

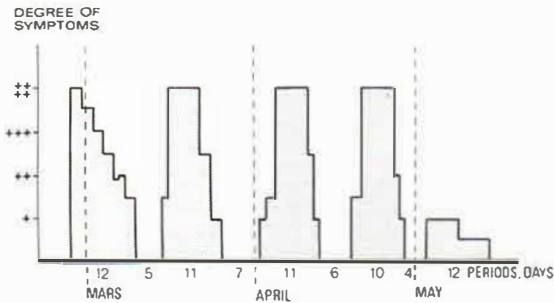


Fig. 2. The patient's arbitrary scores of complaints caused by periods of erythralgia.

Laboratory examinations revealed no abnormalities in routine tests on the blood and urine. Serum triglycerides and cholesterol levels were normal, but had at the most reached 5 and 12 mmol/l, respectively. Autoantibodies were not found. A standard patch test was negative.

Histological examination of the lesion on the left foot revealed a thin epidermis with hydropic degeneration of the basal layer, hyperkeratosis and a sub-epidermal edema below which an infiltrate of lymphocytes was present.

X-ray examination of the feet proved normal. Oscillometry and plethysmography showed normal arterial circulation in both legs. Capillar microscopy of the toes gave normal results.

Subsequent course. In January 1977, 3 g aspirin per day was prescribed. The painful erythema disappeared and the dosis could be reduced without any recurrence during the next 6 months. A leg ulcer appeared in the vicinity of the medial ankle. Hyperkeratotic plugs appeared within the keratotic lesions which gave discomfort on walking. Topical keratolytic and steroid treatment was sufficient to alleviate these complaints.

Comments. The clinical and histological diagnoses were lichen sclerosus et atrophicus (LSA), erythralgia (E) and a post-thrombotic syndrome of the right leg. LSA has been reported once on the feet, in twin sisters (1) and seems to affect the palms and soles only very rarely (2). Boneu et al. (3) and Redding (4) have associated the effect of aspirin with its depression of the aggregation of platelets in thrombocythemia. This mechanism would seem to be a possible cause of the beneficial effect obtained in this case with normal thrombocyte levels. On the other hand no effect of aspirin on LSA could be ascertained.

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Chronic Hypertrophic Vulvitis— A Condition with Similarities to Cheilitis Granulomatosa (Melkersson-Rosenthal Syndrome)

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Abstract. Two cases of chronic hypertrophic vulvitis are described, showing histological changes identical with cheilitis granulomatosa.

Key words: Vulvitis; Cheilitis granulomatosa; Melkersson-Rosenthal syndrome

The syndrome consisting of relapsing swelling of the lips, paresis of the facial nerve and sometimes fissured tongue was first described by Melkersson (2) and Rosenthal (5). The disease is sometimes complicated by paresis of other cranial nerves, mostly the trigeminal nerve, and disturbance of the salivary gland function leading to hypo- or asialia in more than 50% of cases (4). The cheilitis granulomatosa, which is the cutaneous component of the syndrome, usually has a typical histological appearance.

Besides involvement of the lips, alterations indistinguishable from cheilitis granulomatosa have been found in the eye lids (1). In the present paper we describe two patients with vulvar lesions, which were histologically identical with cheilitis granulomatosa.

Case 1. A 35-year-old woman, who had had recurring furuncles in her labia majus since 30 years of age. The last year she had no infection but had noticed a swelling of her labia majus. Because of a lack of success with local treatment, the major part of her vulva was surgically removed.

Case 2. A 22-year-old woman with chronic vulvitis the last 5 years. There was no history of previous disease. She was treated locally and signs of infection disappeared, but a severe swelling of both labia majus and minus remained. A biopsy showed a microscopic picture resembling that of cheilitis granulomatosa. Subsequently both labia majus and minus were removed surgically.

MICROSCOPICAL FINDINGS

Both patients showed the same types of histological alteration. The epidermis was irregularly hyperplastic and showed areas of spongiosis. The dermis revealed a highly remarkable thickening with a pronounced edema and many dilated lymphatic vessels. Especially around vessels, infiltrates consisting of lymphocytes and plasma cells were found.

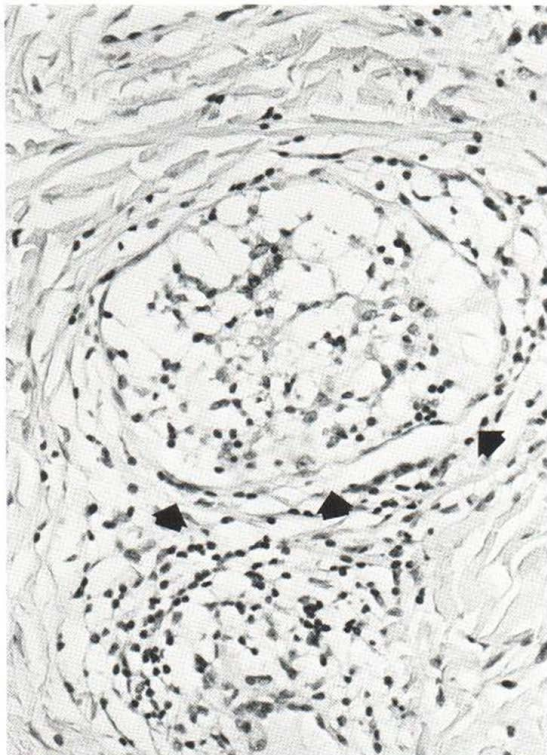


Fig. 1. Dermal granuloma containing epithelioid cells and lymphocytes. The granuloma is at least partially bulging into a lymph capillary (arrows). Outside the granuloma, the collagen bundles are widely separated by a pronounced edema. H. & E. $\times 200$.

Furthermore, scattered small granulomas occurred, containing epithelioid cells, multinuclear cells, lymphocytes and plasma cells (Fig. 1). The giant cells were most commonly of foreign body type.

Most granulomas lay close to or surrounded dilated lymph vessels. Some lymph vessels were obviously compressed by the granulomas (Fig. 1) and small invaginations with epithelioid cells into the lumen of the vessels was sometimes seen. There were no necroses.

DISCUSSION

The histological findings in the vulvar lesions of our two patients are identical with those in cheilitis granulomatosa. Clinically, the swelling of the labia was also very prominent, but not intermittent as is seen in typical cases of cheilitis granulomatosa. The similarity between the latter and cases of hypertrophic vulvitis does not seem to have been noted previously.

The vulvar lesions appeared after recurrent infections. The etiology of cheilitis granulomatosa is unknown but high incidences of chronic or recurrent infections in the oral and sinus regions have been reported (3).

Obstruction of lymphatic vessels by granulomas has been proposed as a factor in the pathogenesis of the edema in cheilitis granulomatosa (3). It is possible that even if there are differences in the etiology of cheilitis granulomatosa and chronic hypertrophic vulvitis, similar pathogenetic mechanisms may be active, giving corresponding histological lesions.

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