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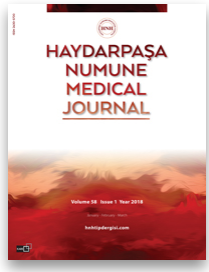


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CASE REPORT

# Intrathoracic Giant Desmoid Tumor: A Case Report

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**Abstract**

Desmoid tumors are uncommon, histologically benign tumors, originating in the facial and musculoaponeurotic tissues; however, they are locally aggressive. They are frequently observed in the abdominal zone. Nevertheless, it is rare in the chest wall. The most efficient and outstanding treatment is surgery. Here we present the case of a 66-years-old male patient who was admitted with the shortness of breath and back pain in the past 4 months, and we learned that he had bypass surgery in 2010, and a lesion mass was found in the left lung in the chest X-ray. This lesion was detected in the upper lobe of the left lung, which was 135×98×74 mm in size at the thoracic computed tomography. A left thoracotomic incision was performed, and a 25×15×6 cm (1195 gr) mass was removed. In this case, the patient had no complications and recurrence in the postoperative period.

Keywords: Desmoid tumor; intrathoracic; surgical treatment.

Desmoid tumor is a rare fibromatous tumor of the chest wall, and a low-grade malignant sarcoma of the soft tissue [1]. Desmoid tumor is a proliferative disease of fibrous tissue, originating from the muscle and aponeuroses [2]. There is no local invasion and recurrence. They do not present with distant metastases. In addition, they form 0.3% of solid tumors, and 3.5% of fibrous tumors [3]. Desmoid tumors, which are frequently abdominal, also hold the chest wall by 10%–28% and are often holding the anterior wall in the thorax [4]. In this article, we present a case of intrathoracic giant desmoid tumor without intrathoracic pathology in the angiography after the bypass operation 4 months before.

## Case Report

A 66-years-old male patient who had been diagnosed with dyspnea and back pain for the past 4 months had a bypass history in another center in 2010 and smoked story was 40 packs/year cigarettes. The patient underwent angiography 4 months before, and no pathology was detected in the angiography. We found the decrease in the left breath sounds, and maturity was present with percussion.

A regular contoured opacity was observed on the chest X-ray, which was starting from the lung apex level, and filling two-thirds of the left hemithorax (Fig. 1). Thorax computed tomography (CT) was performed on the lesions detected

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on the chest X-ray, and a cystic lesion and accompanying pleural effusion were detected in the left-upper lobe of the lung 13×9×7 cm in size (Fig. 2). Because the patient had undergone angiography 4 months before and no lesions were detected in angiography, this lesion was thought to be a hematoma after angiography, and the patient was prepared for the operation. In the preoperative Respiratory Function Test (RFT) of the patient, the FEV1 was 1.68% 64. In October 2012, video-assisted thoracoscopic surgery was performed on the left side of the thorax, and no hematoma was observed, but a mass approximately 15 cm in length was observed. Thoracotomy was discontinued, the left posterolateral incision was inserted into the thorax from the fifth intercostal space, and a biopsy was performed, frozen/section was reported as a result of the mesenchymal tumor. An advanced examination was planned to determine the presence of distant metastases and vascular invasion in the patient, and the operation was terminated.

The drain was terminated when the patient's drainage on the 1st postoperative day reached approximately 100cc. The patient was discharged on the postoperative 7th day without any complications in the postoperative follow-up.

The pathology was reported as a desmoid tumor, the patient underwent from thoracic magnetic resonance imaging (MRI) in November. A thoracic MRI showed a mass lesion measuring 15×14.5×17.5 cm. Starting from the left lung apex to the inferior pulmonary vein, the lesion had a close proximity to the aortic arch and the right pulmonary artery and the descending aorta, but lumen flows and contour integrity of defined arterial structures preserved, reduced the aeration by compressing the left lung. The lesion was compressed to the left main bronchus, which narrowed the lower lobe bronchi at this level. The lesion in particularly posterior mediastinum was revealed to the correct paravertebral area (Fig. 3).

The operation was performed in our clinic, and the left posterolateral incision was inserted into the thorax from the fifth intercostal space. The mass adhered to the anterior and posterior wall of the chest, and the advanced adhesions were separated with the dissection. It could be seen that the lower lobe stuck behind the ascending aorta. After the dissection, the lower lobe was separated from the thoracic wall and mediastinum. The lesion was fully removed (Fig. 4). The lacerations over the lungs were repaired. Two drains were placed, and the operation was terminated. As a result of the pathological examination of the removed mass, a definitive diagnosis was reported as desmoid tumor (Fig. 5), the size of the lesion was 25×15×6 cm, and the measured weight was 1195 gr. On the postoperative 1st day, drainage was 450cc, so we followed the patient at the clinic. On the postoperative 2nd day, drainage was 150cc,

