A relapsing case of Wegener's granulomatosis presenting as an endobronchial mass

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ÖZET

Endobronşiyal kitle gibi nükseden Wegener granülomatozis olgusu

Wegener granülomatozis (WG) nüksü sıktır. Akciğer tutulumu hastaların %85'inde meydana gelmesine rağmen, endobronşiyal lezyon yaygın değildir. Biz, endobronşiyal kitleyle seyreden bir WG nüks olgusunu sunduk. Ellialtı yaşında erkek hasta 14 aylık siklofosfamid ve prednizolon tedavisini ve 36 aylık tam remisyonu takiben nüks olarak başvurdu. İlk hastalıkta böbrek, akciğer, üst solunum yolu, deri, eklemler ve göz tutulumlu olgu olarak tanı konulmuştu. Akciğer grafisi bilateral yama tarzında konsolidasyonlar gösteriyordu. Sitoplazmik antinötrofil sitoplazmik antikorlar (c-ANCA) yüksek titredeydi. Tedavi sonrası c-ANCA negatifleşti. Nüks zamanında, nazal yakınmalar ve hemoptiziyle başvurdu. Göğüs grafisi sağ parakardiyak bölgede yoğunluk artışı gösteriyordu. Fiberoptik bronkoskopi sağ alt lob girişini totale yakın kapatan kitle saptadı. Hastaya 10 aydır siklofosfamid ve azalan dozlarda prednizolon verildi.

Anahtar Kelimeler: *Wegener granülomatozis, nüks, endobronşiyal kitle.*

SUMMARY

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Wegener's granulomatosis (WG) relapse is frequent. Although lung involvement occurs in 85% of patients, endobronchial presentation of the disease is uncommon. We reported a relapsing case of WG presenting as an endobronchial mass. A 56-year-old man present ed with recurrence of WG following 14 months of cyclophosphamide and prednisolone therapy and 36 months of complete remission. At his first presentation, he was diagnosed as having WG with involvement of kidney, lung, upper airways, skin, joints and eyes. His chest X-ray showed bilateral patchy consolidation. Cytoplasmic-anti-neutrophil cytoplasmic antibodies (c-ANCA) was also present in high titres. c-ANCA was negative after therapy. At the time of relapse, he presented with nasal symptoms and hemoptysis. His chest X-ray showed right paracardiac opacity. Fiberoptic bronchoscopy revealed a mass lesion subtotally obstructing the proximity of right lower lobe. He has been given prednisolone in tapering doses and cyclophosphamide for 10 months.

Key Words: *Wegener's granulomatosis, relapse, endobronchial mass.*

Wegener's granulomatosis (WG) is a vasculitic syndrome which affects mainly the upper airways, lungs and kidneys (1). Other organs are commonly involved as well, including the joints, skin, eyes and nervous system (2). In WG, lung involvement occurs in 85% of patients (3). The usual radiological features of lung involvement include nodules with or without cavitation, infiltrates and diffuse alveolar haemorrhage (1,4-6). Endobronchial lesions are uncommon (1,5,7,8). Endobronchial involvement can be as tracheobronchial stenosis, bronchial wall thickening, ulcerations, hemorrhage, secretions and intraluminal masses (6,9).

The patients with WG had 18% of 5-month survival rate before the era of immunosuppressive therapy (2). Introduction of combination therapy with cyclophosphamide and glucocorticoids have dramatically improved the prognosis with 75% of complete remission rate (10). Relapses are frequent among patients with WG (11,12). A previous report pointed out a relapse rate of 50 percent (11). This paper presents a relapsing case of WG presenting as an endobronchial mass.

CASE REPORT

A 52-year-old man presented in January 2000, complaining of arthralgias and nasal symptoms for six months, cough and shortness of breath for two months and hemoptysis for 15 days. Medical history was unremarkable and he had been taking no medications. Physical examination showed red eyes, oral aphtosis and mycotic lesions, and purpuric lesions on bilateral lower extremites. His axillary temparature was 36.8°C, blood pressure was 115/70 mmHg, pulse was 94 beats per minute, respiratory rate was 21 per minute. Haemogram showed a white blood cell count of 14300 and haemoglobin of 11.6 g/dL. Westergren erythrocyte sedimentation rate was 80 mm/hour. Serum urea level was 65 mg/dL and creatinine value was 2.2 mg/dL. Urinalyses revealed proteinuria and microscopic hematuria. Chest X-ray showed bilateral patchy consolidation pattern. Computed tomography (CT) of the thorax demonstrated bilateral patchy consolidation pattern and nodules with sparing of the apices (Figure 1). Bronchoscopic examination detected minimal hemorrhage in left upper lobe bronchus. There was no endobronchial mass. Renal ultrasonography was normal. Skin biopsy revealed a diagnosis of leukocytoclastic vasculitis. c-ANCA was present in high titres. He was treated with prednisone in tapering doses and cyclophosphamide, 150 mg daily for 14 months. He also received trimethoprim-sulfamethoxazole one double strength daily. Therapy improved symptoms between 10 days and 30 days. Serum urea and creatinine levels and urinalysis were within normal levels two months after initiation of therapy. Computed tomography of the thorax detected that pul-

