightly displacing the upper trachea and esophagus to the left, with no evidence of invasion. It is seen incontact with the right aspect of the upper thoracic vertebral bodies, with no evidence of invasioninto the vertebral body, neural foramen or spinal canal (Figure 1).

The patient was then admitted to the hospital for a scheduled total surgical excision of the mass. Upon admission, she was mildly dyspneic and had a bulging mass seen at the right aspect of her neck. During operation, a mid upper mini-sternotomy was done.

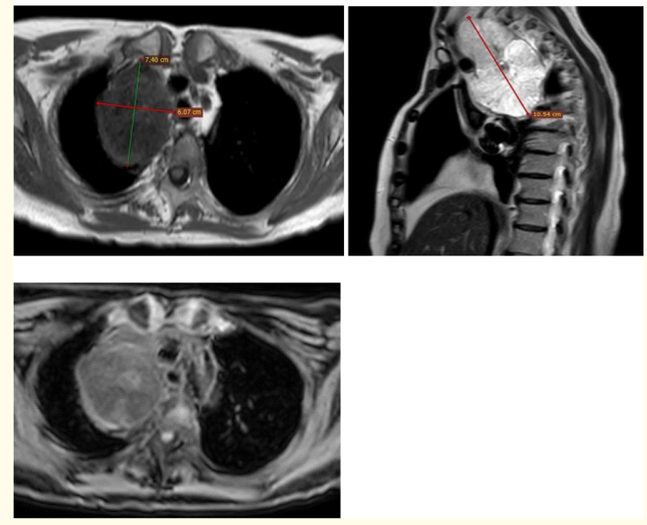

Figure 1: Axial T1-weighted, sagittal T2-weighted and axial T1 post-contrast images of the chest showing a 7.4 x 6 x 10.5 cm right superior mediastinal mass appearing predominantly hyperintense on T2 and hypointense on T1 with heterogeneous enhancement.

Evidence of a huge tumor in theretro-caval space with compression of the trachea, esophagus and the superior vena cava was identified which was responsible for the patient's preoperative clinical picture. An incisional biopsy was taken from the tumor and sent for frozen section examination, which revealed tumoral tissue with myxoid background and large polygonal eosinophilic cells with no evidenceof lymphomatous process. Thus, a decision was taken to proceed with a total resection of this tumor. Prolongation of the mini-sternotomy by a mini-thoracotomy at the 4th intercostal space and total resection of the tumor with capsulectomy was done. Evidence of mucinous tissue wasfound within the tumor. Hemostasis was adequately done followed by closure. The patient tolerated the procedure well and was transferred to the cardiac surgery unit (CSU) in a stable condition. She was discharged home a few days later.

Following up on the mass's histopathology results, it showed tumoral proliferation composed oflobules separated by fibro-inflammatory bands, in a myxoid background positive with Alcian blue stain. It is composed of cells forming short cords , sheets, nests, and sometimes single cells suggestive of signet-ring cells; the tumoral cells are epithelioid with abundant clear or eosinophilic cytoplasm, sometimes with a bubbly/vacuolated appearance (physaliphorous cells); the nuclei are heterogeneous frequently low-grade and occasionally high-grade with no evidence of frequent mitosis; and occasional necrotic areas were also seen. Tumoral cells were extensively positive for CK 1/AE 3, EMA, CD 56, and IDI 1, with Ki-67: 10 to 15% confirmingdiagnosis of conventional chordoma.

Case Presentation 2

We present the case of a 76-year-old male patient known to have hypertension and chronic kidney disease related to polycystic kidney disease (baseline creatinine 2), presenting to the emergency room (ER) for severe dyspnea and pleuritic chest pain. Upon presentation, the patientwas dyspneic with however normal oxygen saturation, conscious, cooperative and oriented, withdecreased air entry on the left lung. A non-enhanced computed tomography (CT) scan of the chest done showed an 18.2 x 13 x 10.6 cm (CC x AP x TR) heterogeneous lobulated mass occupying the left hemi-thorax, more likely pleural-based, showing intermixed fat and soft tissue components, with the solid component predominating at its lower aspect (Figure 2).

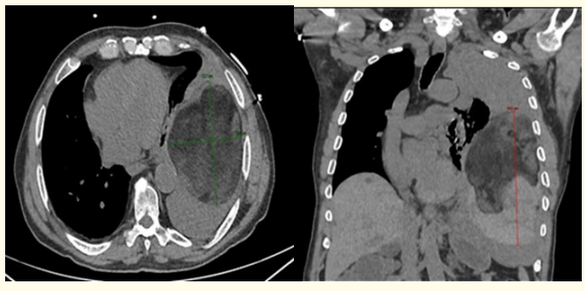

Figure 2: Axial and coronal non-enhanced CT scan of the chest showing an 18 cm pleural-based mass with fat and soft tissue components.

