with aggressive treatment of any infection in an attempt to prevent progression to bronchiectasis.

In summary, this child had bronchomalacia of the pattern seen in the Williams Campbell syndrome and other congenital abnormalities, previously not encountered together, which supports a congenital cause for this syndrome. The case also emphasises the need critically to assess all wheezy children. For many years this boy was thought to have asthma, and was even treated with systemic corticosteroids. Flow-volume loops can show large airway disease and, as in this child, may alert the clinician to the possibility of another diagnosis.

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Malignant pleural effusion in chronic myelomonocytic leukaemia

A Manoharan

Abstract

A case of malignant pleural effusion due to chronic myelomonocytic leukaemia is reported.

Malignant pleural effusions are extremely rare in patients with chronic myelomonocytic leukaemia, a haemopoietic stem cell disorder with myeloproliferative and myelodysplastic features.[1 2]

Case report

A 52 year old woman presented with a three week history of lethargy, weakness, and progressive dyspnoea. Examination disclosed bilateral pleural effusions, greater on the left than the right side, minimal generalised lymphadenopathy, and splenomegaly of 7 cm. Chest radiography and computed tomography showed mediastinal lymphadenopathy and bilateral pleural effusions. Abdominal computed tomography showed hepatosplenomegaly and para-aortic lymphadenopathy. A full blood count showed: haemoglobin 12·8 g/dl, white blood cells 87 × 10^9/l (neutrophils 52%, monocytes 27%, blasts 4%, promyelocytes 1%, myelocytes 4%, metamyelocytes 4%, and band forms 8%), and platelets 205 × 10^9/l; some neutrophils were hypogranular and some platelets were noted to be large on the blood film. Bone marrow examination showed hypercellularity, active erythropoiesis, active myelopoiesis with 28% monocytes or monocytoid cells and an increased number of megakaryocytes with abnormal forms, including micronuclear and mononuclear forms. Cytogenetic studies showed an abnormal chromosome constitution: 46,XX,del(7)(q22), del(20)(q11). Clotting studies gave normal results, arterial blood gas analysis showed hypoxaemia (oxygen tension 5·7 kPa), and results of liver function tests were abnormal. Pleural fluid biochemical investigations showed: protein 38 g/l, glucose 3·7 mmol/l, and lactic dehydrogenase 482 (normal 55–120) U/30 l. Cytological examination of pleural fluid (figure) showed features of chronic myelomonocytic leukaemia. A pleural biopsy was not done. Excision biopsy of an axillary lymph node showed diffuse monocytoid leukaemic infiltration, with frequent mitotic figures.

During the four days when investigations were in progress, repeated left sided thoracocenteses were required for symptomatic relief, and her white blood cell count increased from 87 to 130 × 10^9/l. She was started on treatment with low doses of cytarabine (10 mg/m^2 every 12 hours subcutaneously), 6-thioguanine (40 mg every 12 hours orally), and etoposide (100 mg a day orally) for 10 days, with good results. Six weeks later she was clinically well with reduced splenomegaly (4 cm); her chest radiograph showed a small opacity at the left base posteriorly. Her haemoglobin (11·5 g/dl), white blood count (32 × 10^9/l), and platelets (60 × 10^9/l) had also improved. Further treatment, at the patient's request, consisted only of intermittent low dose single agent therapy with 6-thioguanine or hydroxyurea.

In March 1989 four months after her initial

Department of Clinical Haematology, St George Hospital, Kogarah, Sydney, New South Wales 2217, Australia

A Manoharan

Reprint requests to: Dr Manoharan

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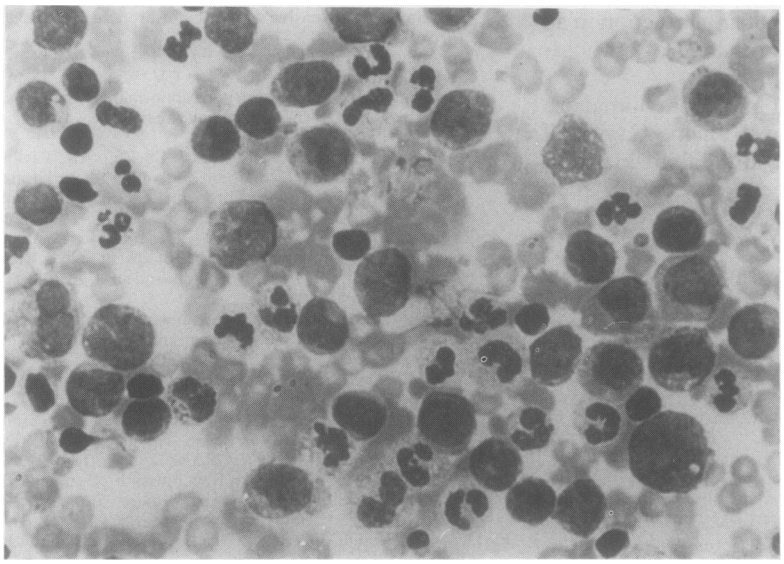

Pleural fluid cytospin preparation showing neutrophils and monocytes. (May-Grünwald-Giemsa stain.)

presentation, she had a clinical relapse and her previous drug regimen was restarted. During the ensuing cytopenic phase, she developed septicaemia but declined active treatment and died at home several days later.

Discussion

Although the peripheral blood film and bone marrow examinations clearly established a diagnosis of chronic myelomonocytic leukaemia, the nature of the coexisting lymphadenopathy and pleural effusion was not immediately clear. These are rare findings in such patients,[3] although a simultaneous lymphoproliferative disorder occurs in some patients with myeloproliferative disorder.[4] Cytological examination of the pleural fluid and lymph node biopsy confirmed leukaemic infiltration and excluded a coexistent lymphoma.

Four of the five reports of chronic myelomonocytic leukaemia with malignant pleural effusion have been in women; the age of the patients has ranged from 52 to 82 years.[1 2] Two of the patients also had concurrent pericardial effusions; other associated clinical features have included splenomegaly (6–16 cm) in three patients, hepatomegaly in two, gum hypertrophy in one, and generalised lymphadenopathy in one. The white blood cell count at presentation has ranged from 33 to 87 $\times 10^9$/l, the absolute monocyte count being 20 $\times 10^9$/l or more in four patients (range 4·3–38·4 $\times 10^9$/l; normal 0·8 $\times 10^9$/l).

Many patients with chronic myelomonocytic leukaemia are symptomless with only a slight monocytosis and it is generally agreed that these patients should be kept under observation only.[2] Our patient had several features associated with an aggressive clinical course, including hepatosplenomegaly, a leukaemic pleural effusion, and rapidly increasing leucocytosis. The clinical response achieved with the low dose combination chemotherapy was similar to that documented in four patients with chronic myelomonocytic leukaemia and pleural effusions treated with razoxane or etoposide, given as a single agent.[1 2] The patient declined further courses of such treatment, although it was well tolerated, because of the need for frequent follow up visits to the hospital. Single agent treatment with hydroxyurea and thioguanine did not give sustained benefit. A recent report has documented an excellent response to low dose etoposide,[2] and this agent deserves further evaluation in patients with chronic myelomonocytic leukaemia and malignant pleural and other serous effusions.

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