OLGU SUNUMU CASE REPORT

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Two Cases with Anterior Mediastinal, Periaortic and Pleural Multicentric Idiopathic Fibrosis: Long-term Follow-up Results with Computed Tomography

Anterior Mediastinal, Periaortik ve Plevral Multisentrik İdiyopatik Fibrozis Tanılı İki Olgu: Bilgisayarlı Tomografi ile Uzun Süreli Takip Sonuçları

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ABSTRACT Intrathoracic fibrosis is a rare disease with a benign histopathology mimicking malignant mass lesions which can be seen in infiltrative or limited mass form and can be followed by computed tomography as a primary imaging modality. In the literature, there are cases of mediastinal or pleural fibrosis. However, in our 2 cases, pleural involvement, retrosternal mass, and posterior mediastinal mass surrounding the aorta are seen at the same time. In particular, both parasternal pleural thickening in the form of bat wings is noteworthy. We could not find a published case in which 3 separate components were involved at the same time, followed up for a long time such as 8 and 6 years. Multicentric intrathoracic fibrosis cases were investigated with pre-diagnoses of thymoma, lymphoma, asbestos, and mesothelioma, diagnosed by biopsy and followed up in our hospital for 6-8 years, are presented because they are rare and instructive.

Keywords: Fibrosis; pleura; mediastinum

ÖZET İntratorasik fibrozis, malign kitle lezyonlarını taklit eden, infiltratif veya sınırlı kitle formunda görülebilen, benign histopatolojiye sahip, tanısında ve takibinde bilgisayarlı tomografinin primer görüntüleme yöntemi olduğu nadir bir hastalıktır. Literatürde, mediastinal ya da plevral fibrozis olgularına rastlanmaktadır. Ancak 2 olgumuzda da plevral tutulum, retrosternal kitle ve aortayı saran posterior mediastinal kitle aynı anda görülmektedir. Özellikle yarasa kanadı şeklinde her 2 parasternal plevral kalınlaşma dikkati çekmektedir. Literatürde 3 ayrı komponentin aynı anda tutulum gösterdiği, 8 ve 6 yıl gibi uzun süre takip edilen olguya rastlamadık. Timoma, lenfoma, asbest ve mezotelyoma ön tanıları ile araştırılan, biyopsi ile tanı alan, 6 ve 8 yıl boyunca hastanemizde takip edilen, multisentrik intratorasik fibrozis olguları, nadir ve öğretici olmaları nedeniyle sunulmuştur.

Anahtar Kelimeler: Fibrozis; plevra; mediastinum

Fibrosis, an abnormal proliferation of collagen connective tissue, can be seen in any organ or body system. Retroperitoneal, pleural or mediastinal fibrosis are often present individually and with different types of etiopathogenesis.[1-3] In this group, retroperitoneal fibrosis (RPF), or Ormond's disease which was initially defined in 1948 is the best known and most common form.[4] More than 70% of cases are idiopathic.[5] Only a few intrathoracic periaortic, pleural, or pericardial fibrosis cases accompany RPF in the

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literature.[5-7] Granulomatous infections such as histoplasmosis and tuberculosis are forefront in the secondary form's etiology in mediastinal fibrosis. It is mostly seen paratracheal, subcarinal, and perivascularly in the middle mediastinal compartment.[8,9] Trachea, esophagus, and vena cava superior stricture can be seen.[7] Asbestosis constitutes an essential group in benign pleural thickening etiology.[10] The mass lesion surrounding the aorta in the posterior mediastinum suggests infiltrating malignant processes, primarily lymphoma or metastasis.[11] We have not encountered synchronous fibrosis cases in the anterior and posterior mediastinum and bilateral pleura without RPF.

Computed tomography (CT) imaging, play an important role in management and follow-up of in this group of diseases. The lesion can be seen in the form of a mass that surrounds and infiltrates the vascular and other visceral structures with unclear margins or in the form of a lobulated smoothly confined mass that has the isodensity of muscle on CT. While there is no necrosis or hemorrhage within the mass lesion, increasing calcification may occur over time. Contrast absorption is dependent on the phase of the disease; in the late stage, it is minimal or absent. The idiopathic form and the secondary form cannot be separated radiologically.[2,12]

Our aim to present these rare cases, which mimic malignancy with imaging findings but have benign histopathology, with long-term follow-up results, especially the mass in the posterior mediastinum, without biopsy, is followed by CT to show the benign prognosis.

