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Case Report

Pleural desmoid tumor: A rare site of presentation☆,☆☆

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ABSTRACT

Desmoids are a rare type of tumor with an unpredictable natural history. The annual incidence in the general population is estimated at 2.4-4.3 cases per million. This rare case is considered a diagnostic challenge. Therefore, the emergence of knowledge of CT features and other findings is an important aspect in the diagnosis. In this case study, we present a 57-year-old female patient with a 2-year history of chest pain in the right upper chest. To establish a proper diagnosis a chest X-ray was performed and showed opacities in the right hemithorax, and computed tomography revealed a solid pleural mass measuring 4.4 cm × 4 cm × 3 cm. Along with imaging results, histopathological examination and immunohistochemical analysis of an open biopsy revealed a desmoid tumor.

Introduction

Desmoid tumors are rare mesenchymal neoplasms also known as deep or aggressive fibromatosis. While these tumors are locally aggressive, they do not metastasize to distant regions. The rate of recurrence for these tumors can reach up to 39%. The morbidity and mortality associated with desmoid tumors is related to local recurrence and the involvement of adjacent organs. These tumors have an annual prevalence of 2.4-4.3 cases per million in the general population, and account for less than 0.03% of all neoplasms. Females have a slightly larger preponderance than males, and the peak incidence occurs in the late third and fourth decades of life. The emergence of chest discomfort, dyspnea, and pleural effusion are all possible symptoms [1–3]. However, despite occult findings on imaging, it is uncommon for desmoid tumors to be detected. The radiologic findings are unique and different from those seen in other pleural tumors, and diagnosis relies on pathologic examination [4].

According to the WHO categorization, desmoid-type fibromatosis is a tumor of fibroblastic or myofibroblastic origin [5]. These tumors are also categorized by their location as intra-abdominal, extra-abdominal, or within the abdominal wall [7]. Intrapleural desmoids tend to remain asymptomatic for a long time and appear larger at presentation compared to other

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desmoid tumors. Intrapleural tumors do not cause symptoms until they expand to a size that creates discomfort from chest wall invasion or dyspnea from lung parenchyma compression. This is unlike superficial chest wall tumors, which cause a palpable mass [6].

Even though the etiology of these tumors is uncertain, some variables are associated with their development and growth [3], including trauma, hormonal changes, pregnancy, and genetic factors. The main aim in managing desmoid tumors concerns identifying potential causes of morbidity and mortality, and involves an assessment of the ability to grow and the tendency to recur [1].

Desmoid tumors have been described in variety anatomical locations, but the abdomen and abdominal wall are the most common sites of occurrence. The chest wall is the most common extra-abdominal site [6]. True primary pleural desmoid tumors, such as the one in the current case, are extremely uncommon.

Case report

A 57-year-old female presented with right chest pain that was present for the previous 2 years. The pain was initially located in her back and radiated to the ventral side of her right chest. The pain was inconsistent and became worse when the patient felt fatigued. The patient also complained of intermittent coughing. Therefore, physical and laboratory examinations were carried out, and the tumor markers CA-125, CA-19.9, and CEA were within normal limits. In addition, a chest radiograph was performed, and it showed that there was an opacity in the right upper hemithorax (Fig. 1). There was no trauma history or previous surgery. The patient was in menopause and had no family history of such tumors.

To pursue a proper diagnosis, a contrast chest computed tomography (CT) was performed and confirmed the presence of a pleural-based mass of approximately 4.4 cm × 4 cm × 3 cm with no evidence of rib destruction and a lack of enhancement (Figs. 2 and 3). An open of the mass revealed the presence of neutrophils, lymphocytes, and macrophages, and an absence malignant cells. The mass was finally resected using video-assisted thoracic surgery, and 2 months after surgery showed there was no residual mass (Fig. 4). A histopathological examination was conducted, which identified the mass as a spindle mesenchymal tumor (Fig. 5A). In addition, immunohistochemical were positive for vimentin and smooth muscle actin, which revealed a desmoid type of fibromatosis (Figs. 5B and C).

