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Case Study

Desmoid tumors of the pleura: a clinicopathologic mimic of localized fibrous tumor

Healthcare

Keywords: Desmoidal tumor ,complete resection,Intrathoracic.

Dr. Fadil Gradica		**University Hospital "Shefqet Ndroqi",Thoracic Surgery Service, Tirana, Albania.**
Dr. Flora Gradica		**Pharmacy Service Tirana Albania**
Dr. Lutfi Lisha		**University Hospital "Shefqet Ndroqi",Thoracic Surgery Service, Tirana, Albania.**
Dr. Fahri Kokici		**Anestesi-reanimation Service**
Dr. Alma Cami		**Anestesi-reanimation Service**
Dr. Dhimitraq Argjiri		**Pneumology Department**
Dr. Rinard Kertoci		**Anestesi-reanimation Service**
Dr. Ylber Vata		**Visceral Surgery Department**
Dr. Leon Shpataraku		**Visceral Surgery Department**
Dr. Eliana Shima		**Anestesi-reanimation Service**
Prof. Perlat Kapisyzi		**Pneumology Department**

Abstract

Introduiction: Desmoid tumor is an aggressive form of fibromatosis of musculoaponeurotic origin. It is a histologically benign, slow growing tumor commonly presenting in the age group of 15-60 years. Desmoid tumor is a rare, benign soft tissue tumor having potential for local invasion. It commonly arises in abdominal wall, presenting as a palpable mass. We describe a case of thoracic desmoid tumor in a middle aged male arising from the chest wall.**Objective**:Intrathoracic desmoid tumors of the pleura are unusual tumors that are often clinically and histologically confused with localized fibrous tumor of the pleura or benign neurogenic tumors. **Matherial and Methods:**We studied three cases of intrathoracic desmoid tumor of the pleura and reviewed the clinical, histopathologic, and immunohistochemical features of the thre patients. Three women, ranging in age from 30 to 40 years (mean, 35 yr) comprised the study group. Treatment included complete resection (two cases)by antrolateral thoracotomy in one case and by VATS in another one , subtotal resection (one case), followed by radiation therapy.**Results**: Follow-up to date shows stable residual disease at 12 months (one case) was done reintervent also expected again local recidive and recomandation radiotherapy treatment and two patients with no evidence of disease at 12 and 36 months, respectively.
Conclusions: Desmoid tumor should be considered in the differential of localized fibrous tumor of the pleura.

Introduction: Desmoid tumor is an aggressive form of fibromatosis of musculoaponeurotic origin. It is a histologically benign, slow growing tumor commonly presenting in the age group of 15-60 years. Apart from abdominal sites, it may also arise from the chest wall,[3] shoulder, foot, thigh, or calf.[4] Despite being indolent in nature it has characteristic tendency for local infiltration causing pressure effects on surrounding organs.

Objective:Intrathoracic desmoid tumors of the pleura are unusual tumors that are often clinically and histologically confused with localized fibrous tumor of the pleura or benign neurogenic tumors.

Matherial and Methods

We studied three cases of intrathoracic desmoid tumor of the pleura and reviewed the clinical, histopathologic, and immunohistochemical features of the thre patients. Three women, ranging in age from 30 to 40 years (mean, 35 yr) comprised the study group. Three patients presented with chest pain and one with shortness of breath. Three of the lesions were based in the parietal pleura and one case lesion invasion of chest wall left side coste II-III-rd and one in the visceral pleura also sternocostal I-II-en and clavicula invasion right site also on left site only pleural parital invasion on case. Treatment included complete resection (two cases)by

antrolateral thoracotomy in one case and by VATS in another one , subtotal resection (one case), followed by radiation therapy.

Results :The mean tumor size was 12.5 cm, varying 1.5-18.5 cm , and all of the tumors exhibited a bosselated, firm, white, cut surface. The histologic features of intrathoracic desmoid tumors were similar to those of desmoid tumors at more conventional sites. Infiltration of the adjacent fat and skeletal muscle was in two cases present. The tumor cells were immunoreactive for vimentin, desmin, smooth muscle actin, and muscle-specific actin in three cases and were negative for S-100 protein. Follow-up to date shows stable residual disease at 12 months (one case) was done reintervent also expected again local recidive and recomandation radiotherapy treatment and two patients with no evidence of disease at 12 and 36 months, respectively. One patient after 36 mounths had severe relaxation left hemidiafragma et respiratore insuficience.Reintervent doublication hemidiaphragama sinister .

