

CASE STUDY

Idiopathic lobular panniculitis with specific pleural involvement

N. Kumagai-Kurata*, H. Kunitoh*, M. Nagamine-Nishizawa*,
K. Watanbe*, N. Nakamura**

Idiopathic lobular panniculitis with specific pleural involvement. N. Kumagai-Kurata, H. Kunitoh, M. Nagamine-Nishizawa, K. Watanbe, N. Nakamura. ©ERS Journals Ltd 1995.

ABSTRACT: A 19 year old man presented with fever and left pleural effusion. Biopsy of a subcutaneous nodule and bone marrow led to the diagnosis of idiopathic lobular panniculitis (Weber-Christian disease).

Thoracoscopically guided biopsy of the visceral pleura also revealed panniculitis change in the pleural adipose tissue.

Eur Respir J., 1995, 8, 1613–1614.

*Dept of Respiratory Medicine, and **Dept of Pathology, Yokohama Munciple Citizen's Hospital, Yokohama City, Japan.

Correspondence: H. Kunitoh, Dept of Respiratory Medicine, Yokohama Munciple Citizen's Hospital, 56 Okazawa-cho, Hodogaya-ku, Yokohama City, Kanagawa 240, Japan.

Keywords: Adenosine deaminase, panniculitis, pleural effusion, thoracoscopy, Weber-Christian disease

Received: November 16 1994

Accepted after revision March 27 1995

Idiopathic lobular panniculitis is characterized by a systemic inflammatory process of adipose tissue [1, 2] of unknown aetiology. It was originally described as a disease entity by WEBER [3] and CHRISTIAN [4]. The term Weber-Christian disease is applied when cutaneous lesions are associated with systemic manifestations [1]. The diagnosis is based on the characteristic lobular panniculitis seen on biopsy of skin inflammatory nodules [2], after other types of panniculitis, such as those associated with pancreatitis or alpha₁-antitrypsin deficiency [5], have been excluded [1].

There have been few reports of pleural involvement in Weber-Christian disease [6] and other types of panniculitis [5], although the presence of fat tissue in the pleura could lead to pleural panniculitis.

Case report

A 19 year old man was admitted to the hospital in February 1991 because of fever and left pleural effusion. He had been in good health until 1 month prior to admission, when he first noticed subcutaneous nodules in the lower abdomen. One week before admission, he developed fever that did not respond to antibiotics. When he was referred to our hospital, left-sided pleural effusion was documented on the chest roentgenogram. On physical examination, he did not appear gravely ill, although his body temperature was 39.1°C, pulse rate 108 and respiratory rate 36 breaths·min⁻¹. No respiratory crackles or heart murmurs were audible. No hepatosplenomegaly was noted. There were painful, erythematous subcutaneous nodules in the lower abdomen.

Laboratory data revealed an erythrocyte sedimentation

rate of 15 mm·h⁻¹ and C-reactive protein (CRP) of 2.7 mg·dL⁻¹. White blood cell count was 2,200 cells·mm⁻³ (2.2×10⁹·L⁻¹), with 16% segmented neutrophils, 36% band forms, 3% metamyelocytes, 1% myelocyte, 4% monocytes, 39% lymphocytes and 1% atypical lymphocytes. Haemoglobin concentration was 131 g·L⁻¹, and platelet count 210,000 platelets·mm⁻³ (210×10⁹·L⁻¹). Serum glutamic oxaloacetic transaminase (GOT) was 85 IU·L⁻¹ (normal 5–27 IU·L⁻¹), and glutamic pyruvic transaminase (GPT) 59 IU·dL⁻¹ (normal 5–24 IU·L⁻¹). Lactate dehydrogenase (LDH) was elevated to 1,650 IU·L⁻¹, (normal 200–450 IU·L⁻¹) and creatine phosphokinase (CPK) was 568 IU·L⁻¹ (normal 40–220 IU·L⁻¹). No particular isozyme pattern of LDH or CPK was noted. Serum amylase was 131 IU·L⁻¹ (normal 60–250 IU·L⁻¹), and lipase was 39 IU·L⁻¹ (normal ≤65 IU·L⁻¹). Serum protein was 6.0 g·dL⁻¹, and albumin 3.1 g·dL⁻¹. Renal function was normal, with normal electrolytes. Tests for antinuclear antibody, rheumatoid factor and immunocomplex were all negative. Serum alpha₁-antitrypsin was 204 mg·dL⁻¹ (normal 170–274 mg·dL⁻¹). The skin test for purified protein derivation (PPD) of tuberculin was negative. Chest roentgenogram and computed tomography (CT) scan revealed moderate pleural effusion, without pulmonary lesions. Abdominal ultra-sound and CT scan showed no pancreatic abnormality.

