A case of endobronchial inflammatory pseudotumor invading the mediastinum

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ÖZET

Mediasteni invaze eden endobronşiyal inflamatuvar psödotümör olgusu

İnflamatuvar psödotümör, tüm akciğer tümörlerinin %1'den daha azını oluşturur. Endobronşiyal lokalizasyon, olguların % 12'sinden daha azında görülür. Bu tümörler mediastinal yapılara invazyon gösterebilir. Bu yazıda, mediastene invazyon gösteren endobronşiyal inflamatuvar psödotümör olgusunu sunduk. Elli yaşında erkek hasta, üç aydır devam eden öksürük ve kanlı balgam yakınmaları ile Mart 2007 tarihinde merkezimize başvurdu. Arka-ön akciğer grafisinde sağ üst zonda opasite saptandı. Sigara alışkanlığı dışında öz geçmişinde bir özellik yoktu. Tam kan sayımı, idrar tahlili ve biyokimyasal incelemeler normal sınırlarda idi. Bilgisayarlı toraks tomografisinde sağ üst lob ile sağ alt lob superior segmenti tutan ve mediastene invazyon gösteren kitle rapor edildi. Fiberoptik bronkoskopi incelemesinde sağ üst lob girişini tamamen kapatan kitle ve ara bronşa uzanım gösteren infiltrasyon görüldü. Bronkoskopik biyopsiler tanısal değildi. PET-BT incelemesinde SUDmaks değeri 18.8 olarak ölçüldü. Solunum fonksiyon testinde FEV_1 2.52 L (%70) ve FVC 2.94 L (%66) idi. Hastaya sağ torakotomi uygulandı. Torakotomide, kitlenin vena kava süperior ve pulmoner arteri invaze ettiği saptandı. Biyopsi örnekleri alındı. Patolojik tanı inflamatuvar psödotümör olarak rapor edildi. Kortikosteroid tedavi başlandı. Klinik ve radyolojik yanıt alınamayan hasta radyoterapi amacı ile onkoloji kliniğine sevk edildi. Hasta Kasım 2007 tarihinde öldü.

Anahtar Kelimeler: *İnflamatuvar psödotümör, akciğer, mediasten, invazyon.*

SUMMARY

A case of endobronchial inflammatory pseudotumor invading the mediastinum

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A 50-year-old-male was admitted to our hospital in March 2007, complaining of cough and hemoptysis for 3 months. Postero-anterior chest X-ray showed an opacity on right upper zone. Computed tomography of the thorax showed a mass lesion occupying the right upper lobe and superior segment of the lower lobe and invading the mediastinum. Fiberoptic bronc-

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hoscopy showed total occlusion of the right upper lobe bronchus by the mass and infiltration of the bronchus intermedius. Bronchoscopic biopsies were nondiagnostic. PET-CT revealed SUVmax of 18.8. Right thoracotomy was performed. Vena cava superior and right pulmonary artery was invaded by the mass. Biopsies were performed. Histopathologic examination demonstrated an inflammatory pseudotumor. Corticosteroid treatment was started. The tumor was clinically and radiologically unresponsive to corticosteroids. He was referred to oncology department for radiotherapy. The patient died on November 2007.

Key Words: *Inflammatory pseudotumor, lung, mediastinum, invading.*

Inflammatory pseudotumor is an uncommon, benign, essentially nonneoplastic, tumor-like process that usually occurs in children and young people (1). It consists of mixed components of collagen fibers, inflammatory cells, and mesenchymal cells (2). Since the initial report of this lesion by Brunn in 1939, it has also been called as plasma-cell granuloma, histiocytoma, xanthoma, xanthofibroma, xanthogranuloma, solitary mast-cell granuloma, fibrohistiocytoma, postinflammatory tumor, but the most commonly used terms include plasma-cell granuloma and inflammatory pseudotumor (3,4). It most commonly presents as a solitary pulmonary nodule, and the endobronchial presentation is uncommon (5). Although regarded as a benign lesion, it may have aggressive behavior (6). We presented a case of endobronchial inflammatory pseudotumor invading the mediastinum.

