

Dilated Lymphatics in Gottron's Papules

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SUMMARY The histopathology of Gottron's papules has not been often reported. Morphological descriptions of these lesions are mainly focused on changes of the epidermis and interface, and on the nature of the inflammatory infiltrate. Dilated vascular vessels of Gottron's papules are rarely mentioned in the literature. We studied these vessels in a biopsy specimen obtained from Gottron's papule in a 65-year-old woman with D2-40 antibody. Although most of the vessels were identified as blood vessels (venules and capillaries), some were demonstrated to be dilated lymphatic vessels.

KEY WORDS: dermatomyositis, Gottron's papule, D2-40, telangiectasia

INTRODUCTION

Gottron's papules are a well known feature of cutaneous involvement by dermatomyositis. Microscopy studies of these lesions have not been frequently reported, whereas their morphological descriptions are mainly focused on epidermal and interface changes, and on the nature of the dermal inflammatory infiltrate. The dilated dermal vessels of Gottron's papules are rarely described, although illustrations showing these dilated vessels can usually be seen in such reports as well as in book chapters. Although the few papers referring to these telangiectasias assume them to be venules, to the best of our knowledge, proper immunohistochemical study of the nature of these vessels has not yet been performed. Therefore, we embarked upon the study of podoplanin expression in the dilated dermal vessels of Gottron's papule.

CASE REPORT

A 65-year-old woman complained of pruritus and swollen eyelids. The pruritus had started 1 month before and was first located on both soles, soon thereafter involving the trunk, neck and scalp. Physical examination revealed desquamative erythema on the scalp and back. There was also palpebral edema and the eyelids appeared erythematous and desquamative. Four to six months later, the patient developed periungual erythema with red papules on the dorsum of her fingers at the level of the interphalangeal joints and metacarpophalangeal joints, which were diagnosed as Gottron's papules. Similar lesions were seen on her right elbow. The patient started complaining of muscular weakness.

Electromyography revealed changes that suggested nonspecific myopathy. Laboratory parameters including muscular enzymes were normal.

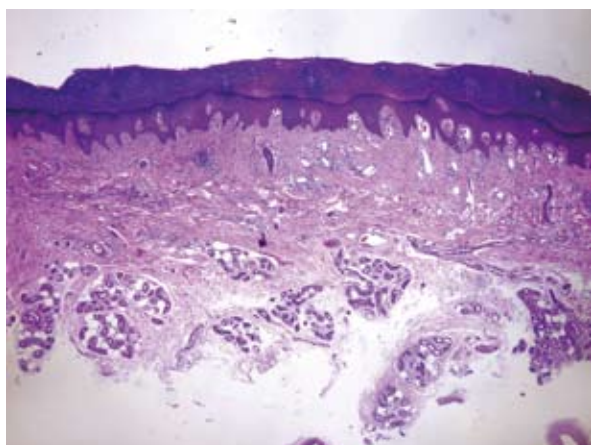


Figure 1. Low power view of the biopsy specimen obtained from one of the Gottron's papules. There is a psoriasiform hyperplasia of the epidermis, with mild inflammatory infiltrate, perivascular, superficial and deep. Even at this magnification (X20), dilated vessels are remarkable in the papillary and reticular dermis.

She was diagnosed with dermatomyositis and investigated for internal malignancy, which was not found.

A biopsy was obtained from one of Gottron's papules of the hand. The epidermis showed psoriasiform hyperplasia. There was a perivascular chronic infiltrate which was superficial and deep (Fig. 1). Dermal vessels from the dermis (papillary and reticular) were dilated (Fig. 2). The inflammatory infiltrate was mainly composed of CD4+ T-cells (Dakocytomation, monoclonal mouse anti-human, clone MT310, code M0716), with a lesser amount of CD8+ T-cells (Dakocytomation, monoclonal mouse anti-human, clone C8/144B, code M0103) and scattered CD57+ cells (Dakocytomation, monoclonal mouse anti-human, clone TB01, code M7271) (Fig. 3). There was a small amount of CD68+ cells (Dakocytomation, monoclonal mouse anti-human, clone PG-M1, code M0876) (Fig. 3). The study with D2-40 (Dakocytomation, monoclonal mouse anti-human, clone D2-40, code M3619) showed immunoexpression in only some of the dilated vessels, while many of them did not express the marker (Fig. 4).

