Case Report

Mediastinal angiomatosis: a rare case report

Min Gyoung Pak[1], Phil Jo Choi[2], Won Suk Choi[3], Eun Ju Kang[4], Mee Sook Roh[1]

[1]Department of Pathology, [2]Department of Thoracic and Cardiovascular Surgery, Dong-A University College of Medicine, Busan, South Korea; [3]Department of Thoracic and Cardiovascular Surgery, Eulji University School of Medicine, Daejeon, South Korea; [4]Department of Radiology, Dong-A University College of Medicine, Busan, South Korea

Correspondence to: Mee Sook Roh, MD, PhD. Department of Pathology, Dong-A University College of Medicine, 26 Daesingongwon-ro, Seo-gu, Busan 602-715, South Korea. Email: msroh@dau.ac.kr.

Abstract: Angiomatosis refers to a rare condition of large hamartomatous vascular lesions that extensively affect a region of the body or several different tissue types in a contiguous way. Several cases have been reported in the mediastinum. We experienced a histologically proven case of mediastinal angiomatosis in a 56-year-old woman that was incidentally detected as multiple conglomerated masses mimicking metastatic lymph nodes on chest radiography. Despite its rareness, our case emphasizes that pathologists and radiologists need to be aware of the rare diagnosis of angiomatosis in the mediastinum.

Keywords: Angiomatosis; mediastinum; vascular malformation

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Introduction

Angiomatosis refers to a rare condition characterized by diffuse proliferation of benign, architecturally well-developed vascular structures with accompanying mature fat that affects a large segment of the body in a contiguous fashion, either extending vertically to involve multiple tissue plans (e.g., skin, subcutis) or by crossing multiple compartments to involve similar tissue types (e.g., multiple muscles) (1,2). It is a histologically benign but clinically extensive and serious vascular lesion due to the high recurrence rate after initial excision (2). Furthermore, angiomatosis could be misdiagnosed as a malignancy on radiological studies because of its infiltrative nature (3). It most commonly affects the lower extremities but also localizes in the abdomen, chest wall, head and neck region, back, and retroperitoneum (2). However, several cases of angiomatosis have been reported in the mediastinal region (3-8). Herein, we report an extremely rare case of mediastinal angiomatosis presenting as multiple mediastinal masses that mimicked metastatic lymph nodes on chest radiography.

Case presentation

Multiple mediastinal masses were found incidentally on a 56-year-old Korean woman during a radiological evaluation for a thyroid mass at another hospital. The patient underwent a left thyroidectomy, and a diagnosis of adenomatous goiter was made at the other hospital. Then, the patient was referred to Dong-A University Medical Center to confirm the diagnosis and undergo further treatment for the mediastinal masses. The patient had a medical history of adenomatous goiter of the left thyroid 6 months ago, acute appendicitis 10 years ago, and uterine leiomyoma 20 years ago. There was no history of hereditary etiology of vascular disease such as von Hippel-Lindau disease. The patient had no remarkable family history. A physical examination revealed a healthy looking woman with no obvious signs. Routine laboratory investigations were normal. A computed tomography (CT) scan of the chest with and without contrast revealed multiple conglomerated, nonhomogeneous, low-density masses with mild peripheral enhancement, measuring from 1.8 to 3.0 cm in diameter,