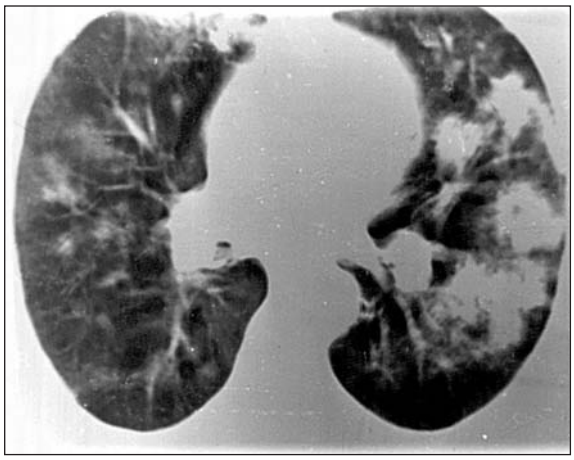

Figure 1. At the time of initial presentation, CT showed bilateral patchy consolidation and nodules.

monary lesions disappeared completely without scarring (Figure 2). c-ANCA was found to be negative three months after therapy.

The patient had 36 months of complete remission before relapse. At the time of relapse, he had presented with nasal symptoms, cough, hemoptysis and shortness of breath for 10 days. His physical examination was unremarkable. CBC and urinalysis was normal. Westergren erythrocyte sedimentation rate was 50 mm/hour. His chest-X ray showed right paracardiac opacity. CT demonstrated opacity on right lower lobe (Figure 3). c-ANCA was present in high titres. Fiberoptic bronchoscopy detected mass lesion subtotally obstructing the proximity of right lower lobe (Figure 4). Bronchial lavage, endobronchial needle aspiration biopsy and bronchoscopic biopsies were performed. Bronchial lavage was negative for tuberculosis bacilli and fungi. Pathological examination did not detected a malignancy. Biopsies showed fibrinoid necrosis, geographic necrosis, microabscesses without granulomas, epithelioid histiocytes and neutrophils. These features are major manifestations of pulmonary WG (2). There was not any features of vasculitis. He has been given prednisolone in tapering doses and cyclophos-

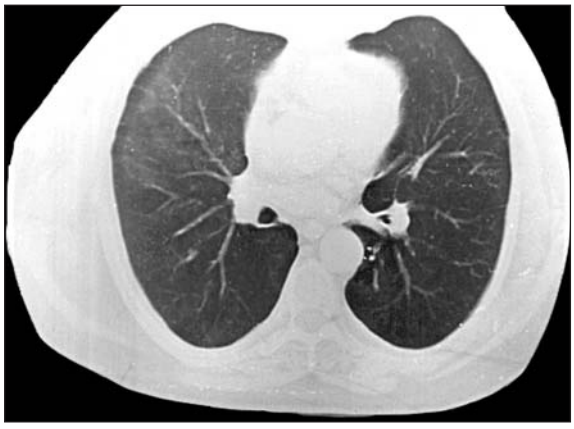

Figure 2. At the time of initial presentation, CT demonstrated that consolidation and nodules disappeared after therapy.

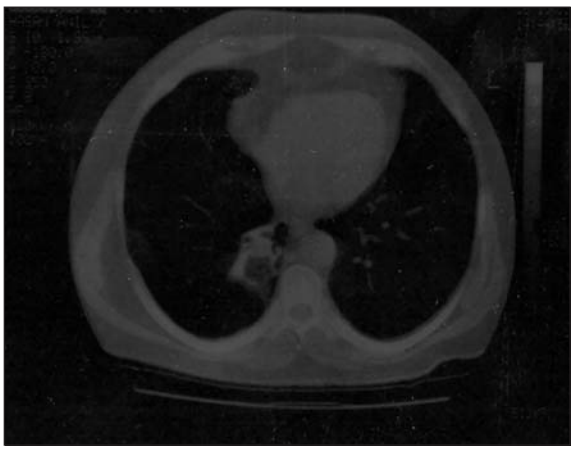

Figure 3. CT shows opacity on right lower lobe at the time of relapse.