CASE REPORTS

CASE 1

The first patient is a 62-year-old male patient presenting with right flank pain and cough. Postero-anterior chest x-ray demonstrated a mediastinal mass. CT examination revealed a 6x4 cm well-defined retrosternal solid mass lesion with lobulated contour on the level of the brachiocephalic vein and aortic arch showing pleural extension into each hemithorax (Figure 1, Figure 2). A more extensive and infiltrative second solid mass lesion was seen in the posterior mediastinum, surrounding the aorta, and extended around the paravertebral pleura on both sides (Figure 3). There was no pressure or wall irregularity on the aorta or vertebral invasion. Scattered nodular

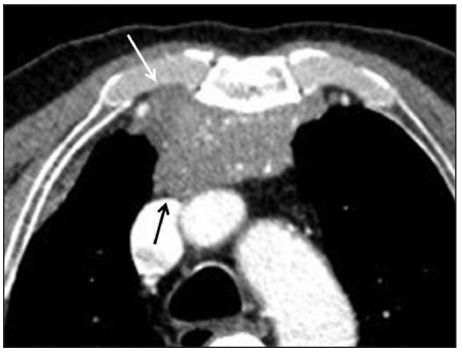

FIGURE 1: Fat tissue has been preserved between the retrosternal solid lesion with left brachiocephalic vein and vena cava superior. The fat plan between the sternum and the mass cannot be distinguished. Right parasternal extrapleural lobulation and nodular calcifications within the mass are noteworthy.

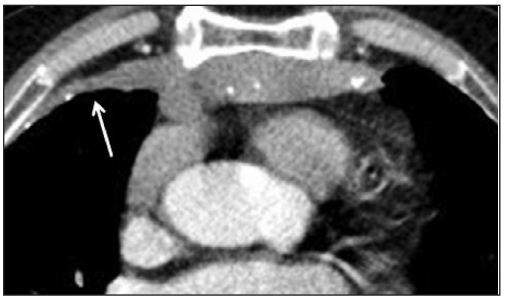

FIGURE 2: Retroternal solid lesion showed pleural extension to the right and epicardial fat plan is preserved.

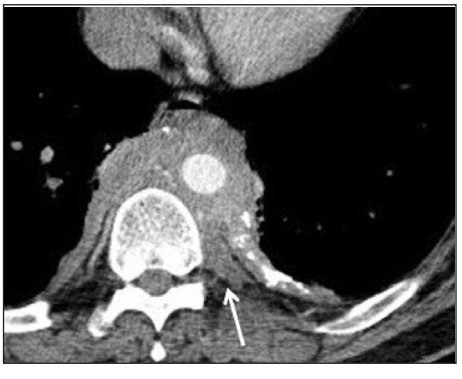

FIGURE 3: The thoracic aorta is wrapped around with the mass lesion, the left pre-vertebral fat tissue is obliterated and oesophagus is replaced to anterior. On both sides, pleural thickening continuing with the mass on both sides and calcifications on the left are seen.

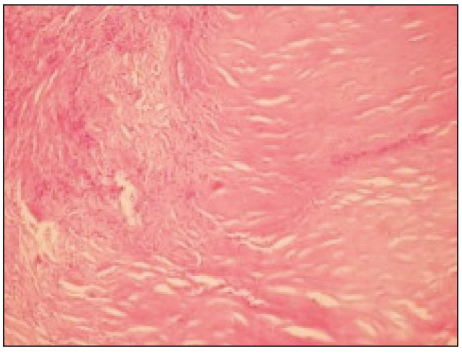

FIGURE 4: A 62-year-old male patient who underwent tru-cut biopsy from a mass in the anterior mediastinum; High-power photomicrograph (original magnification, H&E, x40) demonstrates acellular hyalinized fibrotic tissue.

calcifications were observed in the lesions. Diffuse and mild contrast enhancement was observed. There was no calcific plaque or thickening on other pleural surfaces, and no pathology was seen in the lung parenchyma. Sedimentation and acute phase reactants were normal. Leukocytosis was not detected. Respiratory function tests were routine. Tru-cut needle biopsy was performed on the anterior mediastinal mass in the case under examination for differential diagnosis in pleural malignancy, thymoma, or lymphoma. Acellular hyalinized fibrotic tissue was seen microscopically (Figure 4). The patient, who was not considered for surgical treatment, was followed up with CT because of the absence of pressure findings and clinical signs. In the 8-year follow-up process, focal thickening developed as a particular focus on the right apical pleura, but other lesions and clinics were stable. Approval of the patient was obtained for this case report.

