

An intrathoracic giant tumour and vanishing lung cyst after tumour resection

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Abstract

A 65-year-old woman was brought to the emergency unit with an approximately 6-month history of persistent fever and cough. Chest computed tomography (CT) demonstrated a 16-cm heterogeneous mass with adjacent large cyst (approximately 4.0 cm). The patient underwent CT-guided biopsy, and benign solitary fibrous tumour (SFT) was immunohistochemically diagnosed. As the symptoms were thought to be due to enlargement of the tumour, surgery was deemed necessary, and the tumour was successfully resected. Based on morphological and immunohistochemical examination of the resected specimen, the final diagnosis was dedifferentiated SFT (DSFT). Follow-up CT verified disappearance of the pulmonary cyst. The cyst was speculated to be caused by a check valve mechanism, which may also suggest a rapid growth of the tumour. At the time of writing, 2 years post-operatively, no tumour recurrence has been identified. This represents the first report of intrathoracic giant DSFT with a cystic lesion returning to normal lung parenchyma.

KEYWORDS

cyst, dedifferentiated solitary fibrous tumour (DSFT), giant tumour, three-dimensional computed tomography (3D-CT)

INTRODUCTION

Solitary fibrous tumour (SFT) originates from mesenchymal cells. Dedifferentiated SFT (DSFT) is a rare and relatively recently characterized concept. DSFT reportedly occurs most often in the retroperitoneum or pelvic cavity.¹ Among them, there are several case reports of intrathoracic SFT.^{2,3} Although it was diagnosed benign preoperatively, this case was diagnosed as a DSFT on post-operative pathology. In addition, this case was a 16-cm DSFT with adjacent large cyst, which was not resected and returned to normal. To the best of our knowledge, a case of giant DSFT with cyst has not been reported and is rare, thus prompting us to report the present case.

CASE REPORT

A 65-year-old Japanese woman was brought to the emergency unit with a persistent fever and cough. The fever that had

persisted for 6 months had been low grade and could be managed with over-the-counter cold remedies. The cough was also slight and tolerable. However, for 5 days prior to the visit, she had had a fever of over 38°C. She had no medical or family history of note and was not a smoker. There were no recent complaints of weight loss. On clinical examination, her blood pressure was 145/85 mmHg, temperature was 38.6°C, heart rate was 120 beats/min, respiratory rate was 18 breaths/min and oxygen saturation was 94% (room air). Laboratory testing showed a high inflammatory reaction; C-reactive protein, 19.38 mg/dl (reference range 0–0.5 mg/dl); white blood cell count, 19,800/mm³ (reference range 3000–9000/mm³). Chest x-ray showed an approximately 20-cm mass shadow in the left lower lung field (Figure 1A). Contrast-enhanced computed tomography (CT) of the chest demonstrated a 16-cm heterogeneous mass containing calcification and contrast effect. Interestingly, a cyst (approximately 4.0 cm) was apparent adjacent to the mass. The mass appeared to be originating from the left dorsal side and compressing the left lower lobe (Figure 1B–D).

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On the basis of these findings, pneumonia, lung abscess or thoracic empyema were suspected, so antimicrobial treatment was initiated. Although inflammatory reactions improved with treatment, the mass did not shrink and our patient still suffered intractable cough. The patient underwent CT-guided biopsy on hospital day 9. Histopathological examination demonstrated

a tumour comprising haphazardly arranged, spindle cells admixed within thin collagen fibres, but no apparent mitosis. Immunohistochemical staining yielded positive results for CD34 and BCL2. On the basis of these morphological and immunohistochemical findings, conventional SFT was diagnosed. As the fever and high inflammatory response were

