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# It smells and tastes like cancer but it's not cancer: A case report of aggressive benign lung neoplasm

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ABSTRACT

Desmoid tumors are rare, locally aggressive benign tumors with a high rate of recurrence even after complete resection. Only twenty percent are intrathoracic and they are often painless or minimally painful with slow growth. Rarely they can grow quite rapidly, mimicking a malignant tumor. Here we present a rare case of a rapidly growing, intrathoracic desmoid tumor mimicking lung malignancy in an elderly male ex-smoker. This case demonstrates the importance of including desmoid tumors in the differential diagnosis of lung masses, raising awareness of benign tumors mimicking malignant behavior, and emphasizing the potential benefit of timely follow-up and early diagnosis.

## 1. Introduction

Desmoid tumors are rare and they account for approximately 0.03% of all neoplasms and fewer than three percent of all soft tissue tumors. The estimated incidence in the general population is two to four per million population per year [1]. Most desmoids arise sporadically, although between five and fifteen percent are associated with familial adenomatous polyposis [1,2]. Risk factors for sporadic development of desmoid tumors include previous surgical incision, pregnancy, hormonal exposure, and trauma [1,2]. Desmoids are benign but locally infiltrative and can be deadly via the destruction of adjacent vital structures and organs [1,3]. Herein we present an elderly male ex-smoker who presented with superior vena cava syndrome as a result of a rapidly growing, locally aggressive desmoid tumor. This case report illustrates the importance of considering this pathology early in the differential diagnosis of lung masses. It also emphasizes the need for timely follow-up and management. The invasive nature of the tumor can prevent life-saving resection if the diagnosis is delayed.

## 2. Case presentation

This is a 79-year-old male with a past medical history of hypertension and renal transplant who presented for evaluation of a three-week history of worsening right-sided facial pain and swelling, chest pain, and shortness of breath. The patient denied having similar symptoms in the past. He reported quitting smoking about fifty years ago. He endorsed a thirty-pound weight loss in the past three months without a significant change in appetite. Notably, while being worked up for pneumonia six months prior, the patient was incidentally found to have a three-centimeter focal pleural density in the right upper lateral lung on computed tomography with intravenous contrast, but he failed to follow up for a biopsy of the lesion.

On physical examination, he was hypertensive, there was erythema and swelling on the right side of the face, bilateral neck swelling right greater than the left, and decreased breath sounds over the right upper and mid lung. His brain natriuretic peptide was 330 pg per milliliter (<100). A chest X-ray revealed a right twelve-centimeter upper-to-mid hemithorax lesion with decreased aeration (Fig. 1). A subsequent chest computed tomography with angiography confirmed a twelve-centimeter right upper lobe mass with a small pleural effusion (Fig. 2). Pulmonary medicine and oncology were consulted. The patient was started on high-dose intravenous methylprednisolone with subsequent improvement of the symptoms. A needle core biopsy of the lung lesion was obtained and revealed spindle cells with wavy and fusiform nuclei with the expression of beta-catenin (Fig. 3); favoring desmoid fibromatosis.

The patient was discharged home with instructions to follow up with the cancer center for possible radiation therapy as he is a poor surgical candidate given the extent of the disease.

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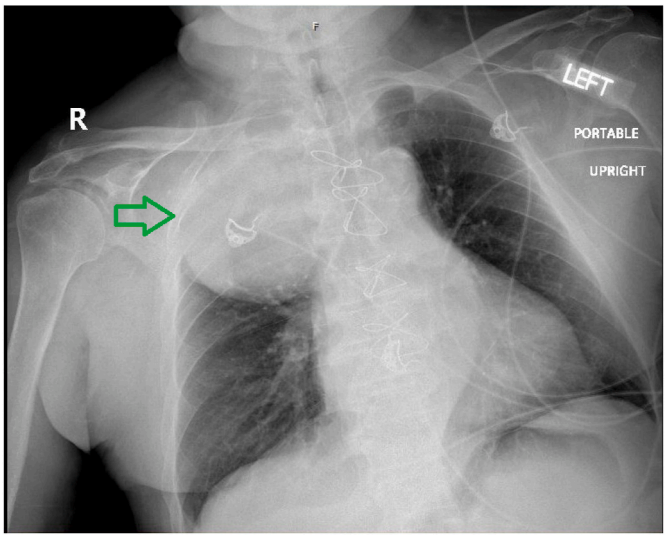


**Fig. 1.** Chest X-ray illustrating a large 12 × 10 cm right mid to upper hemithorax dense lesion (green arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