DISCUSSION

Cutaneous involvement by dermatomyositis includes violaceous heliotropic swelling of the eyelids and cutaneous eruption on the sun exposed areas (1). The histopathology of such lesions has been presented in many reports before (2-5). On the contrary, the histopathology of Gottron's

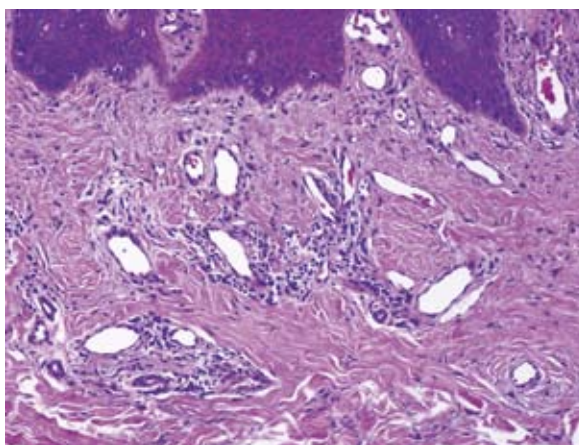


Figure 2. A closer view of the dilated vessels of the dermis and of the perivascular inflammatory infiltrate (X20).

papules, which also are a clinical feature of dermatomyositis, has been rarely reported (6-8). Moreover, the publications referring to the histopathology of Gottron's papules have mainly focused on changes in the epidermis and the interface, and on the nature of the inflammatory infiltrate (6-8). For instance, Hanno et al. studied 11 cases of Gottron's papules and describe changes in the epidermis, the basement membrane zone and the dermis (6). In the dermis, they mention mucin deposit and inflammatory infiltrate. Although the term 'telangiectasia' is not included in their description, their Figures 2 and 3 show biopsies with dilated vessels.

Kovacs and Kovacs mention that in cutaneous lesions of dermatomyositis "hyperkeratosis,

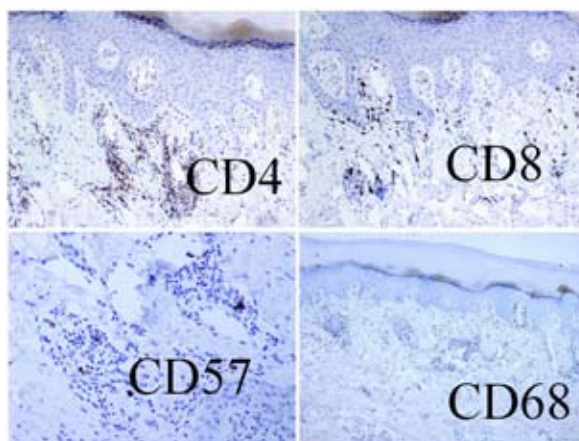


Figure 3. Immunohistochemical study of the inflammatory infiltrate showing it to be mainly composed of CD4+ T-cells (X10), with a lesser amount of CD8+ T-cells (X10), scattered CD57+ cells (X4) and CD68+ cells present in a moderate amount (X20).

vacuolization of the basal keratinocytes, melanin incontinence, perivascular lymphocytic infiltrate, increased dermal edema and mucin deposition, acanthosis and epidermal atrophy are the characteristic light microscopic findings" (9). They also claim that "*hyperkeratosis is seen in Gottron's papules, whereas poikilodermatous lesions have an atrophic epidermis*" (9). Therefore, dilated cutaneous vessels are not mentioned and no histopathologic figures are included in the report.

