MALAKOPLAKIA OF LUNG IN AN IMMUNOCOMPROMISED PATIENT

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Introduction

Malakoplakia of the lung is rare and till 1999 only 14 cases had been described in medical literature, in AIDS patients. It is an unusual inflammatory process of unknown cause which is characterised by yellow brown plaques composed primarily of macrophages with large PAS positive intracytoplasmic inclusions and concentrically laminated calcific spherules known as Michaelis Guttman bodies [4]. It clinically simulates malignancy in a variety of organs and most commonly it affects the lower urinary tract [5]. We describe here a case of pulmonary malakoplakia in an immunocompromised host.

Case Report

A 24 year old asymptomatic male was found to be HIV positive during screening for blood transfusion. Two years later, he presented with five months history of low grade fever, non productive cough, dull aching pain over left anterior chest wall, 5 Kg weight loss and 4 episodes of streaky haemoptysis. Chest x-ray showed a homogenous opacity in left middle zone (Fig-1).

On examination he was averagely built and nourished. General examination was normal. The percussion note was impaired and breath sounds were decreased over left mammary area. Haematological and biochemical parameters were normal. Examination of sputum smears failed to show AFB. Fibreoptic bronchoscopy (FOB) revealed a fleshy growth almost completely obstructing the superior segment bronchus of the lingula. Biopsy from this growth was suggestive of chronic inflammatory granulomatous tissue. Broncho alveolar lavage and post bronchoscopy sputum failed to demonstrate AFB, or malignant cells but grew *Candida*. On clinical suspicion of pulmonary tuberculosis, antitubercular drugs were started along with ketoconazole for candida infection.

The radiological opacity further increased while on therapy (Fig-2). CT scan chest showed a 8x9 cm soft tissue mass attached to the lateral chest wall anteriorly alongwith spread to the superior lingular segment of lung. FNAC of this mass was inconclusive except showing chronic granulation tissue. At this time culture of post FOB sputum grew AFB resistant to Streptomycin and Rifam-

picin. Patient was symptomatic with weight loss. Chest x-ray showed further increase in size of opacity. He was finally submitted to thoracotomy to establish the diagnosis.

On gross examination, a 12x10 cm mass was seen occupying the upper half of the left upper lobe. It was adherent to the anterior chest wall. Necrotic material was seen inside it. Most of it could be excised. Histopathology from this mass showed necrotising inflammatory areas with macrophages containing Michaelis Guttman bodies pathognomonic of malakoplakia (Fig-3). A careful search for mycobacteria or a tuberculous granuloma from the excised specimen did not reveal any nor the specimen culture grew mycobacteria subsequently. He was treated with intravenous ceftazidime and gentamycin to which he responded.

Since post FOB sputum had grown mycobacteria resistant to

Fig. 1 : X-ray chest showing left mid zone lesion

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Fig. 2 : X-ray chest showing increase in lesion

streptomycin and rifampicin, he was treated with, EHRZ, ciprofloxacin, PAS regimen. Patient later developed cryptococcal meningitis proved by CSF examination. CD4- count was 100/cm. He was treated with fluconazole. However patient died later, nearly 5 months after the diagnosis of malakoplakia and S&R resistant tuberculosis.

Discussion

Malakoplakia was first described by Michaelis and Gutmann in 1902 and elaborated by von Hanseman in 1903, who coined the term which is derived from the Greek words 'malakos' (soft) and 'plakos' (plaque) [6]. Stanton and Maxted reviewed 153 of 198 cases reported till 1981 and found genitourinary tract to be involved in 58% of cases, gastrointestinal in 12%, retroperitoneal in 12% and skin in 3%. There were 2 cases involving adrenal gland and cerebrum and 1 case each involving lung, vertebrae, pancreas, pleura, muscle, spleen and hip joint [6]. 40 of these cases had an inter-current systemic disorder, carcinoma, an immune deficiency syndrome or an autoimmune disease.

The pathophysiology of malakoplakia is not well understood. At different times, it was thought to be a tumor, complication of tuberculosis, mast cell disorder or a form of sarcoidosis, but currently it is thought to be arising from acquired defect of macrophage bactericidal digestion [4,7]. All cases of pulmonary malakoplakia reported and reviewed till 1999 had underlying disease states as found in extrapulmonary cases of malakoplakia. They include cardiac and renal trans-

Fig. 3 : Microsection from biopsy specimen showing Michaelis Guttmann bodies in a background of chronic inflammatory cells.

plantation, Hodgkin's disease, AIDS and alcohol abuse. Mortality rate is high in these cases and either one or both lungs may be involved. It does not favour any particular site and is often accompanied by extrapulmonary malakoplakia [2]. The case reported by us had only pulmonary involvement and no extrapulmonary site.

Infection has been accepted as a causative factor for malakoplakia. Three fourths of all cases are related to infection by *E.Coli*. Other bacteria include *Klebsiella*, *Corynebacterium*, *Mycobacteria* and *Staphylococcus aureus*. On rare occasions even fungi have been reported as a putative cause [8]. Of reported cases of pulmonary malakoplakia, 2 were positive for *Rhodococcus equi* cultures [2,3,9]. Shin et al diagnosed a case of pulmonary malakoplakia in an AIDS patient based on histopathological examination of a lung mass by transbronchial biopsy and it was associated with *Rhodococcus equi* infection [2]. Other authors have also found that there appears to be more than coincidental association between pulmonary *R. equi* infection, malakoplakia, and human immunodeficiency virus infection. In the case reported here we could only isolate candida initially, though AFB was grown later, resistant to two antituberculous drugs.

The diagnosis of malakoplakia in patients with HIV infection may represent additional evidence of immunological aberrations that could have important clinical consequence [1,2].

Treatment of malakoplakia has been varied. In lower urinary tract malakoplakia, cures have been reported with long term ampicillin and sulfisoxazole [10]. Other drugs tried include antituberculous drugs (para aminosalicylic acid, INH, rifampicin and streptomycin) and unspecified long term antibiotics, along

with fulguration. Ciprofloxacin has been found to be effective in one case [3].

The present case we managed successfully with resection and a combination of ceftazidime and gentamycin. A combination of intracellularly active antibiotic to which the organism is sensitive, such as one of the sulphonamides, trimethoprim-sulphamethoxazole or rifampicin would thus appear to be the first line regimen in eradicating malakoplakia regardless of the organ system involved. Surgical treatment may be necessary if the disease progresses despite medical regimen [6].

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