CASE REPORT

A 50-year-old male presented on march 2007 with a three-month history of cough and hemoptysis. Posteroanterior chest X-ray showed an opacity on right upper zone. His past medical history was unremarkable except smoking. Complete blood count, urinalysis and blood biochemistry were normal. Computed tomography (CT) of the thorax revealed a mass lesion that invades right pulmonary artery and pulmonary vein and occupies the right upper lobe and upper segment of the lower lobe and measures 10.5 x 9.5 x 11 cm (Figure 1). Fiberoptic bronchoscopy showed total occlusion of the right upper lobe bronchus by the mass and infiltration of the bronchus intermedius (Figure 2). Bronchoscopic biopsies were nondiagnostic. PET-CT revealed SUVmax of 18.8 (Figure 3). FEV_1 was 2.52 L (70%) and FVC was 2.94 L (66%). Right thoracotomy was performed. Vena cava superior and right pulmonary artery was invaded by the mass. Biopsies were performed. Histopathologic diagnosis was an inflammatory pseudotumor. Corticosteroid treatment was started. The tumor was clinically and radiologically unresponsive to corticosteroids. He was referred to oncology department for radiotherapy. The patient died on November 2007.

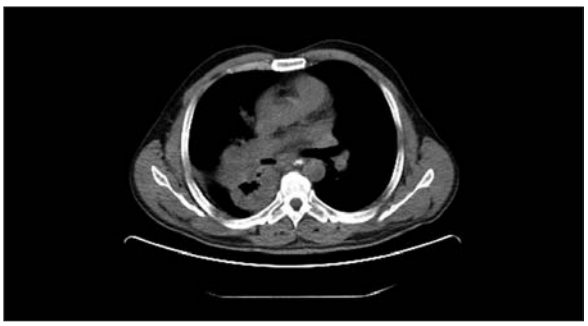

Figure 1. Computed tomography of the thorax shows a mass lesion that invades right pulmonary artery and pulmonary vein and occupies the right upper lobe.

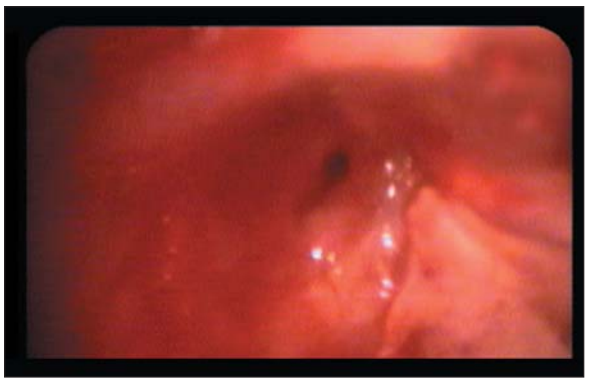

Figure 2. Fiberoptic bronchoscopy shows total occlusion of the right upper lobe bronchus by the mass.

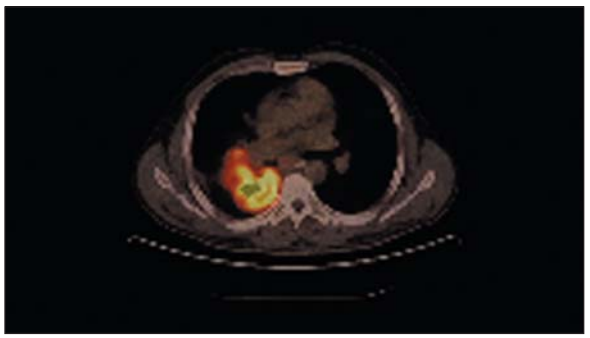

Figure 3. PET-CT reveals SUVmax of 18.8.

DISCUSSION

Inflammatory pseudotumor is a nonneoplastic process characterized by unregulated growth of inflammatory cells. Although its exact pathogenesis is not known at present, two opinions are held: one is that it is tumorous in origin because it presents with few symptoms, has tumorous expanding growth, and has a 10% occurrence rate. The other opinion is that it is inflammatory in origin, because it presents with intraalveolar organic lesions, a past history of respiratory tract infection, and it consists of a variety of inflammatory cells (7). A history of upper respiratory tract infection or pneumonia was reported in 28% of patients (8). This tumor can be associated with trauma, paraneoplastic syndrome or other inflammatory reactions (8,9). There was not any history of trauma or infection in our patient.