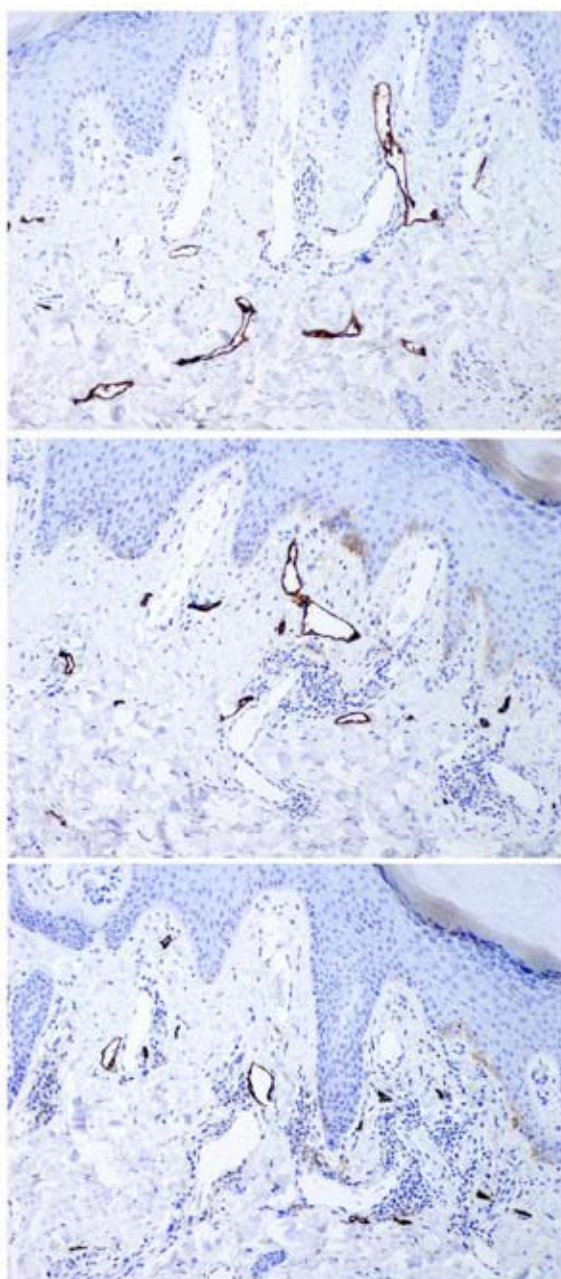


Figure 4. Immunostaining with D2-40 for podoplanin showing telangiectasias that do not express the marker. Some dilated lymphatic vessels are strongly marked with the antibody (X100).

Mendese *et al.* present two cases of Gottron's papules, in which they describe hyperkeratosis, papillomatosis, hypergranulosis, epidermal hyperplasia or atrophy, spongiosis, necrotic dyskeratotic keratinocytes, thickened basal membrane and interface change. Although they do not describe it in words, their Figures 5, 6 and 7 referring to case 2 show prominent telangiectasias (8).

Some classic books also neglect description of these dilated vessels when talking about Gottron's papules, and focus on epidermal changes instead. In their book, McKee *et al.* describe Gottron's papules as being characterized by "*hyperkeratosis, mild papillomatosis, acanthosis or, less often, epidermal atrophy and the features of an interface dermatitis*" (11). Nevertheless, they show only a histopathologic picture of a Gottron's papule, in which telangiectasias are a prominent feature (10). In Lever's Pathology of the Skin, Jaworsky describes Gottron's papules as having "*vacuolization of the basal cell layer, but [with] acanthosis rather than epidermal atrophy*", with no mention of telangiectasias (11).

Other texts recognize that there are dilated blood vessels in Gottron's papules, but they claim they are venules. For instance, Ackerman *et al.* in their chapter on dermatomyositis in their Histologic Diagnosis of Inflammatory Skin Diseases claim that "*Gottron's papules are formed by all the changes that characterize lesions of dermatomyositis*" (12). Among the latter, they include "*venules of the superficial plexus dilated widely*" (12). Dilated vessels are also described in cutaneous biopsies of dermatomyositis in poikilodermatous lesions (10). In fact, the first large case series on dermatomyositis demonstrated poikiloderma by histopathologic criteria in 19 of 24 dermatomyositis specimens (although the type of clinical lesions was not specified) (3). In a very recent report, Smith *et al.* (13) studied 13 biopsies of Gottron's papules and found telangiectasias in 7 (54 %) of them, although they were only mild or moderate (and not marked). Nevertheless, telangiectasia is defined as "*a condition typified by abnormal, permanent dilation of venules mainly but also, at times, of capillaries and arterioles*" (14).

In our study, we corroborated the venules to be dilated in Gottron's papules, but we also found another type of dilated vessels, which immunostained as lymphatic. Dermal lymphatic vessels are usually inconspicuous in normal skin (15). Therefore, dilated lymphatic vessels can be partly responsible for the clinical aspect as a papule in Gottron's lesions. We used D2-40 marker, which

stains cells containing podoplanin (16) and is considered as a good marker for lymphatic differentiation (17-19).

Regarding the immunophenotype of the inflammatory infiltrate, our study corroborated previous studies demonstrating activated CD4+ Th lymphocytes to predominate in the inflammatory infiltrate of Gottron's papules (7). This infiltrate is similar to the one observed in other organs of dermatomyositis patients. For instance, a predominance of CD4+ T cells was demonstrated in the pericardial fluid of a dermatomyositis patient (20), and these patients have also been shown to have a significantly lower percentage of total CD3+ CD8+ subset in peripheral blood than controls (21). Some authors report on muscle biopsies from dermatomyositis patients to show a prevalence of CD4+ helper/inducer T cells over CD8+ T cells (22), whereas others report on the inflammatory infiltrate composition as follows: 18% T4+ cells; 32% T8+ cells; 18% B cells; and 32% macrophages (23).

CONCLUSIONS

Dilated lymphatic vessels (D240 +) were demonstrated in our case of Gottron's papules. Similar findings are observed yet not mentioned in some papers showing histologic pictures of Gottron's papules. We think that dilated lymphatic vessels may be, at least partly, responsible for the clinical swollen appearance of Gottron's papules.

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