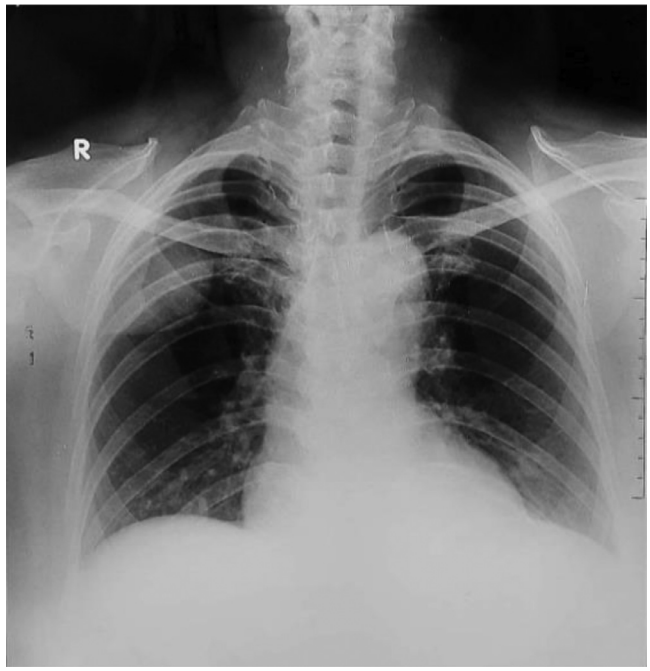

Fig. 1 – Chest radiograph showing an opacity in the right upper hemithorax.

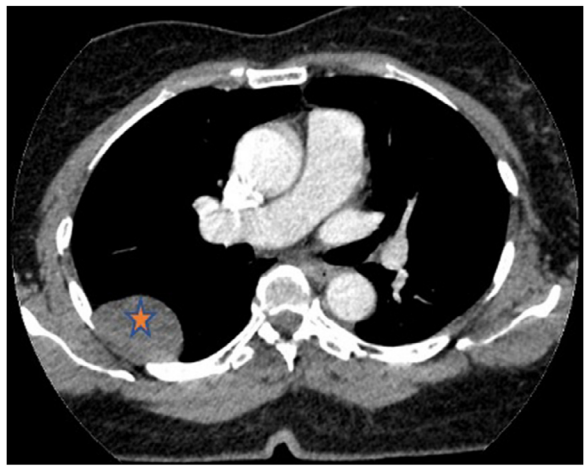

Fig. 2 – Axial view chest computed tomography with a mediastinal window and contrast injection showing a circumscribed, well-defined pleural-based mass (asterisk).

Discussion

Fine intrapleural desmoid tumors are considered a rare clinical entity. Thus, it is a challenge for many clinicians to diagnose such tumors. In this case, the patient complained of pain in the right chest for about 2 years and intermittent coughing. The symptoms of these tumors may vary, and depend on their size and location. Initial imaging for desmoids is usually performed to identify the tumor, determine its extent, define the local mass effect, determine potential resectability for surgical planning, and also for postoperative monitoring [7].

The imaging characteristics of intrathoracic desmoids have been described for several imaging modalities, and the characteristics are similar for desmoids found in other parts of the body. On ultrasound, desmoid tumors present as hypoechoic soft tissue masses with varying vascularity. However, the most commonly used imaging modalities are CT and magnetic resonance imaging (MRI). On CT, isodense-to-slightly hypodense lesions with varying contrast enhancement are well-defined.

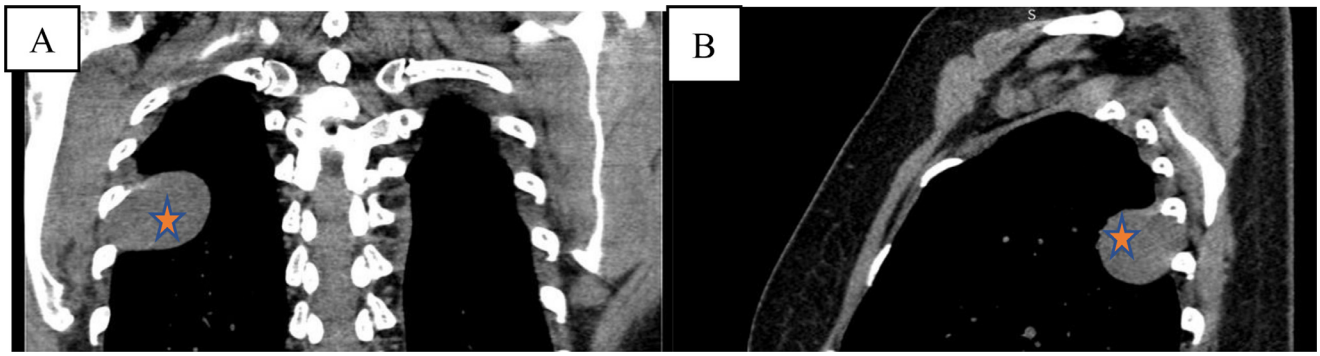

Fig. 3 – Coronal (A) and sagittal views showing the mass attached to the right chest wall (B) (asterisk).