Conclusions:Intrathoracic desmoid tumors often exhibit clinical and radiographic features similar to localized fibrous tumor of the pleura. They generally have histologic and behavioral characteristics identical to those of desmoid tumors at conventional sites. Like desmoid tumors elsewhere, complete resection with negative margins is vital to prevent local recurrence. Desmoid tumor should be considered in the differential of localized fibrous tumor of the pleura.

Case 1. Patient A ,female ,

Her birthday 1950 ,clinically asymptomatic.Ches x –ray routine appeared lesion about 1.5 cm subpleural left hemithorax subcostal ¾ also on CT scaner.

VATS pleural tumorectomy right hemithorax.

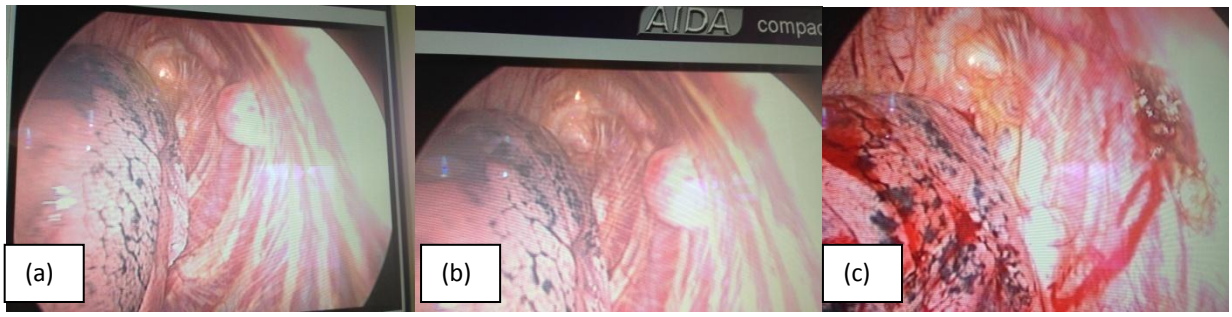

Figure 1,**VATS dexter tumorectomy dexter intercostal III-IV-t. Fig.(a),(b) before excision. Fig.(c) after excision**

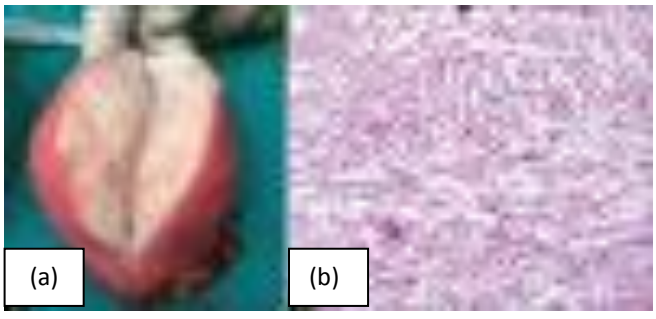

Figure 2 Excised tumor mass with its cut surface. (b) Histopathology section showing spindle cells with bland nuclei and abundant extracellular collagen in the stroma (H and E, ×200).

Case 2.Patient B,Female

Her birthday 1983 .Ches pain .short breathing ,X-ray and Ct scanner appeared pleural apical mass right hemithorax ,costal space II-III-rd ,and chest wall invasion.On 2002 en block resection ,2004 local recidive reintervent ,also 2015 local recidive suspected observation.

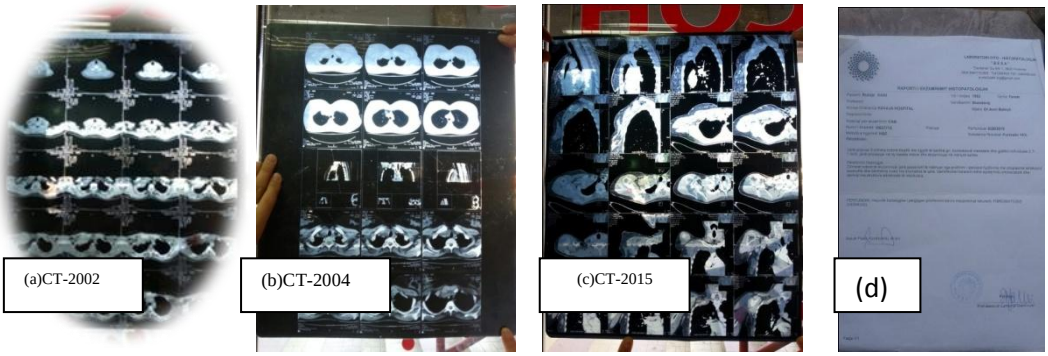

Fig. 3, (a,b**)** Chest wall tumor anterior superior hemithorax dexter. 2002,2004. (c).CT scanner 2015 apical anterior ches wall tumor right hemithorax .Local recarence. (d) Microscopy result Desmoidal tumor

Case 3.

