Massive pleural effusion due to IgG multiple myeloma

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Abstract

Pleural effusion directly attributable to multiple myeloma is exceedingly uncommon and is said to occur in only 1-2% of cases. Of these around 80% occur in IgA disease. We report a case of myelomatous pleural effusion (proven on cytological and immunohistochemical analysis) in a patient with the IgG-κ subtype. We describe the diagnosis, pathogenesis and management of this condition and show the radiological and cytological evidence of the case.

Introduction

Multiple myeloma is a clonal B-cell malignancy characterized by proliferation of plasma cells accumulating mainly in the bone marrow and secreting paraprotein.1 Myeloma accounts for around 1% of all malignancies and UK national statistics for 2006 report an incidence of 6.4 cases per 100,000 annually.2 Mean age at diagnosis is 65 years.

Pleural effusion directly attributable to myeloma is exceedingly rare (1-2% of cases).3 We present a case of pleural effusion arising rapidly in a patient with long-standing multiple myeloma and found to be a myelomatous effusion.

Case Report

A 74-year old man with longstanding IgG kappa myeloma presented with a two week history of increasing dyspnoea on exertion. Blood tests showed a normocytic anaemia (Hb: 7.4 g/dL, MCV: 90.1 fl), and renal impairment (urea: 9.2 mmol/L, creatinine: 248 µmol/L). Serum electrophoresis suggested rapid disease progression with an increase in paraprotein from 14 g/L to 50 g/L over a six week period. Chest radiographs showed a large left sided pleural effusion (Figure 1). Diagnostic tap of the effusion produced heavily blood stained fluid with features suggestive of an exudate (pH: 7.62, total protein: 93 g/L, LDH: 506 U/L).

The effusion persisted, necessitating therapeutic drainage which produced four liters of bloodstained fluid. Computed tomography (CT) of the chest was radiologically suggestive of mesothelioma (Figure 2) but cytological analysis of the pleural fluid showed plasma cells consistent with myeloma (Figure 3).

Subsequent immunohistochemistry revealed κ light chain restriction and VS38 positivity. A diagnosis of pleural effusion secondary to multiple myeloma was made and the patient was commenced on bortezomib and dexamethasone therapy seven days after first presenting. No further pleural aspiration was necessary from this point and repeat chest radiography (Figure 4) taken seven days after starting treatment showed resolving pleural disease and the patient reported improved symptomatology.

Discussion

Pleural effusion is uncommon in multiple myeloma; it has been suggested that it occurs in approximately 6% of cases. Most of these are not directly due to myelomatous infiltra-
natural history of the disease or a feature of
the aggressive behavior of myeloma. Diagnosis is based on cytological analysis and
immunohistochemical staining, pleural biopsy
may also yield a diagnosis of myeloma.
Malignant plasma cells within the cytological
preparation show a typical basophilic cyto-
plasm with large, eccentric nuclei and promi-
nent nucleoli.4

Several possible mechanisms are postulat-
ed for myelomatous pleural effusion: invasion
from adjacent skeletal lesions; extension from
chest wall plasmacytomas; tumor infiltration
of the pleura; and mediastinal lymph node
involvement causing lymphatic obstruction.4,5
Currently, the exact mechanism of pathogene-
sis remains unknown.

The management is two-stage. First, the
effects of the effusion must be alleviated with
therapeutic and diagnostic thoracocentesis.
Once myelomatous pleural effusion is con-
formed on cytology, and/or immunohistochem-
istry, systemic chemotherapy is required to
slow the advance of the disease. There is as
yet no consensus as to how best to manage
this rapid and aggressive phase of disease but
we anecdotally show evidence of a good
response to bortezomib and dexamethasone
therapy.

We suggest that this case highlights the
vital importance of diagnostic clarity, where
possible, in finding the underlying etiology of
a pleural effusion. Myelomatous pleural effu-
sion carries a poorer prognosis and more work
is needed to understand the exact pathogene-
sis of this condition.

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