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Multicentric Reticulohistiocytosis: A Case Report

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Received July 1, 1982

Abstract. With the present case report we call attention to the clinical entity of multicentric reticulohistiocytosis (MR). Referring to earlier case reports on this rare condition we present the history, clinical and histopathological findings. We stress the importance of revealing possible underlying internal diseases, including malignancies, and also the importance of histological examination of synovium when synovectomies are carried out in patients with arthritis of unknown cause, as arthritis commonly precedes the mucocutaneous lesions by years.

Key words: Polyarthritis; Papulo-nodular dermatosis; Histiocytic multinucleated giant cells of foreign body type

Multicentric reticulohistiocytosis (MR), first described by Weber and Freudenthal in 1937 (see 5), is a rare, generalized disorder of unknown etiology. It has been associated with tuberculosis and thyroid diseases (9), lipid storage diseases (7), and in 24% with malignancies (5). In 1980 Belaich (2) reviewed 70 cases from the literature. Since then only 7 cases have been reported (3, 4, 8, 10).

The condition is characterized by xanthomatous giant cell granulomata involving the skin, the mucous and synovial membranes, and occasionally other organs (1).

The papulo-nodular lesions of the mucocutaneous membranes are preceded in two-thirds of the cases by arthritis, by months or even years. Pruritus is common. Associated symptoms are weakness, weight loss and febrile periods. Females are afflicted more often than males; the mean age of onset is the fifth decade.

The patient to be described is to the best of our knowledge the first case of MR reported from Scandinavia.

CASE REPORT

The patient is a 58-year-old housewife. Rheumatoid arthritis was present among several family members. At the age of 45 she developed a mutilating arthritis, gradually involving almost all joints, necessitating hospitalization in the Department of Rheumatology six times during the last 3 years.

Clinical examinations have shown symmetrical swelling and deformity of the MCP and PIP joints and signs of arthritis in most other joints. Radiograms showed destructive changes.

Laboratory examinations showed ESR elevation, slight anemia, a positive latex RF test, a negative Waaler-Rose test, and negative ANA. Other examinations monitoring hematologic and serologic parameters were within normal limits.

In addition to conventional physical and systemic anti-rheumatic treatment the patient underwent several synovectomies. Unfortunately the synovium was not examined histologically.

In 1981 the patient developed a pruritic papulo-nodular eruption on the back of her hands and fingers. On admission to the Department of Dermatology a few months later she presented myriads of reddish-brown papules and nodules, 2–20 mm in diameter, on the hands and fingers, on the upper chest, upper arms, face, lips and tongue. She had lost weight and suffered from weakness, and presented a crippling arthritis.

During the following weeks some of the lesions involuted spontaneously. A later exacerbation located to the forearms faded after about 4 weeks.

Clinical and laboratory examination did not disclose internal disease or signs of malignancies.

Histopathological findings

Biopsies were taken from lesions on the skin, lower lip and oral mucosa. They were fixed in 4% buffered formaldehyde and embedded in paraffin wax. The sections were stained with hematoxylin and eosin (HE) and periodic acid-Schiff (PAS).

All specimens showed accumulations of large cells with a faint eosinophilic cytoplasm. Many of the larger cells contained multiple nuclei. Most cells had PAS-positive material. In two of the biopsy specimens the histiocytes were found growing in between the collagen bundles of the reticular dermis. In other specimens nodular accumulations of cells were found in the papillary dermis and also down to the subcutaneous tissue.

Other cell types such as lymphocytes and granulocytes were only sparsely present.

DISCUSSION

Reticulohistiocytoma occurs in two clinical distinctive, but histologically indistinguishable forms: a local variety, reticulohistiocytoma, and MR.

The history as well as the clinical and histological findings in our patient were fully consistent with the diagnosis MR.

In clinically inconclusive cases, such as ours, histological examination may give the diagnosis as the histopathological findings are distinctive. Therefore histological examination of synovium after synovectomy should be carried out in patients with arthritis of unknown cause.

The differential diagnoses regarding the arthritis are sero-positive rheumatoid arthritis, sero-negative polyarthritis, paraneoplastic arthritis and crystal arthritis, which may all be accompanied by skin nodules, whereas the described skin lesions may resemble granuloma annulare, histiocytoma and sarcoidosis.

The prognosis is clearly influenced by a possible underlying malignancy. In our case, malignancy has so far been excluded.

As regards the skin lesions, about one-half will clear completely or improve after a few years; the other half will remain stationary or progress. The arthritis remains stationary in 50%; in the other half it progresses and may result in crippling arthritis of the hands.

There is no adequate treatment for the disease.

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Atopic Dermatitis and Hodgkin's Disease

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Received June 21, 1982

Abstract. Five patients with atopic dermatitis developed Hodgkin's disease. This rare clinical occurrence was discovered when adenopathy revealed Hodgkin's granuloma in a biopsy specimen, thus adenopathy in chronic atopic dermatitis should not be dismissed casually.

Because atopy occurs so frequently in the general population, its concurrence with other abnormalities may be expected. However, two coexisting phenomena can influence one another. This appeared to be so in the study of atopy and Hodgkin's disease by Amlot & Green (1). These authors studied 115 patients who had active Hodgkin's disease. The patients with Hodgkin's disease and a high IgE serum concentration were separated into atopic and non-atopic groups. In the atopic patients, the IgE levels were not influenced by treatment, whereas in the non-atopic patients, the IgE levels decreased with successful therapy of the Hodgkin's disease.

No relationship of elevated IgE level to duration of remission or life expectancy in Hodgkin's disease has been observed. Atopic patients were different clinically, having less fever and feverish night sweatings and less loss of weight.

CASE REPORTS

We have analysed the patient material of the Mayo Clinic between 1954 and 1979 and discovered 5 cases of atopic dermatitis and Hodgkin's disease. The essential data of these cases are summarized in Table I.