Patent C .Her birthday 1981.She has ches pain ,short breath .X ray and CT scaner result tumor mass anterior chest hemithorax sinister with coste II-III-rd invasion .Intervent enbloc resection and chest wall defect reconstruction with propiletilen mesh 10 x 10 cm et musculocutano plastic.Biopsi result Desmoidal tumor.Als 5 –years after intervent patents had relaxation hemidiafralmal sinister et severe respiratory insuficience.We did reintervent ,left hemidiafragmal doublication ,know she has gut status.

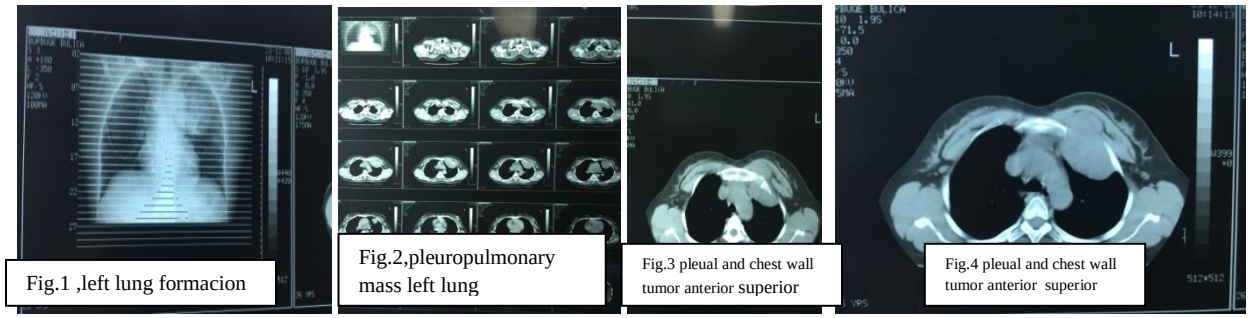
Fig.1 ,left lung formacion

Fig.2,pleuropulmonary mass left lung

Fig.3 pleual and chest wall tumor anterior superior

Fig.4 pleual and chest wall tumor anterior superior

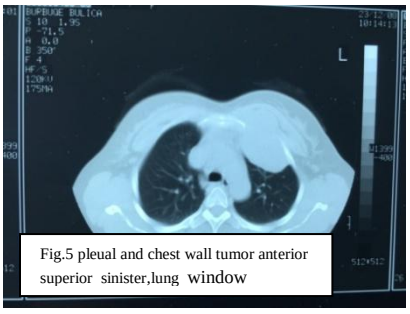
Fig.5 pleual and chest wall tumor anterior superior sinister,lung window

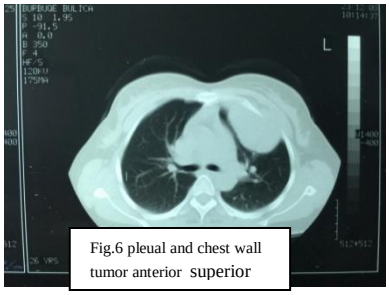
Fig.6 pleual and chest wall tumor anterior superior