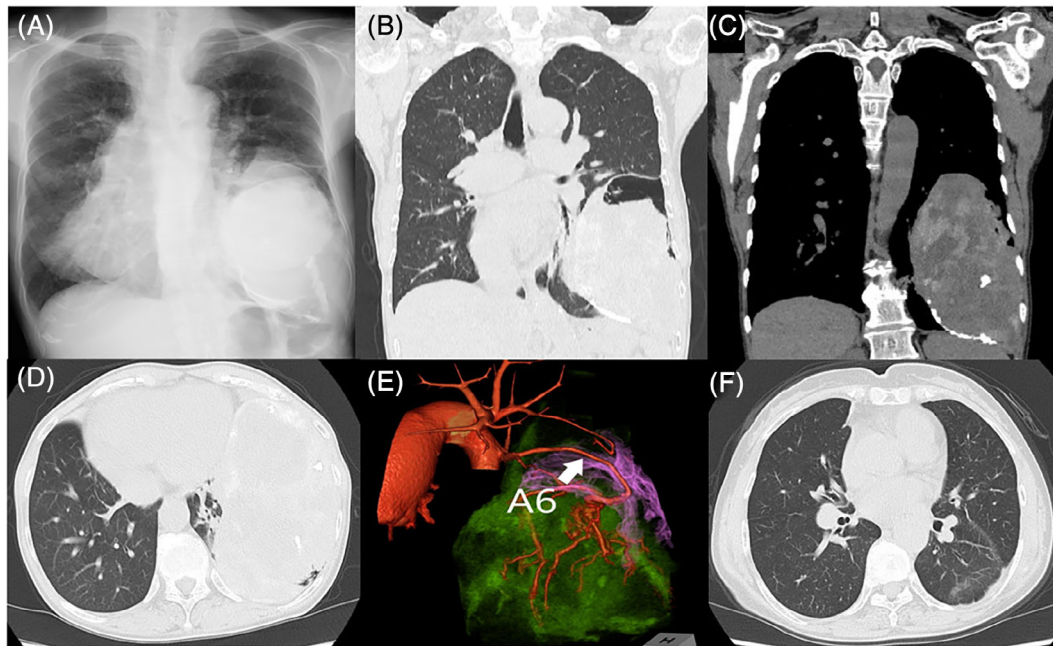


FIGURE 1 (A) Chest radiograph on admission shows a 20-cm diameter mass shadow in the left lower lung field. (B–D) Chest computed tomography (CT) on admission shows a 16-cm diameter tumour shadow with partial calcifications and cyst-like structure including blood vessels adjacent to the tumour. (E) Preoperative three-dimensional CT image shows a tumour (coloured in green) and a cyst-like structure (coloured in purple). It also shows that the superior artery of the left lung (A6) was involved in the cyst. (F) Chest CT 7 months after surgery shows the cyst-like structure has disappeared

