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CASE REPORT

Presentation of a Behcet case with lung malignancy: a case report

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ABSTRACT
The patient to be presented here was A 44-year-old man who was admitted with the muscle weakness, cough, weight loss, and dyspnea. He was a known case of dermatomyositis since 4 months ago. The characteristic features of his condition included with the purpuric cutaneaous scaling rash, heliotrope rash Gottron's papules, shawl sign, dyspnea and 25 kg weight loss. Thoracic computed tomography revealed ground glass opacities and pneumomediastinum. The patient was diagnosed with dermatomyositis. pneumomediastinum, pneumothorax and subcutaneous emphysema were observed as well. Despite intensive immunosuppressive therapy, clinical deterioration and radiological progression were observed and the patient died.

Key words: behcet disease, lung, malignancy

INTRODUCTION

Rheumatic disorders (RDs) consist of a wide group of systematic conditions with multiple organ damage that could have associations with cancers and neoplastic or paraneoplastic conditions. They are three theories concerning the association between autoimmune diseases and cancer:[1]

1) A rheumatic disorder is directly triggered by a tumor or its metastatic arthritis due to synovial infiltration by leukemic cells;

2) Patients with chronic idiopathic rheumatic disorder who develop cancer within a temporal interval of up to many years, for example Sjögren's syndrome, can be developing lymphoma;

3) Patients with clinical manifestations of a rheumatic disorder, which is actually the expression of an occult cancer that becomes clinically evident within months or years—called paraneoplastic rheumatic disorders.

Behcet's disease (BD) is a chronic relapsing systemic vasculitis, defined by disease manifestations such as urogenital ulcers, uveitis, skin lesions and arthritis, and involvement of the central nervous system, the gastrointestinal tract, and blood vessels.
It is now recognized as a multi-system inflammatory disorder characterized by vasculitis treatment of BD usually consists of systemic corticosteroids and immunosuppressive and/or cytotoxic agent.[2, 3]
However, only a few case reports and case series are available concerning the relationship between malignant disease and BD in the relevant literature.4, 5
Here, we report a case with BD and SVC syndrome that developed solid malignant tumor two years after diagnosis.

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CASE REPORT

A 45-year-old male presented with pain and progressive swelling in his neck and face for two weeks. He was a chronic smoker for the 10 years. He complained of oral aphthus lesion recurrently two times a year but not genital ulcer. He also had a history of solitary papule on both legs with healing now and hyperpigmentatat site without any scar.
His physical examination showed facial puffiness, non-pulsatile engorged neck veins, conjunctival redness, and dilated superficial veins over his chest. He was admitted in the rheumatology ward for evaluation. Doppler ultrasonography of the neck venous and arterial system showed the thrombus in right jugular vein with lymphadenopathy and round shape at both side of neck.
Contrast-enhanced computed tomography (CT) scan of the thorax revealed filling defect on the jugular and subclavian vein with extension to the brachiocephalic vein. SVC diameter was less than the normal size and showed collateral branch around SVC.
In the lower lobe of the right lung were fibrotic band and atelectasis. Oral and intravenous (IV) contrast-enhanced CT of the abdominal pelvic revealed bilateral multiple cortical cysts in the kidneys.
The two-dimensional echocardiography showed nor-

mal left ventricle (LV) cavity size and mild mitral and tricuspide regurgitation. Left ventricular ejection fraction was 50% with increased thickness of the anterior aspect of the mitral valve.

He had normochromic normocytic anemia and microscopic hematuria in the lab data, while other parameters were normal. SVC syndrome was found on CT angiography.

With impression of Behcet's disease and superior Vena Cava syndrome, the patient started consuming 1mg/kg per day prednisolon and anticoagulant followed by IV cyclophosphamide 375mg/m2 monthly.

The patient was discharged with warfarin and prednisolon and cyclophosphamide monthly for six months.

In the follow-up, he received 6 doses of 375 mg/m2 cyclophosphamide monthly, and prednisolon was tapered till 10 mg/day. Then, azathioprine 50 mg/day started for him, oral warfarin was continued 5 mg daily, and he was advised to have INR level check regularly.

Six months after the last dose of cyclophosphamide, he presented with fever and productive cough with dyspnea whereby he was admitted in the rheumatology ward with severe leukocytosis and shift to the left (WBC=21000 with 91% PMN; Hb=9/2, ESR=73, CRP=22). Blood and urine cultures were negative and other lab data were normal.

A chest radiograph showed bilateral air bronchogram and lesions in the upper lobe of the right lung for which the high-resolution CT scan revealed consolidation. Fibroptic lung bronchoscopy were performed which proved normal. Culture of the alveolar lavage was negative for bacteria as were acid fast bacilli and nocardia in smear and culture.