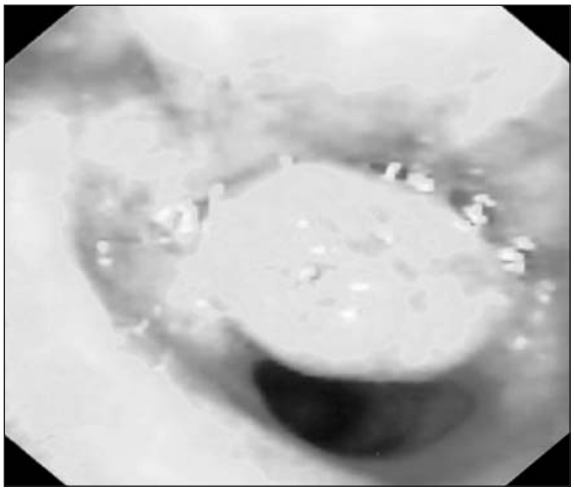

Figure 4. Bronchoscopic examination shows a mass lesion obstructing the proximity of right lower lobe bronchus.

phamide for 10 months. Therapy improved nasal symptoms, cough and hemoptysis within one month. c-ANCA was negative after two months of therapy. Nine months after therapy, bronchoscopic reexamination was performed and it showed no regression or progression in lesion size.

DISCUSSION

The disease now known as WG was first described by Klinger in 1931, followed by other investigators including Rossle in 1933, Wegener in 1936 and 1939, and Ringertz in 1947 (13). Before 1950s therapy was only supportive. Untreated patients have had poor prognosis with a median survival of five months. While the use of prednisolone reduced mortality, the median survival remained at only 12.5 months. Combined treatment with cyclophosphamide and glucocorticoids revealed complete remission in 93 percent of patients, with a mean duration of 48 months (4,14). Relapse is frequent among patients with WG. Langford et al. reported that 52 percent of patients had relapsed (15).

We report a relapsing case of WG. The diagnosis of WG was based on ELK classification system and c-ANCA positivity in both presentations (2). At the time of initial presentation, the patient had upper airways, lung, kidney and other system involvement such as skin and joint. At the time of recurrent disease, there were symptoms of the upper airways and lung involvement, but not the kidney. We observed that the relapsing case has many uncommon features. First, the interval between cessation of combination therapy and relapse was 36 months. A previous report pointed out that this interval is an unusual feature (16). Although relapses are frequent among patients with WG, they generally occur during the tapering of immunosupressive therapy (16,17). Fauci et al. reported that 33 percent of patients achieving an initial remission relapsed, all during withdrawal of medication (18). In other series, authors reported a relapse rate of 50 percent but none with delayed recurrence (11). On the contrary, longer intervals such as 20 years have been published (19).

Second, late relapses generally have the similar clinical and radiological presentation as the initial presentation (1,19). At the time of initial presentation, our patient had symptoms or features of involvement of the upper airways, lung, kidney and other systems. At the time of relapse, the patient had pulmonary and nasal symptoms. There were no features or symptoms of involvement of kidney, skin, joints and eyes. The chest X-ray findings were different at recurrence than at the time of initial presentation. In first presentation, CT demostrated bilateral patchy consolidation and nodules. At the time of recurrence, CT scan showed opacity on right lower lobe. There were no patchy consolidation or nodules on CT scan.

Third, our patient relapsed at a site which was not involved during the first disease episode. The relapsing case had different lesion type compared to initial presentation. At the time of first presentation, lesions were bilaterally located and were parenchymal. At the time of relapse, the lesion was located in right lower lobe bronchus and there were no parenchymal lesion such as nodules and patchy consolidation. Previous reports pointed out that relapses occurred in previously affected areas (1,16,17). In initial presentation, the patient had usual pulmonary lesions including consolidation and nodule. Our patient relapsed with an endobronchial mass. Endobronchial lesions are uncommon findings of WG (6,9).

In conclusion, we reported a relapsing case of WG following 36 months of complete remission. Our patient had uncommon features. At the time of relapse, patients may have different clinical and radiological features compared to initial presentation.

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