Exudative pleural effusion associated with Behcet's disease

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Abstract

Background: Behcet's disease is a rare immune mediated small vessel systemic vasculitis in which diagnosis is mainly clinical and requires the presence of recurrent oral ulcers and at least two additional criteria that includes recurrent mouth and genital ulcers, skin lesions, eye inflammation (uveitis) and a positive pathergy reaction.
Methods: The case records of an 82 year old female Nigerian who presented to the medical outpatient clinic of the Federal Medical Centre, Yenagoa with a six months' history of recurrent oral ulcers, diarrhoea, arthralgia, blurring of vision, breathlessness, abdominal pain and extreme weakness was reviewed.
Results: She responded well to oral corticosteroids.
Conclusion: Behcet's disease should be given consideration in the differential diagnosis of pleural effusion in the presence of recurrent mucosal ulcers.

Keywords: Behcet's disease, Pleural effusion, uveitis, vasculitis

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Introduction

Behcet's disease was named after a Turkish Dermatologist, Hulusi Behcet, who first described the triple symptom complex of recurrent oral apthous ulcers, genital ulcer, and ocular problems in 1937[1].

The international study group for Behcet's disease proposed a separate set of diagnostic criteria for the disease. Based on these criteria, a diagnosis of Behcet's disease requires recurrent oral ulceration and at least two additional criteria, including recurrent general ulcers, ocular lesions, skin lesions and a positive pathergy test[1-3].
The determination of long term prognosis is usually difficult as the clinical course of the disease is variable[1].

Case Report

An 82 year old female Nigerian presented to the medical out-patient department of the Federal Medical Centre, Yenagoa in March 2013 with a six months' history of recurrent oral ulcers, intermittent loose, arthralgia blurring of vision, abdominal pains and extreme weakness. The oral ulcers (Figure 1) were described as very painful such that she was scared of eating. She had a mild cough, left sided pleuritic chest pain and scanty haemoptysis three weeks before presentation. The abdominal pain was non-specific, dull and involved both the lower and upper parts of the abdomen. The arthralgia mainly involved the knee and ankle joints. She had observed a progressive blurring of vision in the preceding six months. She denied history of fever but admitted to history of poor appetite and loss of weight. She had no haematemesis or malaena. She was not diabetic but was hypertensive for which she was on oral hydrochlorothiazide. She neither took alcohol nor smoked cigarettes.

Examination showed a chronically ill looking slim-built, elderly woman with low grade fever (temperature 37.4°C). She was pale, not jaundiced and had no peripheral lymphadenopathy or oedema. Cardiovascular examination showed a pulse rate of 92 beats per minute and her blood pressure was 160/100 mmHg. Jugular venous pressure was normal. The apex was heaving in the 5th left intercostal space but lateral to the mid-clavicular line with a mid-systolic murmur. The examination of the chest showed reduced chest movement in the left lower zone, stony dullness to percussion note, reduced tactile and vocal fremitus and reduced breath sounds in same zone. Examination of the abdomen, rectum and vagina were normal. Evaluation by an Ophthalmologist confirmed uveitis. Pathergy test was positive. A diagnosis of Behcet's disease complicated by anaemia and pleural effusion was made.

Result of laboratory investigations showed a PCV of 12%, WBC of 6.9 x 10^9/L, Neutrophils 52% lymphocytyes 32% eosinophils 4%. Peripheral blood film showed anisocytosis, microcytosis and hypochromic red cells. The erythrocyte sedimentation rate was 38mm/hour. Fasting blood sugar was 3.8mmol/L. HIV test was non-reactive. VDRL test and

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rhematoid factor were negative. Serum electrolytes, urea, creatinine were normal. Total serum protein was 59g/L, albumin 38g/L,LDH 98iu/L (normal range: 70-250iu/L). Urinalysis and stool examination were normal. Her chest radiograph showed an enlarged heart with unfolded aorta and a left sided pleural effusion. Echocardiography showed intact ventricular walls, interventricular and interatrial septae with good and regular cardiac contraction. The left atrium was marginally dilated but other cardiac chambers were normal in size. There was also evidence of gross calcification of the aortic valve with complete loss of valvular motion. M-Mode evaluation showed evidence of rigidity of the mitral value with flattening of its wave pattern-features suggestive of mitral valve stenosis. Other findings were normal. An ECG done was in keeping with long standing hypertensive heart disease.A pleural aspirate was straw coloured with a protein content of 36g/L (Serum protein 59g/L), LDH 199iu/L (serum LDH 98iu/L).The glucose content was 1.6mmol/L.The pleural fluid cytology, culture, gram stain and acid fast bacilli were negative. HLA-B51 assay was not done as the facilities were unavailable. All the sites where she had needle prick developed papules of about 2mm diameter after 48hours.This further confirmed a positive pathergy reaction.

