Solitary Fibrous Tumor Arising from Hyperplastic Thymus

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Abstract

One case of a solitary fibrous tumor arising from the hyperplastic thymus is recorded. The patient was a 37-year-old female who presented with an anterior mediastinal mass. Thoracotomy was performed and revealed that the tumor arose on a pedicle from the posteroinferior surface of the enlarged thymus. The pathologic findings were characteristic of a solitary fibrous tumor. This is a very rare neoplasm that occurred in the mediastinum and had evidence of thymus gland in origin.

Intrathoracic solitary fibrous tumor (SFT) usually occurs in pleura[1,2]. That presents as intrapumonary or mediastinal mass is rare[3-5]. Among the tumors occurring in the mediastinum, very few cases demonstrated firm evidence of thymic origin[5,6]. The rest revealed only the tumor site in the anterosuperior mediastinum, the same location as the thymus. We present a case of SFT that arose on a pedicle from hyperplastic thymus to support the organ of origin for those tumors occurring in the mediastinum.

CASE REPORT

A 37-year-old Thai female was admitted to Central Chest Hospital due to problems of chest discomfort, malaise and weight loss of 4 kg within 3 months. On physical examination, no abnormal clinical findings were detected. Chest X-ray revealed lobulated soft tissue mass at the anterior mediastinum blending with the left heart border. Computed tomography of the chest revealed a homogeneous mass located anterior to the ascending aorta. The mass was 8 cm in length with superior extension 2 cm above the aortic arch. Inferiorly it involved along the pericardium and formed a continuing mass of 6x2 cm at the left heart border (Fig. 1). Median sternotomy was performed and revealed a well-circumscribed anterior mediastinal mass on a pedicle from the posteroinferior surface of the thymus which was generally enlarged.

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Removal of the mass with total thymectomy was performed. The patient recovered well after surgery and showed no evidence of recurrence or metastasis on follow-up.

Grossly the specimen consisted of a tumor from the anterior mediastinum separated from the thymus gland. The tumor, measuring 6x6x2 cm and weighing 50 g, was lobulated, well-encapsulated and hard in consistency. It showed a small round area of raw surface on one side of the smooth capsular surface, indicated by the surgeon as a pedicle of the tumor arising from the thymus (Fig. 2A). Cut surfaces of the tumor revealed homogeneous grey-white tissue with no hemorrhage, necrosis or cystic degeneration. The thymus, measuring 10x8x2 cm and weighing 55 g, showed bilobated structure with generalized enlargement (Fig. 2B). Cut surfaces revealed a large amount of grey-brown thymic tissue.

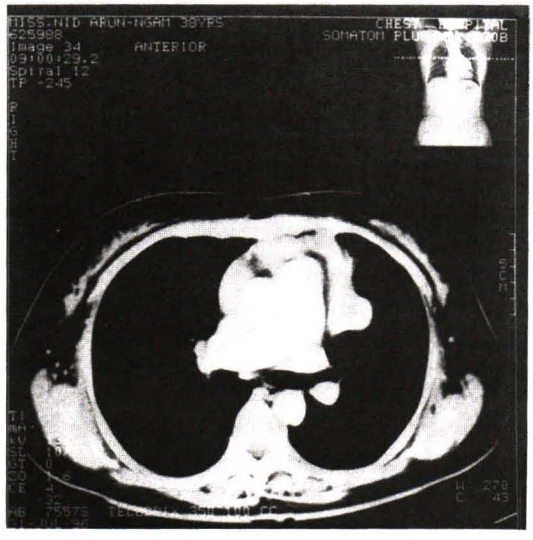
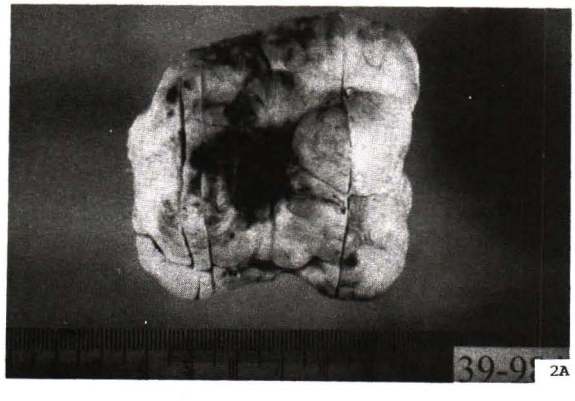


Fig. 1. CT of the chest reveals a homogeneous mass located anterior to the ascending aorta with a continuing lobulated mass at the left heart border.