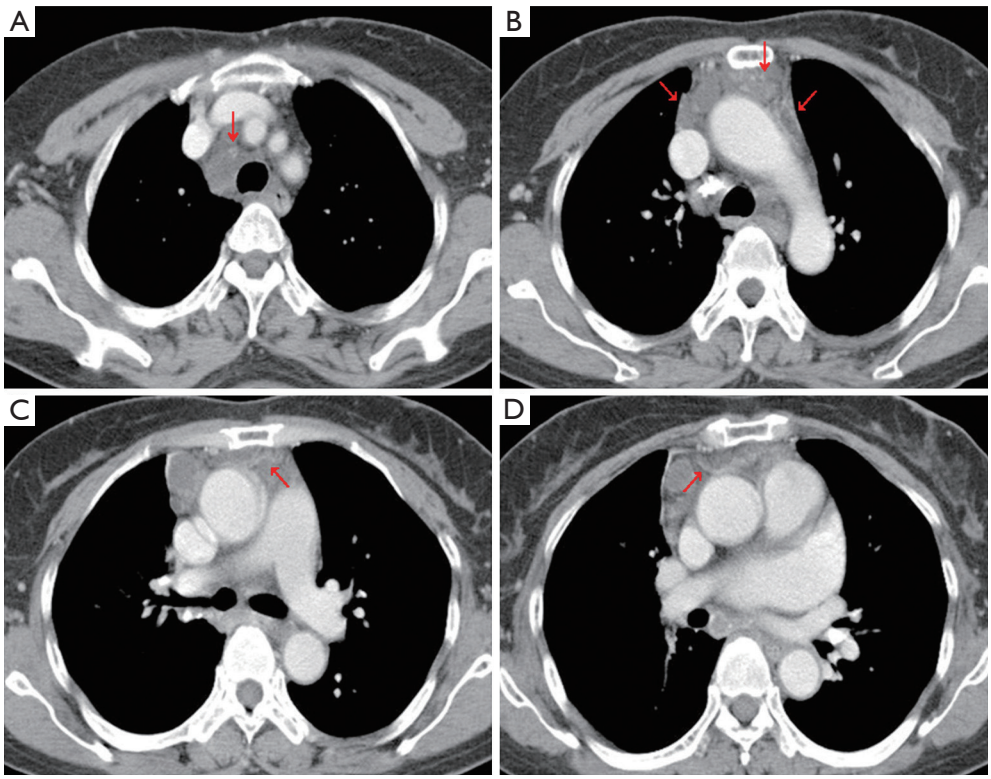

Figure 1 Contrast-enhanced chest CT scan shows multiple conglomerated, nonhomogeneous, low-density masses of 1.8 to 3.0 cm in diameter in the right upper-paratracheal area (A) and anterior mediastinum (B-D). Somewhat streaky, serpiginous, and heterogeneous peripheral enhancements are seen through the small supplying vessels (arrows). CT, computed tomography.

in the superior and anterior mediastinum (*Figure 1*). The radiological studies did not allow us to postulate a specific diagnosis; metastatic lymphadenopathy was suggested initially and malignant lymphoma and tuberculous lymphadenopathy were also included in the differential diagnosis. An endobronchial ultrasound-guided biopsy was attempted to obtain a tissue specimen for diagnosis. Unfortunately, the results were nondiagnostic because the sample only contained a few histologically normal bronchial epithelial cells. Video-assisted thoracoscopic surgical resection was planned for a definitive diagnosis of the mediastinal mass. Thoracoscopy revealed multiple yellowish fatty masses with poorly defined infiltrative borders within the superior and anterior mediastinal spaces that varied from a few to several centimeters in diameter. No connection to the pulmonary circulation was detected. A lung evaluation did not reveal evidence of a parenchymal lesion. Because of the extensiveness of the lesions and their location, no complete resection was performed. One of the largest masses was thoracoscopically excised for a pathological diagnosis, but residual tumor tissues were left behind. The resected specimen revealed an ill-defined predominantly fatty appearing mass with multiple sponge-form hemorrhagic foci, measuring 3.0 cm in diameter (*Figure 2*). The specimen was a tumor-like lesion, consisting of a haphazard proliferation of variously sized vessels, particularly large veins, intermingled with abundant fat tissue (*Figure 3A,B*). The large venous vessels had irregular thick and disordered muscular walls with occasional irregular attenuations and herniations (*Figure 3C*). Sprout clusters of small venules or capillaries resided adjacent to and in the wall of larger vessels (*Figure 3D*). No actual communication of the arteries and veins was identified. These vessels grew randomly in association with large amounts of mature fat with no cellular atypia or lipoblasts (*Figure 3E*). Patchy areas of lymphocytic infiltration, occasionally in a perivascular arrangement, were also present (*Figure 3F*). To help identify the vascular structure, elastic stain and CD31 and smooth muscle actin immunohistochemistry were performed. Thus, the final diagnosis of angiomatosis arising in mediastinum was rendered. Although a subsequent

CT scan performed 3 months later showed persistence of the mediastinal mass with no significant changes in size or density, the patient's disease has been stable for 14 months after surgery.