She was started on oral prednisolone at 1mg/kg (50mg daily), transfused with 3units of packed cells.She was also treated with amoxicillin at 500mg thrice daily and her usual daily hydrochlorothiazide at 25mg daily orally. A repeat chest radiograph after 2 weeks of treatment showed a resolution of the left sided pleural effusion and the mouth ulcers had healed.The arthralgia and diarhoea had also resolved. She is currently on a maintenance dose of oral prednisolone at 10mg daily.

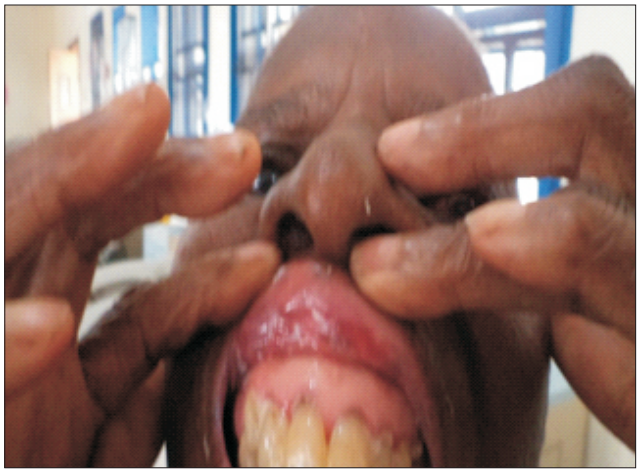

Figure 1. Upper lip showing ulcer with yellow necrotic base

Discussion

Behcet's disease is a rare, multisystemic, immune mediated, small vessel vasculitis affecting the mucocutaneous, ocular, cardiovascular, renal, gastrointestinal, pulmonary, urologic systems. The central nervous system as well as joints, blood vessels and lungs may also be affected[1].The cause and pathogenesis of the disease is not well defined, but the tumour necrosis factor alpha (TNF-alpha) pathway is most likely involved in its path-physiology[2].

Although chronic morbidity is usual and the leading cause is usually ophthalmic involvement, respiratory tract involvement has also been documented. The respiratory tract involvement is usually in the form of haemoptysis, pleuritis, cough or fever[3]. Pulmonary involvement also can take the form of nodules, consolidations, cavities and ground glass lesions[1]. Mortality is low, but patients can die from neurologic involvement, vascular disease, bowel perforation, cardiovascular disease or a complication of immunosuppressive therapy[1].

The disease is more prevalent in the Mediterranean basin and the Orient with an estimated prevalence of 1 case per 10,000 persons, and is strongly associated with the major histocompatibility complex (MHC) antigen HLA-B51.[2,4]. The prevalence of the HLA-B51 in Nigeria has been put at 2.6%[4] with males more affected than females and the peak age of presentation is in the third decade of life [1-2]. However, our index patient is a female Nigeria of Ijaw extraction in her eight decade of life. Similarly, two Nigerians (22-year old female and 31 year old male) presented with features suggestive of Behcet's disease in a report of case series of eight patients in the United Kingdom[5]. The closest findings to that of our patient is the report of pulmonary aneurysm and haemoptysis in an African with Behcet's disease[6].

Although the cause of Behcet's disease is unknown, it is however, widely acknowledged that immunogenetics, immune regulation, vascular abnormalities, bacterial or viral infection may play a role in its genesis and that HLA-B51 or its allele B101 is significantly associated with Behcet's disease[5]. In a study of MHC class 1 chain related gene A (MICA) and HLA-B allele in three Nigerian tribal population (Igbo, Efik and Yoruba) it was found that there was a high level of gene diversity[7]. Our patient did not benefit from HLA-B51 test as the facilities for this were not available.

Because of the absence of a pathognomonic laboratory test for Behcet's disease, the diagnosis of Behcet's disease is based on clinical criteria[1]. Our patient had mouth ulcers, arthralgia, bilateral uveitis and left sided exudative pleural effusion thus meeting the international study group criteria for the diagnosis of Behcet's disease[1]. Test for tuberculosis, syphilis, rheumatoid factor, retroviral (HIV) infection which are usually associated with systemic inflamination were all negative.

The main stay of treatment of Behcet's disease is corticosteroids[1]. The usual regimen is to start with high dose steroids and later low dose maintenance therapy as the inflammatory process is controlled. The usual dose is oral prednisolone 0.5-1mg/kg/day not exceeding 80mg. Since our patient did well on steroids, there was no need to use other modalities of treatment which include the use of immunomodulating drugs like azathioprine, chlorambucil and cyclophosphamide.

Conclusion

The report is to heighten the awareness of physicians of the possibility of this rare disease in our setting in the Niger Delta region where it is thought not to occur and that it should be given consideration in the differential diagnosis of pleural effusion in the presence of recurrent mucosal ulcers.

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