The subsequent mass effect causes a contralateral mediastinal deviation to the right and left pulmonary parenchymal volume loss with diffuse mosaic attenuation. There is an associatedlarge left-sided pleural effusion, of slightly high density suggestive of fluid turbidity.

Due to the CT scan findings and the clinical picture of the patient decision was taken to proceedwith operation. Thus surgery was done for a wide radical resection of his intra-thoracic tumor torelieve the mass effect on the mediastinum, re-expansion of the lung and control of the bleed.

During the operation, the patient was in the antero-lateral position. A left thoracotomy centeredat the 4th space was performed. Evidence of a huge intra-thoracic tumor showing chest wall invasion at the above-mentioned thoracotomy, with adhesions reaching the diaphragm and the pericardium, total collapse of the left lung and right-sided shifting of the mediastinum. During the procedure, decision was taken to go for a second thoracotomy at the level of the 6th space to facilitate the resection of the mass which was eventually completely evacuated from the thorax (Figure 3). Adequate hemostasis was performed, two chest tubes were placed and closure done.

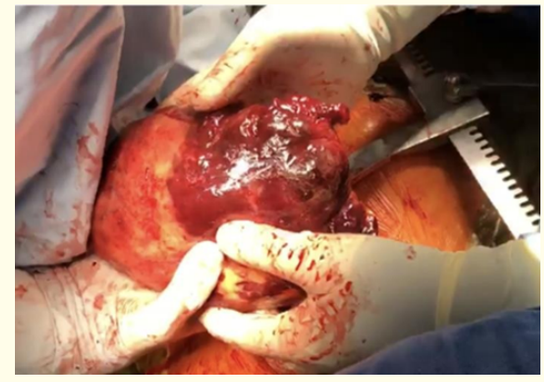

Figure 3: Resected intrathoracic mass.

The patient tolerated the procedure well, he was extubated post-operatively, was hemodynamically stable, off vasopressors and transferred to the cardiac surgery unit (CSU) ingood condition.

The chest tubes were removed on the first post-operative day and the patient was transferred to aregular floor hemodynamically stable, showing improvement in his hemoglobin level and electrolytes with the hematocrit level reaching 37. He was discharged home on the third post- operative day in a stable position.

Following up on the mass's histopathology, it was found to be a myxolipoma with clear surgical margins and no evidence of rib involvement. Immunohistochemical stains performedwith adequate controls were positive for Vimentin, s100, and CD34; PanCK (AE1/AE3), P16,BC12 and MDM2 were negative and Ki-67 was up to 1%.

Discussion

Intrathoracic tumors of aggressive potential are rare, especially when talking about extra axial intrathoracic chordomas and lipomas. Chordomas are rare low-grade slow-growing yet locally aggressive tumors of notochordal origin, affecting around 1 per 1 000 000 people yearly [1-3]. Theembryonic notochord normally regresses and only remains as the nucleus pulposis of the intervertebral discs [4]. Because of their notochord origin, they almost exclusively involve the axialskeleton extending from the clivus to the sacrum, including the vertebral bodies of the mobile spine, with the most common location being the sacrococcygeal region [1-3]. Extra-axial chordomas, also referred to as parachordoma and chordoma peripheric arising outside the axial skeleton are extremely rare, and review of the literature only revealed a few reported cases of extra-axial, namely mediastinal, chordomas [5-8]. These extra-axial chordomas are thought to either arise from ectopic notochordal remnants outside the vertebral bodies or the primary vertebral involvement is non-detectable [6].

Chordomas typically affect people between the ages of 40 and 60 years old with a male to femaleratio of 2:1, and are rarely seen in children and elderly [1]. Based on the 5th edition of the World Health Organization, chordomas are classified into three subtypes: conventional chordoma whichis the most common subtype, poorly differentiated and dedifferentiated chordomas which are much more aggressive subtypes [2,3]. The clinical presentation of chordomas depends on their location, and symptoms may take months to years to develop given the slow growing characteristic of these tumors [2]. The most common symptoms are local pain, neurologic dysfunction, and symptoms related to invasion/compression of adjacent anatomic structures [2].

Computed tomography (CT) and magnetic resonance imaging (MRI) are the modalities of choicefor the evaluation of chordoma, allowing the assessment of location, size and characteristics of the tumor as well as the assessment of nearby structures [2,6]. Tumors involving the axial skeleton present as lytic lesions that may contain calcifications in 30 to 90% of cases and may show extraosseous extension [2]. On MRI, these tumors appear hyperintense on T2-weighted images and iso to hypointense on T1-weighted images owing to the presence of calcifications and/or hemorrhage, showing moderate to avid enhancement on post-contrast sequences depending on the degree of necrosis/hemorrhage [2,7]. On the other hand, extra-axial chordomas appear as well- defined soft tissue lesions with less bony involvement when compared to their intra-osseous counterpart [7].