Biopsy of a subcutaneous nodule (fig. 1a) revealed subacute panniculitis with necrosis of adipose tissue, infiltration of lymphocytes, neutrophils and histiocytes, and formation of micro-oil cysts. Vasculitis was not observed. Biopsy of bone marrow from the iliac crest (fig. 1b) revealed a similar inflammatory process in adipose tissue in the bone marrow.

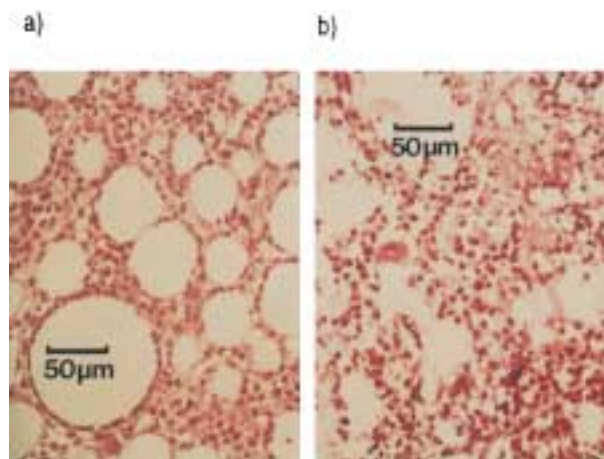


Fig. 1. – Histological sections of: a) the subcutaneous nodule; and b) bone marrow; both of which show inflammatory process of adipose tissue (haematoxylin and eosin stain; internal scale bar=50 µm).

Left-sided thoracentesis recovered a sterile pleural exudate. The pleural glucose was 110 mg·dL⁻¹, protein 3.9 g·dL⁻¹, LDH 3,140 IU·L⁻¹, amylase 108 IU·L⁻¹ and adenosine deaminase (ADA) 153.3 IU·L⁻¹. Thoracoscopic examination, performed on the tenth hospital day, revealed white nodules on the visceral pleura (fig. 2a). Biopsy of the nodules showed scant pleural tissue, and panniculitis change was observed in the adipose tissue (fig. 2b).

The patient was diagnosed as having Weber-Christian disease with involvement of the bone marrow and pleura. Treatment with prednisolone (50 mg·day⁻¹) was initiated, and his symptoms ameliorated. The dose of prednisolone was tapered to 30 mg·day⁻¹ over three weeks, and clinical relapse, characterized by fever and elevated LDH level, occurred. Retreatment with prednisolone at 50 mg·day⁻¹ for two weeks resulted in incomplete clinical response. Oral cyclosporin at 600 mg·day⁻¹ was then

added to the corticosteroid therapy, and total resolution of the disease activity was obtained. The doses of corticosteroid and cyclosporin were then tapered to discontinuation in November 1992. After the disease relapsed in September 1993, the patient was successfully retreated with cyclosporin. He is now disease-free under a maintenance dose of cyclosporin of 100 mg·day⁻¹.

Discussion

Although Weber-Christian disease can involve any fat tissue of the body, including the pleura, pleuritis is a rare complication of this systemic panniculitis. Pleuropulmonary involvement was not mentioned in a large review of 253 cases [2], and in a Japanese series of 70 patients, only four had pleural effusion. Although the pleural involvement was considered to result from panniculitis of the pleural adipose tissue, this is the first report to our knowledge of thoracoscopically examined and histologically documented pleural involvement of the disease.