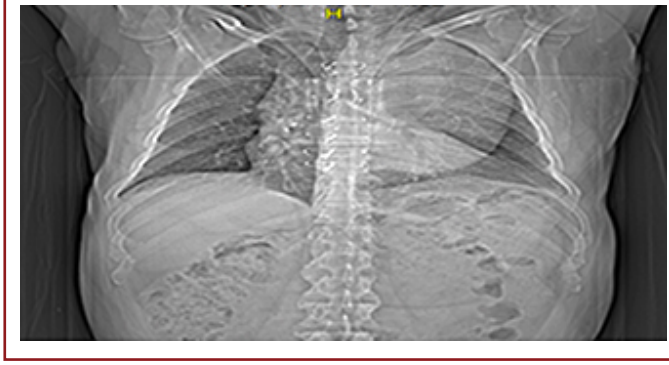

**Figure 1.** The patient's chest x-ray shows a mediastinal mass covering 2/3 of the left lung.

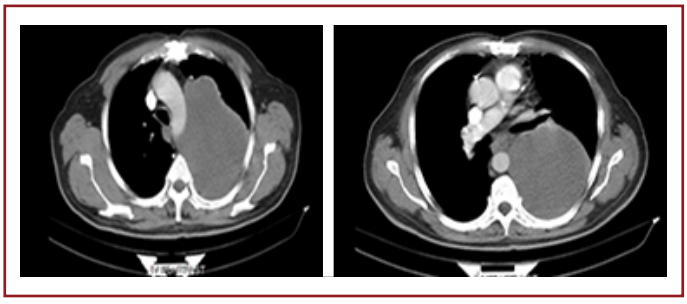

**Figure 2.** The thorax CT image of the Intrathoracic mass is shown.

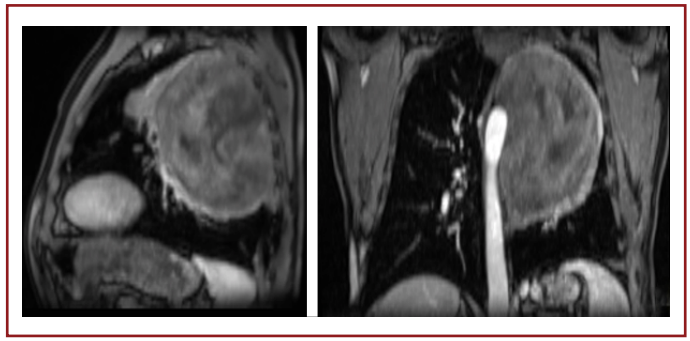

**Figure 3.** The thoracic MRI image of the Intrathoracic mass is shown.

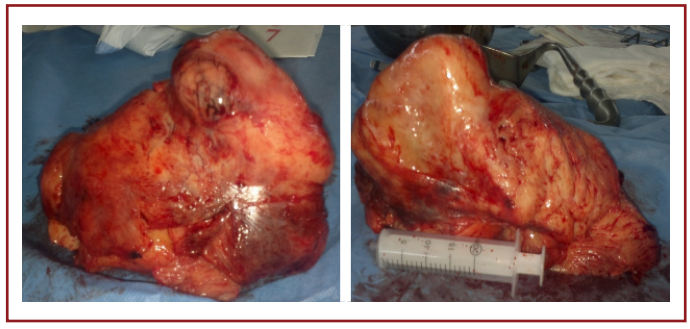

**Figure 4.** The desmoid tumor is seen as macroscopic.

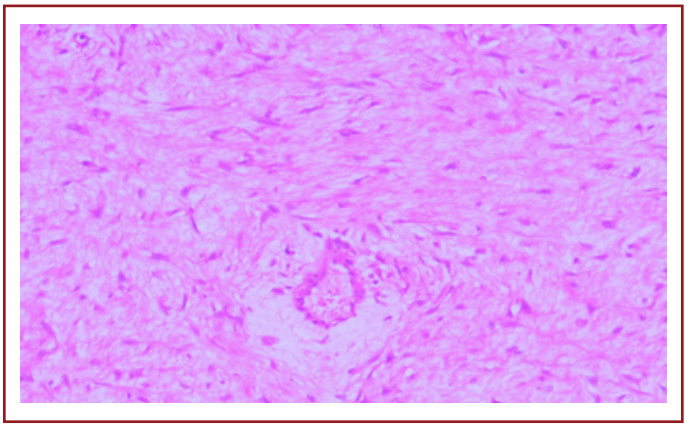

**Figure 5.** With the H×E (×10) the desmoid tumor consisting of needle cells in the mycoloid and fibrous stroma is observed.

so we terminated drain of the patient. At the chest X-ray, the lung was expanded, and we discharged the patient on the 3rd postoperative day. The patient is still followed without complications (Fig. 6) and in the follow-up, the FEV1 was 2.31% 78%.

## Discussion

Desmoid tumor is an aggressive tumor of the fibrous tissue, originating from musculature strikes, muscle aponeuroses, and connective tissue facies. Intrathoracic desmoid tumors are very rare, but they are the most common soft tissue sarcomas. They are evaluated by recurrence and local invasion after resection, but no distant metastases have been reported [2]. Pathogenesis is not clear, and trauma is the most important factor, in addition to the laceration, surgical incision and intramuscular injections are responsible [5]. Thoracic desmoid tumors are seen after mastectomy, breast silicone prosthesis implantation, coronary artery surgery, rib fractures, and thoracotomy. It is also thought to be due to endocrine causes. Estrogen leads to the growth of desmoid tumors [2]. There is evidence of genetic causes. We think that our case developed due to coronary artery surgery. The disease is more common at the age of 30, but our patient was older than this age group.