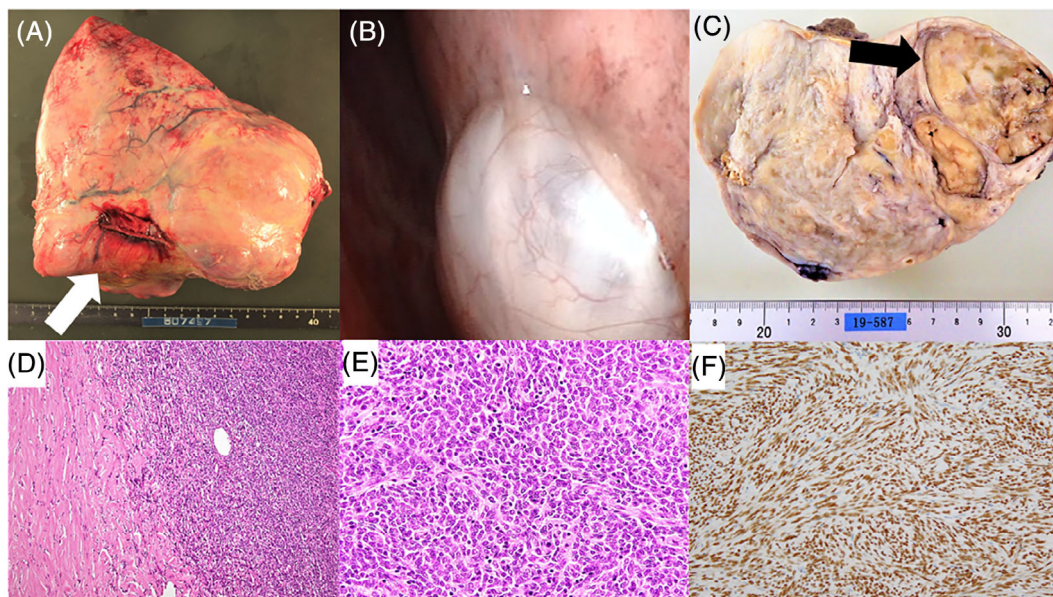


FIGURE 2 (A) Photograph of the excised specimen from the mediastinal side. White arrow indicates resected stalk. (B) Intraoperative findings. A pulmonary cyst with a thick cyst wall is seen. (C) Macroscopic findings. The cut surface of the tumour is well demarcated showing lobulated pattern. Black arrow shows yellowish nodule in the tumour with foci of necrosis. (D) The tumour consists of two regions: a conventional solitary fibrous tumour (SFT) area (left) and a dedifferentiated area (right) (haematoxylin and eosin [HE] stain). (E) The dedifferentiated area shows high cellularity of the tumour cells with many mitotic figures (HE stain). (F) The tumour cells are positive for STAT6. They are positive both for a conventional SFT area and a dedifferentiated area

thought to be most likely secondary to the infection associated with the tumour growth, surgery was planned as a radical treatment. Before surgery, three-dimensional CT (3D-CT) suggested that the cyst-like changes were only adjacent to the tumour and did not originate from the tumour (Figure 1E). Preoperative respiratory function test showed combined ventilatory impairment; expiratory forced vital capacity (FVC) was 1.6 L, FVC was 68.4% of the predicted value, forced expiratory volume in 1 s (FEV₁) was 1.1 L, FEV₁ was 60.4% of the predicted value and FEV_{1%} was 68.6%. Intraoperative findings showed the mass had not invaded into the left lower lobe, chest wall or diaphragm. The tumour showed a stalk with trophic vessels arising from the visceral pleura. Because the tumour was huge, the mediastinal side of the tumour was not visible, but the stalk was manually identified with reference to the 3D-CT findings. The tumour was finally extracted after resection of the stalks using automatic suturing devices (Figure 2A). As preoperative 3D-CT had suggested, a huge cyst with thick cystic walls was also found (Figure 2B), but was expected to disappear with the release of the check valve mechanism, and so was not resected. Macroscopic finding showed that the cut surface of the tumour is well demarcated showing lobulated pattern and yellowish nodule in the tumour with foci of necrosis (Figure 2C). Histopathological examination revealed the tumour comprised two regions: conventional SFT regions and sarcomatous regions that were hypercellular and lacked the morphological features of conventional SFT (Figure 2D). Conventional SFT areas comprised a patternless proliferation of spindle-shaped cells. On the other hand, the dedifferentiated areas lacked characteristic features of SFT and consisted of pleomorphic cells with a high mitotic rate (5/10 high-power fields) (Figure 2E). The tumour cells are positive for STAT6, and both conventional and DSFT were positive (Figure 2F). Immunoreactivities for CD34 and BCL2 were increased in the dedifferentiated areas. The dedifferentiated areas were sharply demarcated from the conventional SFT in most areas, but showed gradual separation in a few areas, indicating dedifferentiation from conventional SFT (Figure 2D). On the basis of these morphological findings, we diagnosed DSFT rather than malignant SFT. Follow-up CT after 7 months showed disappearance of the giant cyst (Figure 1F). The post-operative CT showed no cyst-like lesions, confirming that the preoperative cysts originated from normal lung parenchyma. Post-operative respiratory function test showed improvement; FVC was 2.19 L, FVC was 85.7% of the predicted value, FEV₁ was 1.37 L, FEV₁ was 68.8% of the predicted value and FEV_{1%} was 63.43%. At the time of writing, 2 years post-operatively, the patient has shown no recurrence of tumour, and she remains free of intractable cough.