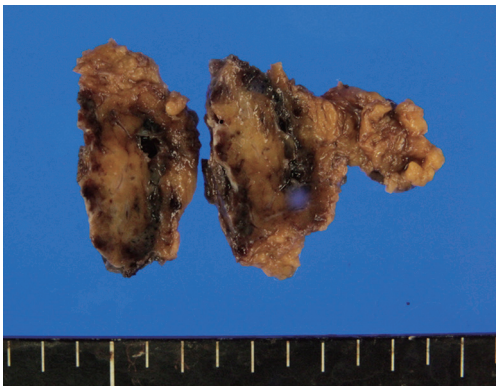

Figure 2 Resected specimen reveals an ill-defined mass with a predominantly fatty appearance and multiple sponge-form hemorrhagic foci, measuring 3.0 cm in diameter.

Discussion

Although the term angiomatosis has been used for a variety of vascular conditions, the single term angiomatosis of soft tissue usually refers to large hamartomatous vascular lesions that are distinctive from an arteriovenous malformation (1,2). Co-existence of vascular proliferation and abundant fat in the infiltrating lesions suggests that angiomatosis is not simply a proliferation of vessels but a more generalized mesenchymal proliferation (2). It has a highly characteristic but not totally specific histological pattern. Therefore, vascular malformation, arteriovenous malformation, and venous malformation have been used as synonyms for angiomatosis. Moreover, because the term angiomatosis has been used previously with many connotations, it is likely that some lesions previously called angiomatosis were examples of various vascular lesions (2). Because of nomenclatural issues, it has been very difficult to gain a clear understanding of this lesion and to compare experiences derived from one specialty with another. Nevertheless, it is clear that angiomatosis in the mediastinum is extremely