Fig.7,(p-a)chest x-ray,after radical

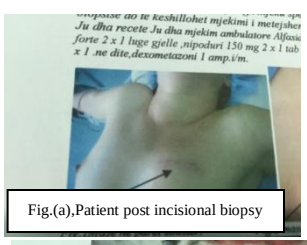

Fig.(a),Patient post incisional biopsy

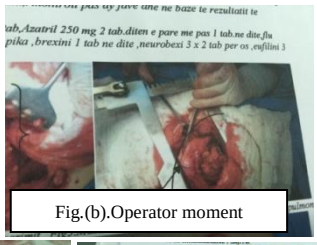

Fig.(b).Operator moment

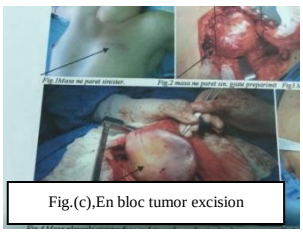

Fig.(c),En bloc tumor excision

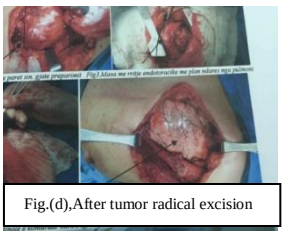

Fig.(d),After tumor radical excision

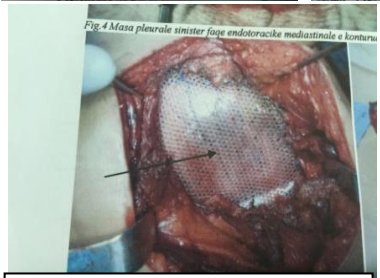

Fig.(e),Meshe chest wall defect reconstruction

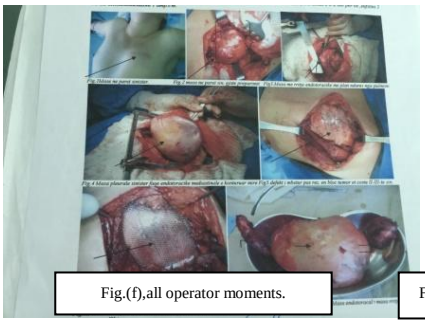

Fig.(f),all operator moments.

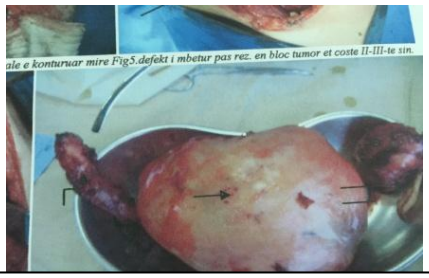

Fig.(g),Specimen ,tumor mas and sternocostal resection Coste II-III-rd.sinister hemithorax

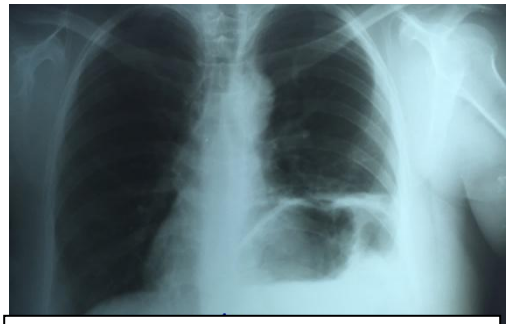

Fig.(h),Chest x-ray Relaxatio hemidiafragmal sinister 5-years after chest wall resection

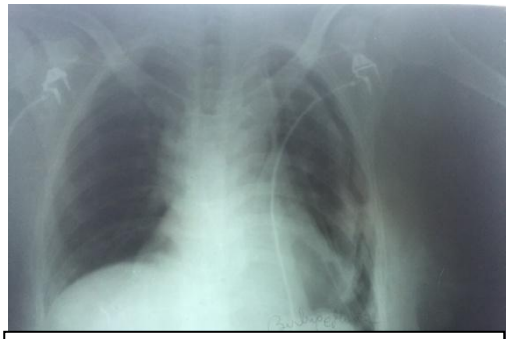

Fig.(i),Chest x-ray After hemidiafragmal doublication sinister .

Discussion

Desmoid tumor is an aggressive form of fibromatosis of musculoaponeurotic origin. It is a histologically benign, slow growing tumor commonly presenting in the age group of 15-60 years. Apart from abdominal sites, it may also arise from the chest wall,[3] shoulder, foot, thigh, or calf.[4] Despite being indolent in nature it has characteristic tendency for local infiltration causing pressure effects on surrounding organs.

Desmoid tumor usually presents as painless palpable mass but, less commonly, may project inwards and is detectable only on imaging.[8] Pain may occur due to nerve involvement giving rise to sensory and motor symptoms in the distribution of the nerve.[8]

Definitive diagnosis of desmoid requires histopathological examination. Fine needle aspiration may not be useful due to hypocellularity of the tumor.[3] Intrathoracically located desmoid may sometimes pose a diagnostic dilemma. Important differentials to be looked for in such cases include neurofibromas, fibrosarcomas, mesothelioma, ganglioneuromas, lung cancers, calcifying fibrous pseudotumors, and localized fibrous tumors of the pleura.[9] Complete surgical excision with wide tumor-free margins is the treatment of choice. Despite complete excision, chest wall desmoid is known for its high rate of recurrence,[6] though exact rate is not known in view of its rarity.

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