Pulmonary and Central Nervous System Involvement in Sweet’s Syndrome: A Very Rare Case Report

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Abstract

Sweet’s syndrome is a multisystem inflammatory disorder characterized by painful, erythematous plaques and aseptic neutrophilic infiltration of various organs. The absence of vasculitis is a histological criterion for diagnosis, but recent reports suggest that vasculitis can occur in Sweet’s syndrome. Involvement of the central nervous system and the pulmonary system is very rare. In this case study we describe a chronic alcoholic man with Sweet’s syndrome associated with acute-onset encephalitis and severe pulmonary involvement. The patient’s symptoms responded dramatically to steroid treatment, and notably, a skin biopsy of his lesions showed vasculitis.

Key words: sweet’s syndrome, neutrophilic dermatosis

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Introduction

Sweet’s syndrome was originally described as an acute febrile neutrophilic dermatosis. It is an uncommon, recurrent skin disease characterized by painful plaque-forming inflammatory papules. Histologically the striking feature is a dense dermal infiltrate composed of mature neutrophils with frequent nuclear fragmentation (1). Although absence of vasculitis is a previously reported histopathological diagnostic criterion, recent reports suggest that the presence of vasculitis should not exclude the diagnosis of Sweet’s syndrome (2). Pulmonary and central nervous system involvements (CNS) have rarely been reported. In this report, we describe the case of a chronic alcoholic man with Sweet’s syndrome with the involvement of the pulmonary and central nervous systems. His skin lesions showed vasculitic changes, a finding infrequently present in Sweet’s syndrome.

Case Report

A chronic alcoholic, 32-year-old man was admitted to our hospital with sudden onset of symptoms of fever, headache, cough with dark coloured sputum, myalgia and non-pruritic painful maculoerythematous skin lesions on his legs and arms (Fig. 1). On physical examination he exhibited somnolence, neck stiffness, and numerous pustules and erythematous skin plaques on the extremities. Focal neurological signs, episcleritis and uveitis were not present. He had an erythematous nodular eruption on the left periocular region (Fig. 2) and painful arthritis of the left ankle. Lumbar puncture was performed with suspicion of central nervous system (CNS) infection. Examination of the cerebrospinal fluid (CSF) revealed a white cell count of 80/mm³ (90% lymphocytes), a protein level of 43 mg/dL, and a glucose level of 71 mg/dL (the concomitant serum glucose level was 113 mg/dL). Laboratory values included a peripheral white blood cell count of 7,700/mm³ with 70% polymorphonuclear leukocytes, and 30% lymphocytes. The hemoglobin level was 13.5 g/dL, the erythrocyte sedimentation rate was 41 mm/h and C-reactive protein was 60 mg/L. Biochemical parameters were normal on admission. The total IgA, IgG, IgM and IgE levels, and the IgG subgroups and the complement levels were normal. CEA, AFP, Ca 19-9 and PSA were also

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within normal range. The rheumatoid factor, antinuclear antibodies, anti-ds DNA, ENA, ANCA, HBs Ag, anti-HBc IgM, anti-HCV, anti-HIV were negative. In order to exclude Behcet disease, HLA B51 was performed and it was found to be negative. Other HLA types were not investigated. Cranial magnetic resonance imaging (MRI), transthoracic echocardiography and abdominal computerized tomography (CT) were normal. Tuberculin skin test and pathergy tests were negative. Meningoencephalitis was considered. A combination treatment of ceftriaxon (2 g IV q12h) and acyclovir (10 mg/kg IV q8h) was started until the CSF culture result was available. Furthermore, antibodies against Herpes simplex virus, Borrelia burgdorferi, and Brucella spp. were undetectable in both the blood and CSF. A skin biopsy showed neutrophilic dermatosis characterized by the presence of dense dermal infiltrates of mature neutrophils and leukocytoclastic vasculitis of the erythematous lesions (Figs. 3, 4). A biopsy of the nodular lesions showed septal panniculitis with small foci of inflammatory cells extending into the adjacent lobular fat. After the fourth day of hospitalization the patient became dispneic. On physical examination crackles were detected in both lungs. A chest computed tomography (CT) revealed bilateral reticulonodululay infiltrations and pleural effusion (Fig. 5). Fiber-optic bronchoscopy did not show any endobronchial abnormality. Bronchoalveolar lavage revealed 170/mm³ leukocytes with 60% neutrophils and 40% mononuclear leukocytes. A transbronchial lung biopsy showed interstitial inflammation, edema, minimal fibrosis and alveolar
infiltration by large numbers of neutrophils and occasional lymphocytes. The patient’s antibiotics were replaced with imipenem and teicoplanin in order to include nosocomial pathogens. CSF, urine, blood, skin, sputum, bronchoalveolar lavage, pleural fluid stains and cultures were negative for bacteria, fungi, parasites and *M. tuberculosis*. Polimerase chain reaction results for *M. tuberculosis* in CSF, urine, sputum, bronchoalveolar lavage, pleural fluid were also negative. Peripheric blood flow cytometric analyses were within normal range. Despite empirical broad spectrum antibiotics, his skin lesions and dyspnea worsened. The diagnosis of Sweet’s syndrome was established on the basis of the previously published diagnostic criteria for this disease (3). A high dose of oral steroid treatment (prednisone 60 mg/day for 7 days) was started. The patient’s respiratory symptoms, mental state and lesions improved dramatically within two days of steroid treatment and he became afebrile. The patient’s control chest X-ray returned to normal. Prednisone treatment was tapered with a slow dose. The patient was followed up for six months subsequent to steroid treatment. No more skin lesions appeared during this period of time.