Microscopically the tumor was made up of spindle cells with hyperchromatic nuclei and abundant eosinophilic collagenized stroma (Fig. 3A, B). The nuclei showed mild anisonucleosis. A few round to oval epthelial-like cells were noted in one region. Thick fibrous capsule, a few loose cellular foci with wavy nuclei were noted. There was no mitosis identified. Special stain for Masson's trichrome stained dark blue cytoplasm. Immunohistochemistry for S-100 protein, neurofilament, cytokeratin, epithelial membrane antigen and CD34 were negative. The neoplastic cells showed strongly positive immunoreactivity for only vimentin. Sections of the thymus showed thymic lobulation made up of hypercellular cortex and medulla. The pathologic findings were typical of solitary fibrous tumor arising from the hyperplastic thymus.

DISCUSSION

Intrathoracic SFT arising from visceral or parietal pleura is not uncommon but that arising in



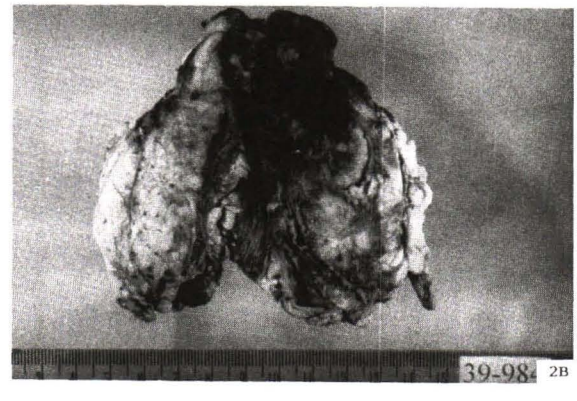


Fig. 2. Grossly the tumor is lobulated, well encapsulated and shows a raw surface of pedicle that was previously separated from the thymus at the middle part of the tumor (A). The thymus gland shows generalized enlargement of bilobated organ (B).

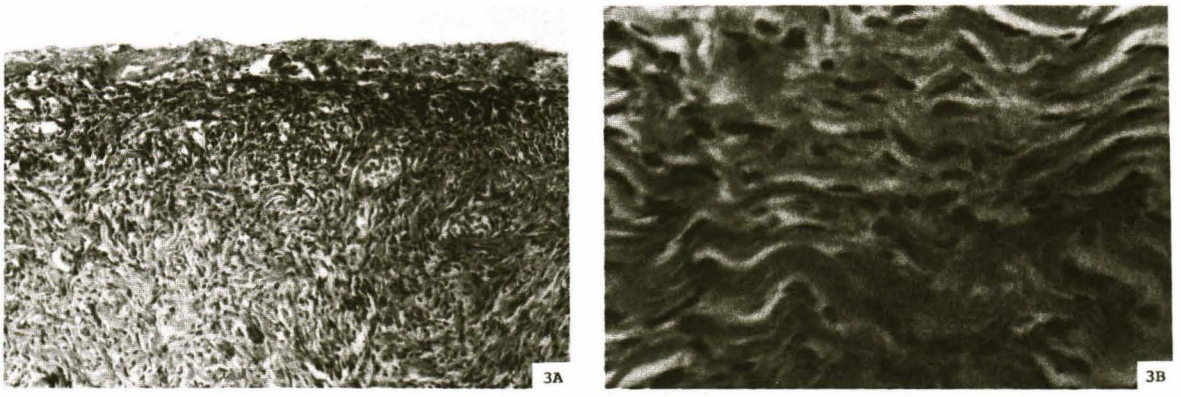


Fig. 3. Microscopically the tumor shows fibrous capsule covering a mass of spindle cells (A) in abundant collagenized stroma (B).