DISCUSSION

DSFT is rare, with about 30 cases reported worldwide.² DSFT often arises in the retroperitoneum or pelvic cavity.¹ Diagnosis of DSFT can only be confirmed through histopathological analysis.⁴ Dedifferentiation is a phenomenon

that is well described in soft tissue and bone tumours. Morphologically, dedifferentiation is characterized most often by an abrupt transition between well-differentiated components and high-grade areas of the tumour, and confers more aggressive biological behaviour.³ Histologically, DSFT comprises conventional SFT and a high-grade, sarcomatous, dedifferentiated area. Dedifferentiated areas generally lack the morphological features characteristic of SFT composed of fibroblast-like cells, collagen fibres and thin-walled blood vessels.² This is in contrast with malignant SFT, in which even the atypical portion contains some features of typical SFT. In addition, an abrupt transition between SFT and a high-grade area is observed with loss of immunoreactivities, such as for CD34.² The morphological findings supported a definitive diagnosis of DSFT in this case. One of the problems in this case was that preoperative needle biopsy could not reach a definitive diagnosis. Interpretation of pathological findings obtained from needle biopsy must consider the possibility that the sufficient specimen was not optimally collected. The huge tumour obstructed fine visual development of the surgical site in this case. Nevertheless, preoperative 3D-CT showed the exact location of the tumour and cyst, and we were able to complete the surgery successfully. Even more interesting in this case was the large cyst adjacent to the tumour. Pulmonary cystic airspaces are reported to be associated with 1% of non-small cell lung cancer, and can be due to a check valve mechanism obstructing the small airways.⁵ In this case, the cyst disappeared after resection of the tumour, suggesting a check valve mechanism as the cause of the cyst, but the evidence is poor. However, the check valve mechanism was speculated to be affected by the rapid growth of the tumour, and this case was also considered to be a mass with a relatively fast growth rate. This case was a giant DSFT with adjacent cyst, which is very rare. And this case was unfortunately diagnosed as conventional SFT at the intraoperative diagnosis. Preoperative diagnosis may not be able to determine whether the mass is benign or malignant, so we need to be careful.

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CONFLICT OF INTEREST

None declared.


AUTHOR CONTRIBUTION

Conception or design of the work: Masashi Nishimura and Sumitaka Yamanaka. Drafting the work or revising it: Masashi Nishimura and Shinichiro Ota. Final approval of the version to be published: All authors.

ETHICS STATEMENT

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

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REFERENCES

1. Collini P, Negri T, Barisella M, Palassini E, Tarantino E, Pastorino U, et al. High-grade sarcomatous overgrowth in solitary fibrous tumors: a clinicopathologic study of 10 cases. *Am J Surg Pathol*. 2012;36(8):1202–15.
2. Yang JW, Song DH, Jang IS, Ko GH. Dedifferentiated solitary fibrous tumor of thoracic cavity. *Korean J Pathol*. 2014;48(3):250–3.
3. Masuda Y, Kurisaki-Arakawa A, Hara K, Arakawa A, Oh S, Suzuki K, et al. A case of dedifferentiated solitary fibrous tumor of the thoracic cavity. *Int J Clin Exp Pathol*. 2014;7(1):386–93.
4. Olson NJ, Linos K. Dedifferentiated solitary fibrous tumor: a concise review. *Arch Pathol Lab Med*. 2018;142(6):761–6.
5. Fintelmann FJ, Brinkmann JK, Jeck WR, Troschel FM, Digumarthy SR, Mino-Kenudson M, et al. Lung cancers associated with cystic airspaces: natural history, pathologic correlation, and mutational analysis. *J Thorac Imaging*. 2017;32:176–788.

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