Primary desmoplastic fibroblastoma of diaphragm

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Abstract
Desmoplastic fibroblastoma is an extremely rare benign soft tissue tumor and desmoplastic fibroblastoma originating from the diaphragm has not been documented previously. In our case, we report the first primary diaphragm desmoplastic fibroblastoma.

KEYWORDS
clinical assessment, diaphragm, desmoplastic fibroblastoma

INTRODUCTION
Desmoplastic fibroblastoma, also known as collagenous fibroma, is a rare benign soft tissue tumor that was first described by Evans in 1995.1 To the best of our knowledge, no case of desmoplastic fibroblastoma originating from the diaphragm has been reported so far. In this report, we describe an extremely rare case of diaphragm-derived fibroblastoma.

CASE REPORT
A 56-year-old woman was admitted to our institute on account of a growing tumor mass in the left pleural cavity. The computed tomography (CT) scan conducted 3 years prior had revealed a pleural mass measuring 3.6 × 1.5 cm with a clear boundary. Since the patient had no other complaint, the tumor was diagnosed as benign and regular follow-up was recommended. The patient underwent left mastectomy 6 months before due to mammary cancer, and the tumor mass measured 6.4 × 2.8 cm during imaging. After eight chemotherapy courses, the patient was referred to the thoracic surgery department.

The contrast-enhanced CT showed a strip-like tumor of soft tissue density in the left pleural cavity that was close to the parietal pleura and diaphragm, measured 8.6 × 2.9 cm, and extended to the lobar fissure. There was slight peripheral enhancement but no obvious enhancement for the rest of the tumor mass (Figure 1a). Given the patient’s history of mammary cancer, fluorine-18 fluorodeoxyglucose positron emission tomography (FDG-PET) was performed and showed a slight and diffuse increase in tumor radioactivity. The maximum standardized uptake value was 1.8 and there were no other high radioactive foci. Accordingly, the tumor was diagnosed as benign or low-grade malignant and unrelated to mammary cancer.

A thoracoscopy probe was inserted into the pleural cavity and revealed a grayish white lobulated solid tumor with a mesentery and three microvascular pedicles connected to the diaphragm (Figure 1b). The tumor was not tethered to the lung tissue and therefore was removed en bloc by cutting the mesentery and pedicles with an ultrasonic scalpel. The freshly resected tumor measured 10 × 7.5 cm in diameter (Figure 1c). Histologically, the tumor was hypocellular and consisted of spindle-shaped and stellate-shaped fibroblastic cells embedded in a dense collagenous or myxoid stroma.

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with infrequent mitotic figures (Figure 2a). Immunohistochemical staining showed that the tumor cells were positive for caldesmon but negative for smooth muscle actin (Figure 2b), desmin, CD34, S-100, AE1/AE3, SOX, and CD117. Furthermore, the Ki-67 index was only 1%. Based on the above results, the tumor was finally diagnosed as desmoplastic fibroblastoma. Four months later, CT revealed no evidence of recurrence.

DISCUSSION

Soft tissue tumors are difficult to diagnose on the basis of histopathological examination and have to be ultimately confirmed by immunohistochemical analysis. Therefore, the initial clinical assessment is also important for diagnosis. In the reported case, the tumor mass was first detected 3 years before. Although the patient did not complain of symptoms like chest pain, the tumor volume doubling time was 409 days, indicating chronic tumor growth. Contrast-enhanced CT showed a homogeneously dense, strip-shaped tumor with slight peripheral enhancement. There was a visible boundary between the tumor and parietal pleural tissues, but the tumor adhered closely to the diaphragm and interlobar visceral pleura. Based on the radiological characteristics, the tumor was considered benign or low-grade malignant. However, given the patient’s history of mammary cancer, PET was additionally performed and showed mild radioactive uptake in the tumor, which excluded metastasis of mammary cancer and confirmed the presence of a second primary tumor in the left pleural cavity. We made a small incision in the chest wall and inserted a thoracoscopy probe into the pleural cavity, which led to complete tumor resection with minimal injury.

Although desmoplastic fibroblastoma commonly affects the soft tissues of the extremities, it may possibly originate from the trunk, bone or even viscous tissues. It is a benign tumor that occasionally exhibits invasive characteristics. Complete removal of the tumor is curative and no recurrence has been documented even for “invasive” desmoplastic fibroblastoma. We observed an irregularly shaped solid tumor connected to the diaphragm, and
separated the benign mass from the diaphragm via a simple disjunction of the mesentery. CT scan performed 4 months later revealed no evidence of recurrence, which was consistent with previous studies.

Primary diaphragm tumors are rare and the published literature is limited to individual case reports. Benign and malignant diaphragm tumors have similar morbidity. Diaphragm cysts are the most common benign tumors, whereas lipomatous masses are the most common solid tumors. In addition, pathologically benign or malignant soft-tissue tumors such as fibroblastic/myofibroblastic, vascular, nerve sheath, and smooth muscle tumors have been reported sporadically. In our case, the tumor was hypocellular and consisted of spindle-shaped and stellate-shaped fibroblastic cells embedded in a dense collagenous or myxoid stroma. The pathological type of the tumor was confirmed by immunostaining.

To summarize, this is the first reported case of primary diaphragm desmoplastic fibroblastoma. Radiological examination indicated the benign behavior of the tumor, which was subsequently removed by a small incision and video-assisted thoracoscopy. The final pathological results were confirmed by immunohistochemical staining and short-term CT showed no signs of recurrence. The patient will be followed up as necessary.

ACKNOWLEDGMENTS
There are no sources of support/funding.

CONFLICT OF INTEREST
The authors have stated that there are no competing interests.

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How to cite this article: Guo C, Wang G, Ma D, Li J, Liu H, Li S. Primary desmoplastic fibroblastoma of diaphragm. Thorac Cancer. 2021;12:2961–3. https://doi.org/10.1111/1759-7714.14168