Desmoid tumor is manifested by pain. It makes frequent pressure [3]. It also causes motor and sensory complaints with the retention of nerves. An increased nerve involvement also increases the pain severity and prevalence [2,6]. In our case, there were complaints of progressive pain and shortness of breath.

Often, chest X-ray is used to detect the presence of a mass. Thoracic CT gives more details about the size and location of the mass. Thoracic MRI is more sensitive in detecting local recurrences and soft tissue infiltrations. The most common location is the anterior chest wall with 47%, posterior with 32%, and lateral wall with 11% [6]. The left hemithorax is more frequent than the right [3]. Intrathoracic desmoid tumors can reach large sizes, and 22% are larger than 10 cm [6]. Definitive diagnosis is made histopathologically. A fine needle aspiration biopsy may not be sufficient, because desmoid tumors are relatively hypocellular. Mitotic activity does not occur, and malignant criteria cannot be determined by cytology [3]. Diagnostic problems of intrathoracic desmoid tumors are more abundant than of abdominal desmoid tumors due to localization [2]. In differential diagnosis, neurofibromatosis, ganglioneuroma, fibrosarcoma, fibrous pseudotumor, local fibrous tumor of pleura, and lung cancer should be considered. Due to the large mass found in our case, we thought of lung cancer or a cancer of the posterior mediastinum.

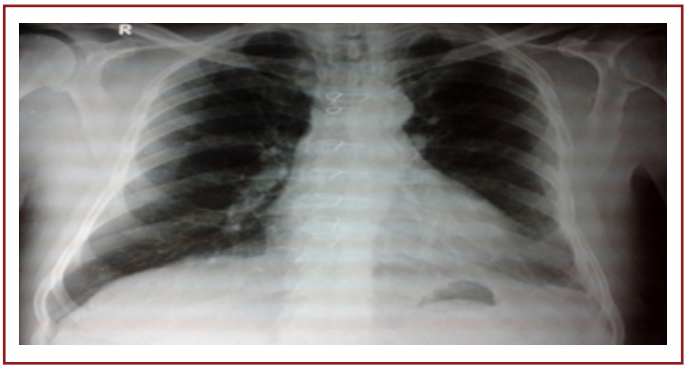

**Figure 6.** Recurrence or complications are not observed in the PA lung chart taken in postoperative 6 months.

Surgery is the treatment of choice. The negativity of the borders decreases the recurrence. Recurrence rate is lower in intrathoracic desmoid tumors compared to abdominal desmoid tumors (30%–60%). The recurrence is seen more frequently in the masses seen in patients over the age of 30 and larger than 10 cm. Because of the localization of intrathoracic desmoid tumors, a complete surgical removal may not always be possible [3]. After incomplete surgery, radiotherapy treatment options are available in large tumors, recurrent, and inoperable tumors [2]. They are in the series that offer different medicines. The c-AMP modulators (theophylline, chlorothiazide, ascorbic acid, testolactone), colchicine, estrogen blockers, prostaglandin [2,4] combined chemotherapy (cyclophosphamide+doxorubicin, mitomycin+doxorubicin and cisplatin, ifosfamide+etoposide) provide long-term benefits for desmoid tumors [1–4].

Consequently, although the desmoid tumors are not frequent, it should be kept in mind whether the patient has already undergone trauma and operation. The treatment should be surgical. The quality and quantity of the surgeon

also determine the later recurrences. If the surgical margin is positive, radiotherapy, chemotherapy, and other modalities are added to treatment. We wanted to show that the surgical approach may be less complicated than predicted radiologically, as in our case. In addition, it is possible to save the patient from the tumor burden and increase the respiratory parameters by surgery in such large masses.

**Informed Consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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## References

1. Okuno SH, Edmonson JH. Combination chemotherapy for desmoid tumors. Cancer 2003;97:1134–5. [CrossRef]
2. Iqbal M, Rossoff LJ, Kahn L, Lackner RP. Intrathoracic desmoid tumor mimicking primary lung neoplasm. Ann Thorac Surg 2001;71:1698–700. [CrossRef]
3. Kabiri EH, Al Aziz S, El Maslout A, Benosman A. Desmoid tumors of the chest wall. Eur J Cardiothorac Surg 2001;19:580–3.
4. Kostolny M Jr, Präuer H, Augustin N, Lange R. Extended resection of a chest wall desmoid tumour with concomitant coronary artery bypass grafting. Eur J Cardiothorac Surg 2001;20:1040–1. [CrossRef]
5. Allen PJ, Shriver CD. Desmoid tumors of the chest wall. Semin Thorac Cardiovasc Surg 1999;11:264–9. [CrossRef]
6. Brodsky JT, Gordon MS, Hajdu SI, Burt M. Desmoid tumors of the chest wall. A locally recurrent problem. J Thorac Cardiovasc Surg 1992;104:900–3.