Exact incidence of pulmonary inflammatory pseudotumor is not known. It is reported to be 0.04-1.2% of all pulmonary tumors (8). Inflammatory pseudotumor of the lung is not sex related. The patients vary greatly in age (1-77 years), about 60% of patients are younger than 40 years of age, and 15% are between the ages of 1 and 10 years (1,10). Melloni et al. reported that all the patients in their series were adults and only 16% were younger than 40 years (8). We presented an older male patient. He had pulmonary symptoms including cough and hemoptysis. Most patients with inflammatory pseudotumor are asymptomatic and tumor is discovered by incidental findings on chest radiographs. However, 26-56% of these patients have been reported to show clinical symptoms such as cough, hemoptysis, dyspnea and chest pain (1,7). Symptoms are strictly related to lesion location: paranchymal or endobronchial (8). The endobronchial presentation is uncommon, accounting for less than 12% of the cases, and constitutes the most symptomatic form, which frequently leads to an early diagnosis (5,10). Our patient had endobronchial presentation.

Radiologically, the tumor usually presents as a well-circumscribed, solitary, round or ovoid peripheral pulmonary nodule or mass measuring 1 to 6 cm in diameter (10). Agrons and coworkers analyzed chest X-ray features in 60 patients with pulmonary inflammatory pseudotumor (11). Fifty-two (87%) patients had solitary peripheral lesions, of which 31 were a masses and 21 were a solitary pulmonary nodule. Three (5%) patients had multiple nodules. Secondary infiltration of hilum, mediastinum, and airways is proven in 16% of patients. In a previous report, CT of the thorax revealed well-circumscribed parenchymal lesions (nodules or masses) without signs of infiltration in 15 patients, whereas 3 patients presented an infiltration of the adjacent pleura (8). Radiographics can demonstrate calcification, cavitation, notch, spicula and pleural indentation in tumor but these features are unusual (1,7). In our case, CT showed a mass lesion 11 cm in diameter. There was no calcification in the tumor. PET-CT examination identified a high standardized uptake value. Higashi et al. reported that FDG PET showed a false-positive result in a case of inflammatory pseudotumor (12). Inflammatory pseudotumors are difficult to diagnose. Preoperative diagnostic methods such as percutaneous fine needle biopsy and bronchoscopy is considered insufficient for diagnosis. The diagnosis is made by surgical procedures such as video-assisted thoracoscopic surgery or open lung biopsy (1,7,8). In our patient, bronchoscopic biopsies were nondiagnostic, we established the diagnosis of inflammatory pseudotumor with open lung biopsy.

Although pulmonary inflammatory pseudotumors are regarded as a benign lesion, they may have aggressive features, and they may encase bronchi or invade chest wall, mediastinal structures, the spine and vessels. Multiple recurrences and metastatic spread can also be seen (6,10,11,13). Corneli et al. reported that the tumor invaded the mediastinal pleura, the pericardium, the esophagus, the thoracic aorta and the diaphragm (6). The treatment of choice of these tumors is surgery. Wedge resection, if radical, is suitable for curative purposes. When wedge resection is not feasible, the lesion is removed with major resections such as lobectomy or pneumonectomy. Nonsurgical treatment modalities including radiotherapy, chemotherapy, and steroids may have a place in the setting of incomplete surgical resection, multifocal disease, tumor recurrence, or contraindication to lung resection (6,8,10,14). The prognosis of patients with completely resected inflammatory pseudotumors is excellent: 78% to 100% of patients were in complete remission after an average follow-up of 3.3 years after surgical resection. Intrathoracic recurrence was seen in 5% of cases (10). The tumor may be fatal. Bahadori and Liebow reported the fatal outcome of a patient with involvement of esophagus and superior vena cava (15). In our case, the tumor invaded the mediastinum. Because the patient was inoperable, he was treated with corticosteroids followed by radiotherapy. The tumor was aggressive and our patient died 8 months after diagnosis.

In conclusion, pulmonary inflammatory pseudotumor is rare. The preoperative diagnosis of this tumor is difficult. Although it is considered as benign, it may have aggressive behavior and poor prognosis.

CONFLICT of INTEREST

None declared.

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