The differential diagnosis of mediastinal chordoma includes neurogenic tumors, neurenterciccysts, lymphadenopathy and extra-medullary hematopoiesis among others [5,9].

An extra-axial chordoma has similar macroscopic and microscopic appearance as the axial counterpart [8]. Macroscopically, it appears as a white to brown lobulated mass showing a fibrous pseudo-capsule and gelatinous consistency [8,10,11]. Microscopically, it appears as multiple lobulesseparated by fibrous septa, with tumor cells arranged in short cords, nests or singly, showing an abundant eosinophilic cytoplasm [8,10,11]. It stains positive for cytokeratin, epithelial membrane antigen, vimentin, brachyury and S-100 [2,8]. When combined, brachyury and cytokeratin are highly sensitive and specific for chordoma [2].

However, for lipomas are slow growing benign soft tissue tumors [12]. Such neoplasms arise frommesenchymal cells and are considered the most common mesenchymal tumors [12]. As such, one in every thousand people will develop one approximately; however, the exact incidence and prevalence of lipomas are not well indicated in the literature [12]. In addition, lipomas are slightlymore common in males than females, especially in the fourth to the sixth decade of life [12].

The most common locations of lipomas are the trunk and the extremities; however, these neoplasms could arise from adipocytes in any other atypical location [13]. One example of rare locations is intrathoracic, where these tumors can arise from the lung, mediastinum, thoracic walland are considered to be extremely rare [14]. Lung lipomas account for only 5-6% of lung tumors generally, especially that only 2% of lung neoplasms are benign and usually lung tumors are of malignant potential [15].

Few cases of intrathoracic lipomas were reported in the literature; such tumors can be located in the mediastinum, bronchial tree, pulmonary parenchyma and pleura [15,16]. Mediastinal lipomas areslow-growing tumors, the reason why patients remain asymptomatic until the tumor is large enough to compress adjacent structures [15]. Bronchial tree lipomas are those arising from the subcutaneous fat of any part of the bronchial tree including the trachea, and parenchymal lipomas are found in the periphery of the parenchyma in most cases [15]. Moreover, pleural lipomas extend from their original location of mesothelial parietal pleura to the sub-pleura and insome cases to extra-pleural locations [15].

Furthermore, different histological subtypes of lipomas were identified; angiolipomas, myelolipomas, fibrolipomas, angiomyolipoma, myelolipomas, ossifying lipoma, hibernomas,pleomorphic lipomas, chondroid lipomas, neural fibrolipomas and myxoid lipomas [16].

Radiologically, imaging of different lipomas subtypes could be similar and they are classicallyseen as well circumscribed tumors of subcutaneous fat on MRI and CT scans [17]. Sonography isthe imaging modality used to mark the interface between the lesion and surrounding fat [17]. As such, a high index of suspicion should be maintained to diagnose and treat such tumors mainly iffound in atypical locations.

To note, myxoid lipomas, as in our case, in contrast to other lipoma subtypes have potential totransform into liposarcomas [16]. Microscopic examination of myxoid lipomas show spindle shaped cells with scant cytoplasm and uniform nuclei in a myxoid background [16]. On the otherhand, liposarcoma shows areas of high vascularity with “chicken wire” pattern and lipoblasts [18,19].

As for immunohistochemistry, both myxoid liposarcoma and lipomas are usually negative for MDM2 and CKD4 [19,20]. But myxoid lipomas almost always stain positive for S100 where liposarcoma expresses variability. Hence the need for a combination of

radiological, histologicaland immunological findings for differentiation.

The treatment of choice of such tumors should be surgical excision of the whole mass especiallyin symptomatic patients, taking into consideration that excision will be both diagnostic and therapeutic [14]. To note, excision will help confirm the histopathological characteristics of the mass, relieving the symptoms and the compression on adjacent structures [14]. Adjuvant radiotherapy is sometimes recommended to improve local control of residual disease in chordomas cases, improving disease-free and local recurrence-free survivals [2,3,8,21].

Conclusions

Intrathoracic tumors with aggressive potential are rare, especially chordomas and lipomas. Imaging is helpful in identifying tumor location, characteristics and effect on adjacent structures. Definitive diagnosis is usually made by histopathology and immunohistochemistry following surgical resection, with total excision being the treatment of choice, aiding in both diagnosis and symptom relief.

Ethical Approval

The work presented in this article goes in accordance with the Declaration of Helsinki in 1964.

Consent

Patients' written consent were taken prior to reporting their cases.

Availability of Data and Material

References used are attached in the manuscript.

Competing Interests

This article has not been presented in any national or international meeting. The authors have no conflicts of interest or disclosures to declare.

Funding

None.

Authors' Contributions

The manuscript has been read and approved by all named authors and there are no other persons who satisfied the criteria for authorship but are not listed. The order of authors listed in the manuscript has been approved by all of us.

Acknowledgements

None.

Principal Investigator

Sarkis Ejbeh.

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