Pneumonectomy in pulmonary mucormycosis complicating Behçet's disease

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Summary: A 37 year old man with Behçet's disease who was maintained on prolonged corticosteroid therapy, developed diabetic ketoacidosis and pneumonia. Secondary infection with mucor intervened with abscess formation cured by pneumonectomy. The association of Behçet's disease and mucormycosis has not been previously reported, although diabetes mellitus was almost certainly the predisposing cause. Surgical treatment offers the best chance of survival in similar cases.

Introduction

Mucormycosis occurs in patients with several underlying diseases, especially haematological malignancies and diabetes mellitus (Murray, 1977). A patient with Behçet's disease complicated by pulmonary mucormycosis, successfully treated by surgical resection is presently reported.

Case report

A 37 year old man was admitted because of arthritis of the right ankle, aphthous stomatitis, scrotal ulcers and erythema nodosum. The laboratory tests showed a mild leucocytosis and elevated erythrocyte sedimentation rate. HLA B₅ was found. These findings were consistent with the diagnosis of Behçet's disease. His condition improved following treatment with diclofenac sodium 200 mg/day for 30 days. Two years later he was readmitted because of headache, blurring of vision and progressive loss of memory. The physical examination revealed left hemiparesis with left homonymous hemianopsia. The cerebrospinal fluid protein level was 140 mg/dl and the computed tomographic (CT) scan showed a space occupying lesion in the right temporoparietal region. These clinical findings were interpreted as a pseudotumour due to neurological manifestation of Behçet's disease. Treatment with prednisone 80 mg/day was started and a progressive improvement occurred, as demonstrated

by the resolution of the CT scan findings and disappearance of the neurological deficits.

A year later, while still under treatment with dexamethasone 3 mg/day, the patient was readmitted because of fever, chills, polydipsia and polyuria. On physical examination he was obtunded, dyspnoeic with signs of consolidation in the right lung. The white blood cell count was $18.9 \times 10^9/l$ with a shift to the left, the blood glucose level was 1000 mg/dl, arterial blood pH 7.1, urine glucose 4% and ketones + 3. Characterization of the T-cell population demonstrated a diminished number of total T-cells and that of helper cells (T – 56%, B – 19%, helper (OKT₄) – 35%, suppressor (OKT₈) – 21%). The chest X-ray revealed a massive pneumonic infiltration of the right lung. Soon acute respiratory failure developed, requiring mechanical ventilation. Repeated sputum cultures grew *Klebsiella* and *Enterobacter* species. Following a week of intensive care treatment including wide-spectrum antibiotic coverage, he slowly improved, as also seen in his repeated chest X-rays.

The patient was discharged on the 39th day of hospitalization. During a follow-up period of one month a right parahilar infiltrate persisted unchanged (Figure 1). Bronchoscopy revealed a few hard endobronchial plaques, which almost completely obstructed the right main upper lobe bronchus. A biopsy specimen stained with PAS showed numerous hyphae of mucor invading blood vessels (Figure 2).

Treatment with intravenous amphotericin B 5 mg/day was started. However, a severe systemic reaction (chills, high fever, retrosternal tightness) appeared and

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Figure 1 Persistent right parahilar infiltrate.

the drug was discontinued after the administration of a total dose of 140 mg.

A right pneumonectomy was performed. A huge abscess cavity was found near the right main bronchus with invasion of the cavity wall by mucor (Figure 2). Attempts to reinstitute amphotericin B following surgery failed again because of the reappearance of the above mentioned systemic reaction. The patient made an uneventful recovery and now, 2 years after surgery, his condition is satisfactory.

Discussion

Pulmonary mucormycosis usually has a fatal outcome and in the majority of these patients the correct diagnosis is established on autopsy (Meyer *et al.*, 1972; Murray, 1977). We are aware of 70 hitherto reported patients with pulmonary mucormycosis. Four of them were cured by amphotericin B and 9 other patients by surgical resection (including one with isolated tracheal involvement) (Gribetz *et al.*, 1980; Record & Ginder, 1976; Schwartz *et al.*, 1982). Usually, the infection accompanies severe systemic illnesses, characterized by impaired host defence mechanisms. More than 75% of the patients had leukaemia or lymphoma (Meyer *et al.*, 1972). Multiple predisposing factors had

Figure 2 Hyphae of mucor invading blood vessels.

been mentioned: diabetes mellitus, corticosteroid treatment and prolonged antibiotic therapy (Gribetz *et al.*, 1980). In our patient the disease was the endpoint of an unusual chain of events: neuro-Behçet's disease successfully treated with corticosteroids, which in turn initiated diabetic ketoacidosis without previous history of diabetes mellitus, followed by a severe pulmonary bacterial infection necessitating prolonged antibiotic treatment. Several immunological abnormalities are described in patients with Behçet's disease including generation of autoantibodies against oral mucosa, circulating immune complexes in the serum, hyperglobulinaemia and impaired cellular immunity with a decrease in the number of T-cells and diminished response to lectins (O'Duffy, 1981). Immunological abnormalities were also demonstrated in our patient.

Some of the problems posed by amphotericin B therapy are demonstrated in our case. Doubts have been raised concerning the efficacy and the exact role of this form of treatment (Gribetz *et al.*, 1980; Murray, 1977). The present case history supports the previously reported view (Gribetz *et al.*, 1980; Schwartz *et al.*, 1982) that whenever the localized pulmonary mucormycosis is encountered, surgical resection may be lifesaving.

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