the mediastinum is rare. The largest series by Witkin et al in 1989 revealed that 11 of 14 mediastinal SFTs arose in the anterosuperior portion, the location compatible with thymic origin[5]. However, only two cases in their series had firm evidence: a pedicle from the thymus in one tumor and entrapped thymic element in the other. Fukushima et al in 1992 reported the first Japanese case of mediastinal SFT and revealed that the tumor also arose from the thymus[6]. Other series revealed only a few tumors arising from the pericardium[2], mediastinal reflection of parietal pleura[1] or failed to demonstrate the exact organ of origin in the mediastinum[3,4]. In Thailand, no case of mediastinal SFT has been reported before. The largest series of mediastinal tumors by Wongsangiem et al did not mention this type of tumor[7].

SFT can behave as a benign or malignant neoplasm. That with malignant behaviour may produce local invasion, recurrence but rare metastasis[1,2,5]. Histologic features of malignancy are high cellularity, mitotic activity, pleomorphism, hemorrhage and necrosis but these do not indicate definitely the clinical behaviour of the tumor[2]. The important indicators of good clinical outcome are circumscription, presence of a pedicle supporting the tumor and the ability to be totally excised[1,2,5].

In our case, by using computed tomography of the chest alone, it was difficult to differentiate a mediastinal tumor connected with hyperplastic thymus from a single lobulated mass occupying the anterior mediastinum with extension to the left hemithorax. Operative findings revealed that the tumor was well-circumscribed, arose on a pedicle from the posteroinferior surface of the thymus and was completely removed. The histologic findings were favourable except for nuclear hyperchromasia and mild anisonucleosis. These features indicated good prognosis.

The thymus gland normally decreases in size and weight with advancing age after puberty. This process is called physiologic involution[8]. But thymic hyperplasia is associated with a large number of diseases. We do not know the exact cause of the non-involuted but hyperplastic change of the thymus gland in our case. But the remaining organ revealed the origin of the tumor. Although we found no entrapped thymic element from histologic examination, the pedicle from the thymus that supported the tumor confirmed the thymic origin of mediastinal SFT in our case.

We also believe that those cases of SFT located in the anterosuperior mediastinum with no connection to pleura, pericardium or any other specific organs might originate from the thymus but the patients were found to have tumor at the end of the thymic involution. Therefore, SFT is still an exceedingly rare neoplasm found to arise from the thymus.

SUMMARY

One case of solitary fibrous tumor arising from hyperplastic thymus is recorded. The patient was a 37-year-old female who presented with an anterior mediastinal mass. Thoracotomy was performed and revealed that the tumor arose on a pedicle from the posteroinferior surface of the enlarged thymus. The pathologic findings were characteristic of solitary fibrous tumor. This is a very rare neoplasm that occurred in the mediastinum and had evidence of thymus gland in origin.

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โซลิทารีไฟบรัสทูเมอร์ ที่เกิดขึ้นจากไฮเปอร์พลาสติคธัยมัส

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รายงานผู้ป่วยหญิงอายุ 37 ปี ตรวจพบเงาของก้อนที่ตำแหน่งเมดิแอสตินัมจากภาพรังสีของทรวงอก และได้รับการผ่าตัดพบว่ามีเนื้องอกเกิดขึ้นจากต่อมธัยมัสที่โตขึ้นกว่าปกติ ลักษณะที่ตรวจพบทางพยาธิวิทยา เข้าได้กับโซลิทารีไฟบรัสทูเมอร์ ซึ่งเป็นเนื้องอกที่พบได้น้อยที่ตำแหน่งเมดิแอสตินัม และมีรายงานผู้ป่วยน้อยมากที่สามารถแสดงต้นกำเนิดของเนื้องอกชนิดนี้จากต่อมธัยมัส

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