CASE 2

The second patient was a 45-year-old male patient presenting with chest pain; he had a history of untreated pleural thickening followed up for approximately five years. A retrosternal, non-calcareous solid mass lesion reaching 3 cm thickness with antero-posterior orientation was observed on the chest CT scan. The lesion had a typical "batwing" shaped extension on the anterior costal pleura, and bridging between the two pleural sides attracted attention (Figure 5). There was no invasion of the vascular structures and pulmonary parenchyma. We saw no accompanying mediastinal lymphadenopathy. A small number of bands were observed in the pulmonary parenchyma. Also, a second solid mass lesion with lobulated contour and clear boundaries showing pleural extension was observed in the posterior mediastinum on the left lateral side of the aorta but not circling the vascular structure (Figure 6). The laboratory findings of the patient did not reveal any pathology except a slight increase in sedimentation rate. The patient underwent a left pleural decortication operation. Histopathological examination of the surgical biopsy specimens of the pleural mass showed pleuritis and hyaline plaque (Figure 7). There was no progression seen during the 6-year follow-up. We obtained the approval of the patient for this case report.

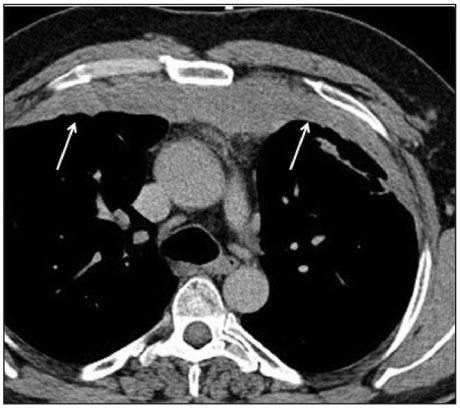

FIGURE 5: Retrosternal mass lesion showing typical "bat wing" shaped pleural extension (or as previously described "bridging fibrosis"). Anterior mediastinal fat tisue is fully preserved.

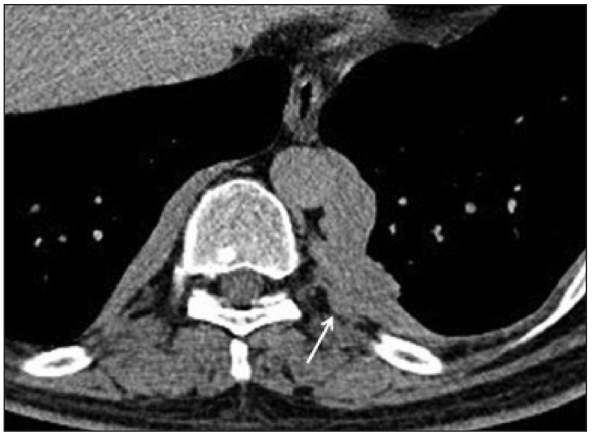

FIGURE 6: Bilateral posterior asymmetric pleural thickening and left paraaortic mass lesion with lobulated margins is seen. Extrapleural fat tissue obliteration is more evident on the left.

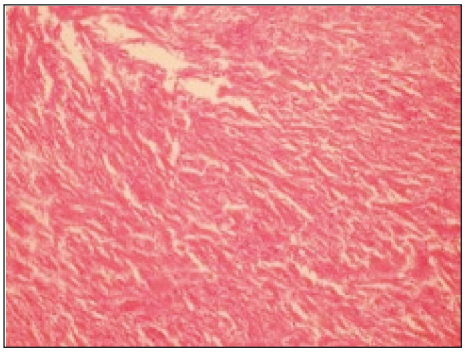

FIGURE 7: A 45-year-old male patient who underwent pleural decortication; pleuritis, and hyaline plaques were observed in the high-power photomicrograph (original magnification, H&E, x40).

