CASE REPORT

Cytophagic histiocytic panniculitis in a 74-year-old man

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Abstract

Cytophagic histiocytic panniculitis is a chronic histiocytic disease of the subcutaneous adipose tissue characterised by lobular panniculitis with histiocytes containing blood cell fragments. It is also associated with marked systemic features such as fever, pancytopenia, hepatosplenomegaly, liver abnormalities and coagulopathy. We report a case of cytophagic histiocytic panniculitis in a 74-year-old man successfully treated using combination therapy with prednisolone and cyclosporine A.

Keywords: cytophagic histiocytic panniculitis, cyclosporine A, haemophagocytosis, older people

Case report

A 74-year-old man was admitted to our hospital with a fever, severe malaise and left chest wall and axillary swelling and pain. His past history included chronic bronchitis and hyperlipidaemia. A painful red lump was observed in his left anterior chest wall and axilla. Laboratory test results were as follows: C-reactive protein, 14.94 mg/dl; white blood cell count, 12,200/mm3 with no leukaemic cells; aspartate aminotransferase, 145 IU/l; alanine aminotransferase, 116 IU/l; lactate dehydrogenase, 788 IU/l; total bilirubin, 0.3 mg/dl; fibrin/fibrinogen degradation products, 56.9 μg/ml; D-dimer, 42.6 μg/ml and ferritin, 31 195 ng/ml. Computed tomography scan revealed hepatosplenomegaly and a high-density area in the soft tissue of the left chest wall and axilla. Gallium-67 scintigraphy showed intense uptake in the same region (Figure 1).

The original diagnosis was infective panniculitis, but empirical antibiotic therapy was ineffective. A left axillary skin biopsy showed aseptic lobular panniculitis with predominant CD68+ histiocytes and ‘bean-bag’ cells with no evidence of infection or lymphoma. A diagnosis of cytophagic histiocytic panniculitis (CHP) was made. We initiated methylprednisolone pulse therapy followed by 50 mg/day of prednisolone (PSL) combined with 150 mg/day of cyclosporine A (CyA). Fever and other systemic symptoms, including left chest wall and axillary swelling and pain, resolved within 10 days after treatment.

Discussion

CHP is a histiocytic disease of the subcutaneous adipose tissue characterised by aseptic lobular panniculitis [1–3]. Fatal cases of CHP have been reported as a result of massive visceral haemorrhage or organ system failure [1]. CHP has been treated with glucocorticoids, including glucocorticoid pulse therapy; however, it was often resistant to glucocorticoid therapy and therefore had a poor prognosis [2, 3]. In the late 1980s, treatment with CyA was found to be effective and Ostov et al. reported that CyA significantly reduced the mortality [4]. In refractory cases, the efficacy of chemotherapy [5], autologous peripheral blood stem cell transplantation [6], plasmapheresis [7] and interleukin-1 (IL-1) receptor antagonists [8] have been reported.

The pathogenesis of CHP is still unclear. However, there is a report that a large number of T cells infiltrated a subcutaneous inflammatory lesion in a patient with CHP [3]. This case was the benign type of CHP; successfully treated with CyA. CHP is considered associated with both benign and malignant types of panniculitis. The benign type is authentic CHP, whereas the malignant type is associated...
with subcutaneous panniculitis-like T-cell lymphoma [9]. These two processes differ in aetiological, clinical, histopathological and biological behavioural aspects [10]. CyA is a lipophilic peptide that targets T cells in infiltrating subcutaneous adipocytes, and is thus effective for the treatment of CHP [4].

There are <100 reported cases of CHP [3], and most cases occurred in patients under 50 years old, including children. CHP in patients over 65 years of age has been rarely reported [2–4], although Winkelmann and Bowie reported an 80-year-old patient [1].

Conclusion

CHP is a rare cause of panniculitis that can be treated with PSL and CyA. Although many cases were observed in children, adolescents and adults younger than 50 years of age, CHP should be considered in cases of panniculitis with systemic symptoms even in an aged population.

Key points

• CHP is a very rare cause of aseptic panniculitis.
• Characteristic histological features are aseptic panniculitis with infiltration of so-called ‘bean-bag’ cells.
• Although sometimes fatal, CHP can be successfully treated with high doses of PSL combined with CyA.
• CHP should be considered in cases of panniculitis with systemic symptoms even in an aged population.

Conflicts of interest

None declared.

References


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Figure 1. Gallium-67 scintigraphy showed intense uptake in the left chest wall and axilla.