In our case, the pleural ADA level was elevated to that seen in active tuberculous pleuritis [7]. ADA is reported to be associated with the proliferation and differentiation of lymphocytes (mainly T-lymphocytes). It has been reported that the ADA level and percentage of CD4+ T-cells in pleural exudates are significantly correlated, and the ADA level is presumed to be a marker of cell-mediated immunity [8].

There are several reports of successful treatment of corticosteroid-resistant Weber-Christian disease with cyclosporin [9–11]. Although this drug appears to be highly effective in those cases, its exact role in the treatment of Weber-Christian disease remains to be established.

Elevated pleural ADA and clinical response to cyclosporin treatment in our patient suggest that T-cell-mediated dysfunction may play a role in the pathogenesis of panniculitis-associated pleuritis, or that of Weber-Christian disease in general.

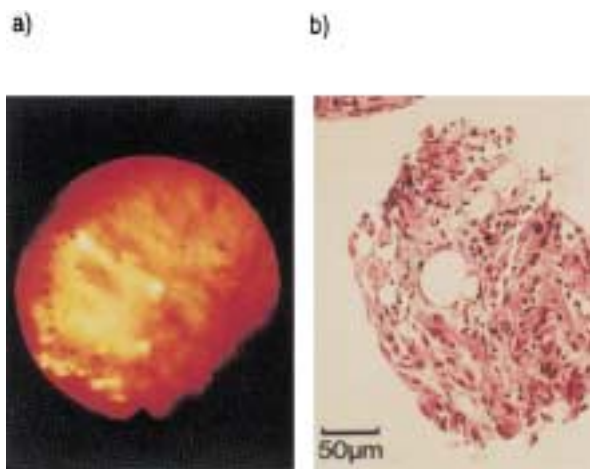


Fig. 2. – a) White nodules on the visceral pleura visualized by thoracoscopy, b) histological section of the nodule revealing panniculitis change of pleural adipose tissue (haematoxylin and eosin stain; internal scale bar=50 µm).

References

1. Lazarus GS. Panniculitis and disorders of the subcutaneous fat. *In*: Wyngaarden JB, Smith LH, eds. *Cecil Textbook of Medicine*, 18th edn. Philadelphia, W.B. Saunders Co., 1988; pp. 2050–2051.
2. Panush RS, Yonker RA, Dlesk AD, Longley S, Caldwell JR. Weber-Christian disease: analysis of 15 cases and review of the literature. *Medicine* 1985; 64: 181–191.
3. Weber FP. A case of relapsing nonsuppurative panniculitis, showing phagocytosis of fat cells by macrophages. *Br J Dermatol* 1925; 37: 301–311.
4. Christian HA. Relapsing febrile nodule nonsuppurative panniculitis. *Arch Intern Med* 1928; 42: 338–351.
5. Smith KC, Pittelkow MR, Su WP. Panniculitis associated with severe alpha₁-antitrypsin deficiency: treatment and review of the literature. *Arch Dermatol* 1987; 123: 1655–1661.

6. Achten G, Moriame-Rousse N, Wanet J, De Coninck J, Vertommen J. Idiopathic liquefying panniculitis with fatal outcome. *Ann Dermatol Venereol* 1977; 104: 693–696.
7. Banales JL, Pineda PR, Fitzgerald JM, Rubio H, Selman M, Salazar-Lezama M. Adenosine deaminase in the diagnosis of tuberculous pleural effusions: a report of 218 patients and review of the literature. *Chest* 1991; 99: 355–357.
8. Baganha MF, Pego A, Lima MA, Gaspar EV, Pharm B, Cordeiro AR. Serum and pleural adenosine deaminase: correlation with lymphocytic populations. *Chest* 1990; 97: 605–610.
9. Entzian P, Barth J, Mong H, Ohnhaus EE, Kirch W. Treatment of Weber-Christian panniculitis with cyclosporin A. *Rheumatol Int* 1987; 7: 131.
10. Usuki K, Kitamura K, Urabe A, Takaku F. Successful treatment of Weber-Christian disease by cyclosporin A. *Am J Med* 1988; 85: 276–278.
11. Royle G, Blacklock H, Miller M. Treatment of cytophagic panniculitis with cyclosporin A. *Am J Med* 1992; 92: 704–705.