DISCUSSION

In both cases, granulomatous infection findings such as tuberculosis or histoplasmosis, or autoimmune disease in another system were not observed. An autoimmune disease was detected in 32% of 84 cases of mediastinal fibrosis published between 2006 and 2016.[3] Multifocal fibrosclerosis, a syndrome characterized by fibroinflammatory involvement in multiple organs and systems, and associated with IgG4, is also included in the differential diagnosis because of its multicentric nature. In this disease, in the RPF series of 491 cases, the mediastinal involvement was only 3.3%.[7] In our cases, RPF is also not seen.

In contrast to what is seen in some cases of granulomatous mediastinitis, stricture is not observed in the trachea, esophagus, and vena cava superior. Lesions are retrosternal and posterior mediastinal. Thymic neoplasms and lymphoma are the first to come to mind in anterior mediastinal mass lesions, and histopathological diagnosis is required. Mediastinal fibrosis secondary to malignancy has a poor prognosis, then differentiation of benign and malignant fibrosis is essential. However, the distinction between benign and malignant fibrosis may be difficult with CT.[1,13] Tissue biopsy was performed on both of our patients. In both cases, it is observed that retrosternal solid lesions do not invade vascular structures and lung parenchyma and are non-progressive during follow-up. Stability or slow progression is an essential finding for benign etiopathogenesis.

Asbestosis constitutes an important group in benign pleural thickening etiology.[10,14] The cases have a history of asbestos exposure. Diffuse pleural thickening, which shows continuity in both hemithorax with the mass, creates a different appearance from asbestosis, pleural metastasis, or mesothelioma. It is also an essential detail that, in the first case, preserved the internal mammarian artery and vein against the aggressive extensor extension on the right. In both cases, a bilateral pleural extension of the retrosternal mass in the form of a "batwing" creates a typical appearance. This appearance was defined as "bridging fibrosis" in 2 cases exposed to asbestos in the literature.[10] The association of mediastinal and pleural fibrosis in asbestosis is very rare in the literature.[10]

The mass lesion surrounding the aorta in the posterior mediastinum suggests infiltrating malignant processes, primarily lymphoma or metastasis. Szarf and Bluemke, as a part of intrathoracic periaortic fibrosis, multifocal fibrosclerosis and mediastinitis in 2005, and Lee et al. published with fluorodeoxyglucose positron emission tomography and CT findings as an isolated mass that cannot be distinguished from malignant lesions without mediastinitis, and emphasized that the lesion may be malignant due to high standardized uptake value and narrowing of the lumen in the aorta.[11] Vertebral column or esophagus invasion and stricture in the aorta were not observed in our cases. An important finding is that the lesions are clinically and racially stable for 4 and 8 years without the need for surgery.

In intrathoracic fibrosis, CT is the primary radiological method in detecting lesions, in their exact localization, in investigating other parenchymal and mediastinal findings that may contribute to differential diagnosis and planning treatment is pressure, and in follow-up.[1,6,8,9] However, because it mimics many other diseases, it is made by histopathology for definitive diagnosis. In our cases, the biopsy was performed from the retrosternal component, and an invasive procedure for diagnostic purposes was not considered for the periaortic area. The cases show that CT imaging is sufficient for the follow-up of the disease.

In conclusion, this presentation is valuable because it is the first case of benign intrathoracic fibrosis with 6 and 8 years of CT follow-up, involving the

anterior and posterior mediastinum and pleura simultaneously, without RPF.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Seher Susam, Kenan Can Ceylan;* ***Design:*** *Seher Susam, Kenan Can Ceylan, Ceyda Anar;* ***Control/Supervision:*** *Seher Susam, Kenan Can Ceylan, Ceyda Anar, Nur Yücel;* ***Data Collection and/or Processing:*** *Kenan Can Ceylan, Nur Yücel, Ceyda Anar;* ***Analysis and/or Interpretation:*** *Seher Susam, Funda Cansun Yakut, Nur Yücel;* ***Literature Review:*** *Seher Susam, Nur Yücel, Funda Cansun Yakut;* ***Writing the Article:*** *Seher Susam Funda Cansun Yakut;* ***Critical Review: References and Fundings:*** *Kenan Can Ceylan.*

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