After 2 weeks of IV cephalosporin and vancomycin use, he was discharged with oral cefecxim and 10 mg/day prednisolon along with 100 mg azathioporin and 5 mg daily warfarin with controlled INR.

He referred back after one month with chills and fever and hemoptysis .

The lab data count was 15700 with PMN=90%, Hb=10/6, ESR=69, and CRP=130.

Vancomycin and meropenem IV were prescribed and chest radiography revealed an irregular cavity in the upper lobe of the right lung. Sputum smear and culture were repeated which were negative for acid fast bacilli and bacteria.

Fine needle aspiration of the lung lesion showed non-specific inflammation of the interstitial tissue without evidence of vasculitis. Bronchoscopy was reconsidered whose result was normal, and BAL cytology was normal and negative for malignancy.

High resolution CT scan of the lung showed fibrocavitary lesion in the right upper lobe and a single nodule of 8 mm diameter in the left upper lobe.

The patient was discharged since he was in good condition in the infectious diseases ward. Oral levofloxacin with other drug and rheumatology consultation were prescribed.

After 8 months, he complained of progressive dyspnea whereby lung CT scan was performed for him spiral. It showed fibrosis with upper right lobe bronchiectasis and fibrotic band in sub segmental collapsed area at both posterior and lower lung lobes accompanied by multiple nodules with spiculated nodules with the mean diameter 8mm without calcification in the left upper lung lobe. He was referred to a cardiothoraxic surgeon

The differential diagnosis by the pathologist was granuloma or neoplasia.

High resolution CT scan was repeated one month later which depicted an increase in the nodule's diameter from 8 mm to 1.4 cm with an irregular and spiculated border. In addition, there was a consolidation in the lower right lobe that was adhered to the plural.

The patient underwent bronchoscopy for a third time, whose results were normal and PCR of the BA fluid for TB was negative.

Levofloxacin and azathioprine along with prednisolon were continued for three weeks. After the last bronchoscopy, however, he developed deep vein thrombosis in the right leg on oral warfarin with WR=2.5.

He referred for open lung biopsy where histopathological report showed invasive adenocarcinoma. He referred to the oncologist who started chemotherapy for him with IV cisplatin and etoposide on a monthly basis.

DISCUSSION

The frequency of malignancy, especially solid cancer, associated with BD is nearly 100 cases across the world, a relatively low prevalence as compared with the prevalence of other autoimmune diseases. A study at the school of medicine of Ajou University in Korea evaluated cancer morbidity in BD patients compared with the general Korean population. It was found that out of 506 patients with BD, 11(2.17%) developed cancer.[6]

This study and other case series studies6, 7 reported that BD may be associated with a lower cancer-related morbidity compared with other auto-immune diseases.5[-7]

According to the literature, BD is substantially linked with hematologic malignancies, particularly myelodysplastic syndrome.8, 9

Our patient is a case of Behcet's disease according to ICBD criteria consisting of oral aphtus lesion, positive pathergy test, vascular thrombosis, and skin manifestation. With 5 scores, he was a definite case of Behcet's disease.

Lung involvement was the typical finding which presented initially with fibrosis, extended to consolidation, and converted ultimately to cavitation. He developed fever, dyspnea, and hemoptysis. Several instances of HRCT and fiberoptic bronchoscopy did not show malignant lesion. Results of smear, culture, and BAL cytology were negative for infection and

malignancy.

Since the patient was treated with prednisolon and azathioprine, the infection was on top of the list for differential diagnosis for lung problem. This was evaluated with direct smear, microbial culture, and PCR of TB. Although all were negative, he received several courses of wide IV and oral antibiotics. However, there was no permanent and effective response after nearly 11 months. A solitary nodule in HRCT in the right upper lobe of the lungs was found which grew in size. Another solitary nodule in spike shape was also found in the left lung a month later.

Invasive adenocarcinoma was reported after open long biopsy and histopathological examination.

The first presentation of this patient was SVC syndrome. Over 85% of SVC syndrome cases are caused by malignant diseases among which the lung cancer is responsible for 60% of the cases. Most of these can be seen in small cell carcinoma cases followed by squamous cell carcinoma, adenocarcinoma, and large cell carcinoma.[6]

SVC syndrome may be due to hypercoagulable state, pulmonary arteriovenous malformations, and Behcet's disease.

CONCLUSION

It is not well defined whether the SVC syndrome in our patient was associated with Behcet's disease or lung cancer, although lung nodules appeared after 10 months of SVC syndrome presentation, and the patient already had clinical criteria of Behcet's disease for many years. This case was interesting for the rare presentation of Behcet's disease albeit in Iran, we find very omnifarious presentations of Behcet's disease for the high incidence and prevalence of BD.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interest.

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