Discussion

Sweet’s syndrome commonly occurs in association with infectious and inflammatory disorders. It is also associated with neoplasms, especially hematological disorders, autoimmune diseases, drugs and trauma (4-6). Through laboratory and radiological test results we excluded these potential underlyng disorders and diseases mimicking Sweet’s syndrome in the patient. Although the patient was a chronic alcoholic, his liver function tests were normal on admission. During his follow up, before steroid treatment, his ALT and AST levels were elevated (ALT was 312 IU/mL, AST was 180 IU/mL) for a short period and returned to normal with steroid treatment. Sweet’s syndrome may be idiopathic (7). We think that chronic alcoholism may also be a trigger for this patient but yet a thorough review of the available literature produced no information regarding an association between Sweet’s syndrome and alcoholism.

Our patient’s symptoms fulfilled the previously published diagnostic criteria for Sweet’s syndrome. He had sudden onset of tender erythematous plaques and dense neutrophilic infiltrates on biopsy as the major diagnostic criteria; and a high fever, elevated erythrocyte sedimentation rate, positive C-reactive protein and an excellent response to treatment with systemic corticosteroids supplemented the diagnosis as minor criteria (3). Even though vasculitis has not been considered a component of Sweet’s syndrome, recently it has been reported that Sweet’s syndrome can demonstrate vasculitis (2, 8).

Although systemic manifestations of Sweet’s syndrome such as fever, arthritis and ocular involvement have been reported frequently in the literature, pulmonary and CNS involvements are very rare (9). In the present patient both pulmonary and CNS involvement occurred. The pulmonary involvement present in our patient was diagnosed based on clinical, radiological, and also histopathological findings. The patient’s chest CT revealed bilateral reticulonodular infiltrations and pleural effusion. Infiltration of a single lobe, extensive infiltrates, nodular infiltrates, and effusions have been reported as radiological findings of Sweet’s syndrome in the literature (10). Our patient’s transbronchial lung biopsy showed interstitial inflammation, edema, minimal fibrosis and alveolar infiltration by large numbers of neutrophils and occasional lymphocytes. Interstitial infiltrates of neutrophils, lymphocytes, macrophages and eosinophils have been demonstrated in the lung biopsies of most patients with pulmonary involvement in the literature. Intraalveolar edema and minimal fibrosis have also been reported as in the present patient (4, 5, 10, 11).

Sweet’s syndrome with central nervous system involvement is also rare in the literature. Encephalitis and meningitis are the most common neurological manifestations. An analysis of 20 previously published case reports showed pleocytosis in 17 of 20 patients with neuro-Sweet disease (12). In CNS involvement, radiological images can be normal in some patients, while in other patients various abnormalities in the basal ganglia, white matter, or brain stem can be seen (13). In our patient the cranial MRI was normal. As many clinical features of Sweet’s syndrome are similar to those of Behcet disease, and as both of them can be seen concurrently in the same patient, the differential diagnosis should be made between these two diseases (12). We investigated and excluded the existence of Behcet disease in our patient.

In conclusion, we emphasize again that although rare, involvement of the pulmonary and central nervous systems can be seen in Sweet’s syndrome. Also vasculitis does not exclude the diagnosis of Sweet’s syndrome. Furthermore, differential diagnosis between Sweet’s syndrome and Behcet disease should be made.

References

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