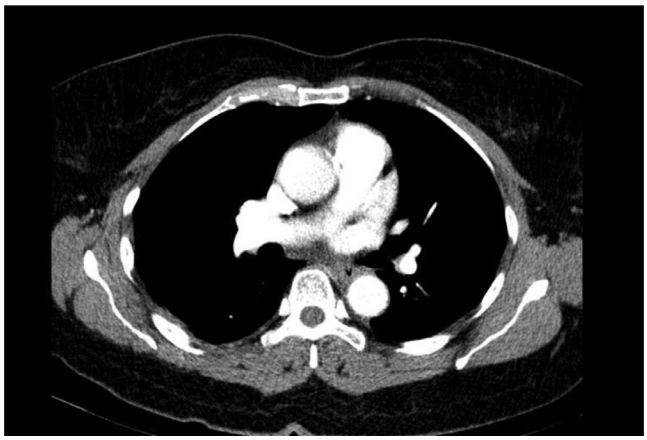

Fig. 4 – Axial view chest computed tomography with a mediastinal window showing no residual mass 2 months after surgery.

Hyperdense masses are those with a high collagen content. On MRI, most desmoid tumors are homogeneously isointense on T1-weighted images. However, they are slightly hyperintense on T2-weighted or short tau inversion recovery images. The imaging appearance can vary depending on the proportions of cellular tissue, myxoid tissue and collagen in the tumor [6]. The emergence of a higher T2 signal on MRI may have better value as it is associated with more rapid growth. Imaging is recommended prior to treatment, and, initially, every 3-6 months after surgery [7]. In the current study, the patient's postoperative CT scan was done 2 months after resection and showed no recurrence or residual mass. In this case, the observation interval and postoperative evaluation interval were concurrent.

In terms of imaging, the major differential diagnosis is malignant soft tissue sarcoma. Desmoid tumors typically show an infiltrative growth pattern, cross fascial boundaries, and do not show central necrosis. However, the majority of soft tissue sarcomas are space-occupying intramuscular lesions that compress, rather than invade, adjacent tissues. Furthermore, soft tissue sarcomas respect fascial boundaries and often show central necrosis. If only primary pleural desmoid tumors are considered, the differential list is more extensive and includes localized fibrous tumors of the pleura, inflammatory pseudotumors, fibroma, mesothelioma, desmoplastic fibroblastoma, and metastatic disease [8].

Histopathological confirmation is required before treatment, even though the imaging is suggestive. The alternatives for treatment include observation, surgical resection, radiation, conventional chemotherapy, hormonal agents, and newer molecular-targeted agents. Based on the location, lo-

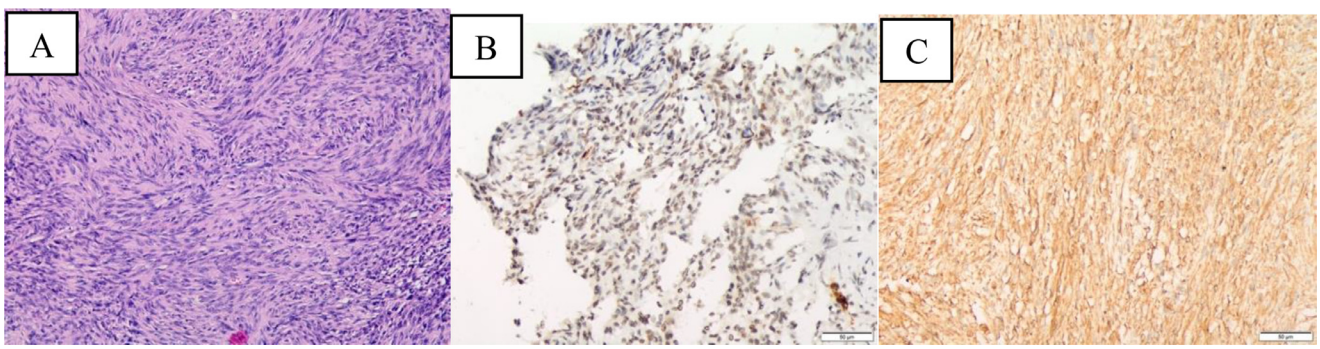

Fig. 5 – The tumor was histologically characterized as a spindle mesenchymal tumor (hematoxylin and eosin stain; original magnification x400) (A), and the immunohistochemistry was positive for vimentin (vimentin stain; original magnification x400) (B) and smooth muscle actin (smooth muscle actin stain; original magnification x400) (C).

cal effects, and clinical course, a multidisciplinary strategy for the individual patient is required [7]. Due to the lack of accumulated experience, intrathoracic desmoid tumors, including pleural masses, should be under long-term observation [8].

Conclusion

It is a challenge for clinicians to diagnose intrapleural desmoid tumors. CT imaging is the primary modality used to characterize these tumors for surgical planning and for postoperative monitoring. Histopathological confirmation should be carried out for a definite diagnosis.

Consent and ethic committee approval

Written consent has been obtained from the patient as there is no patient identifiable data included in this case report. This study has met the ethical principle and already got approval from Research Ethics Committee from Dr. Soetomo General Hospital, Surabaya.

Patient consent statement

Informed consent obtained for publication of a case report.

Written informed consent was obtained from the patient for the publication of this case report.

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