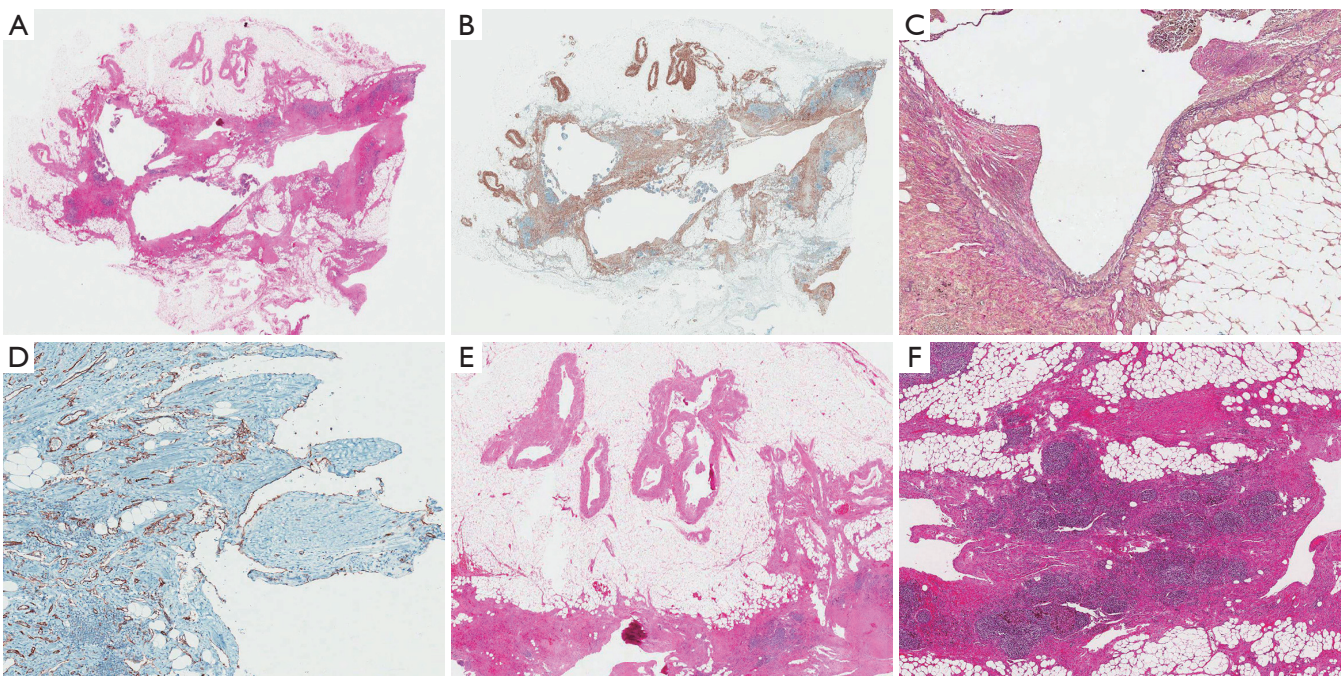

Figure 3 (A,B) The specimen reveals a tumor-like lesion consisting of a haphazard proliferation of variously sized vessels, particularly large veins, intermingled with abundant fat tissue (A: hematoxylin and eosin, ×4 and B: smooth muscle actin immunohistochemistry, ×4); (C) the large venous vessels have irregular thick and disordered muscular walls with occasional irregular attenuations and herniations (elastic stain, ×100); (D) sprout clusters of small venules or capillaries reside adjacent to and in the walls of larger vessels (CD31 immunohistochemistry, ×100); (E) vessels are growing randomly in association with large amounts of mature fat with no cellular atypia or lipoblasts (hematoxylin and eosin, ×40); (F) patchy areas of lymphocytic infiltration, occasionally in a perivascular arrangement, are also present (hematoxylin and eosin, ×100).

rare, as only several cases have been described (3-8). Only a few articles reported earlier was available when the PubMed search was conducted for English-language articles (3,5,6,8). We herein describe an additional case that fits the diagnostic features of angiomatosis in the mediastinum. Our case emphasizes that mediastinum can be site for angiomatosis, although it is very uncommon.

Soft tissue angiomatosis develops almost exclusively in the first two decades of life with female predilection and is very uncommon after the age of 30 years. In the series by Rao and Weiss (2), which included 51 cases of soft tissue angiomatosis, the oldest patient was 54 years, and more than half of the cases occurred in the lower extremities, followed by the chest wall, abdomen, and upper extremities. Interestingly, the lesion in our patient was detected at the age of 56 years, the oldest reported patient, and a lesion was only detected in the mediastinum but not in the extremities. Although soft tissue angiomatosis has no tendency to evolve over time into an aggressive malignant tumor, the importance of recognizing angiomatosis is that most patients develop recurrences probably due to incomplete excision in the face of the infiltrative nature of the extensive disease. Nearly 90% of lesions persist after surgical excision, and 50% recur multiple times within a 5-year period. The mean interval for recurrence is 5 years after the original presentation (2). Very rarely, angiomatosis with massive or progressive bone destruction can occur, which was first described as Gorham's disease by Gorham and Stout (9). Stojsic *et al.* (10) recently reported that Gorham's disease of the ribs associated with angiomatosis of the thoracic wall. Our patient remained stable with a persistent residual tumor at the time of this report (14 months postoperatively).

Despite the rareness, our case emphasizes that pathologists and radiologists need to be aware of the rare diagnosis of angiomatosis in the mediastinum. CT scan showing diffuse nonhomogeneous masses occasionally results in an erroneous impression of malignancy, although the presence of serpiginous density within a low density area is highly characteristic of angiomatosis and corresponds to the large tortuous vessels situated within a fatty background (3). Actually, the lesions appeared as ill-defined nonhomogeneous masses mimicking metastatic lymphadenopathy on the CT scan in our case. The diagnosis of angiomatosis is usually not suspected radiologically but histopathology can reveal the definitive diagnosis. Because of the presence of large amounts of fat, these tumors often appear as predominantly fatty tumors. Histologically, two main patterns are seen. The most common pattern consists of a haphazard proliferation of varying sized vessels, particularly large veins, as in the present case. Less frequent is a capillary hemangioma-like pattern (2). The main histological differential diagnoses include infiltrating lipoma, angiolipoma, angiomyolipoma, angiomyxolipoma, intramuscular angioma, and liposarcoma (11,12). Differentiating these entities and forming the exact histopathological diagnosis of angiomatosis are important due to its high recurrence rate.

In conclusion, our case emphasizes that clinicians, radiologists, and pathologists need to be aware of the rare diagnosis of angiomatosis in the mediastinum and should carefully evaluate the clinical and radiological findings.

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Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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