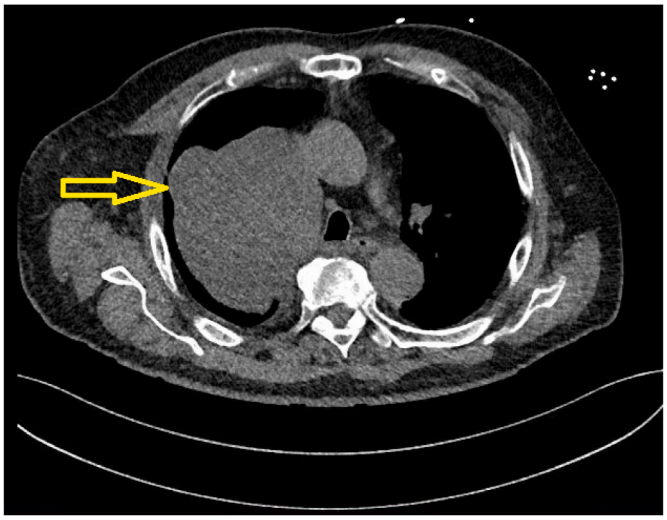

**Fig. 2.** Chest computed tomography with angiography showing a 12.0 × 9.4 cm x 9.4 cm right upper lobe mass (yellow arrow). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

## 3. Discussion

Desmoid tumors are rare locally aggressive tumors with no known potential for metastasis or dedifferentiation [1]. Tumor-related destruction of vital structures and/or organs can be fatal [1]. Rarely, a patient may present with constitutional symptoms, such as fever and/or weight loss [4]. Our patient suffered a 30-pound weight loss that could not be explained by another condition. He also presented emergently with superior vena cava syndrome (SVCS) symptoms that could only be explained by tumor-related compression of the vena cava. Although malignancy represents most SVCS cases, up to 40% of cases can have a benign etiology [5]. Glucocorticoids have only shown benefit in SVCS caused by mediastinal lymphoma masses [5]. However, our patient's symptoms improved with the use of methylprednisolone.

Desmoid tumors can have an unpredictable clinical course, but they tend to grow slowly in the majority of cases [1,3]. Our patient's mass was particularly unique because the mass grew four times larger than its initial size within six months; mimicking a malignancy. While magnetic resonance imaging is the preferred imaging modality for the evaluation of soft tissue masses of the trunk, the patient's inability to remain immobile made a computed tomography scan the more appropriate modality.

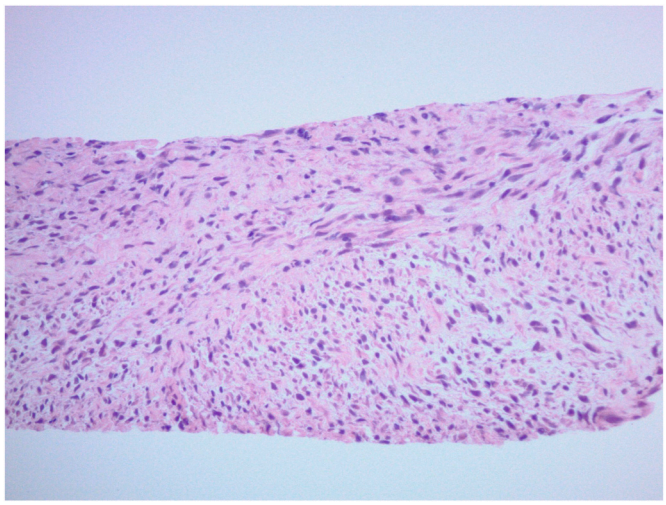

**Fig. 3.** 200x magnification Hematoxylin and Eosin stain of the tumor lesion composed of spindle cells without significant nuclear atypia, increased numbers of mitosis, and necrosis.

Given their lack of ability to metastasize, desmoid tumors can be controlled locally using surgery and radiation [3,6]. However, there is a significant risk of local recurrence, even after complete surgical resection [3,6]. There are no evidence-based or widely accepted guidelines for the management of unresectable desmoids [3], however, radiation therapy alone may be effective in these patients [6]. Despite the high recurrence rate, surgery remains the mainstay of treatment of desmoid tumors. Given the extension and the aggressive local invasion, our patient's tumor was unresectable. He was referred to our local cancer center where he will benefit from radiation treatment. If diagnosed earlier through proper patient follow-up for biopsy, our patient may have benefited from resection of the lesion. This case highlights the importance of counseling our patients on early biopsy and diagnosis while reiterating the need to follow the Fleischner Society Guidelines for incidental pulmonary nodules.

## 4. Conclusion

This case report of a desmoid tumor involving the lung contributes to the literature by raising awareness among clinicians of this benign, yet aggressive pathology. Given the unique rate of growth in this patient, the tumor may have been mistaken for an aggressive lung cancer. Through increased awareness, clinicians can further emphasize to patients how imperative it is to have close follow-up and early biopsy as per the Fleischner Society Guidelines. Through earlier diagnosis, patients would potentially have the opportunity to have the mass resected before the mass gets extensively larger. As seen in this case report, a delay in diagnosis can lead to fatal compression of the local structures.

## Declaration of competing interest

No potential conflicts of interest were reported by the authors.

## Acknowledgments

None.

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