Segmental Neurofibromatosis versus Giant Nevus Spilus

Sir,

Segmental neurofibromatosis (NF-5) is characterized by café-au-lait (CAL) spots, and/or cutaneous neurofibromas limited to a circumscribed body segment (1). However, in the absence of clear diagnostic criteria for NF-5 it is difficult to establish the diagnosis in many cases. The recent reports by Selvaag et al. (2) and Menni et al. (3) in this journal demonstrate the difficulty in diagnosis of NF-5. No unique histological feature defines a lesion as a CAL spot (3). Some immunohistochemical (4) and ultrastructural (5) features may provide a clue for CAL but are not diagnostic.

The diagnosis of NF-5 was occasionally applied to patients with giant nevus spilus (6), despite insufficient clinical and histological evidence (7). Likewise, in Selvaag's case, the lentiginous hyperplasia and the presence of nevus cell nests are characteristic features of speckled lentiginous nevus (8) and are not diagnostic of CAL. Therefore, this case may represent unilateral lentiginosis rather than NF-5.

NF-5 is regarded as a mosaic form of other types of neurofibromatoses (mainly NF-1). Such mosaicism may also involve the gonads (9), resulting in transmission of full-blown NF-1 to an offspring of a parent with NF-5 (10). Consequently, patients with NF-5 may not be reassured that there is no risk of NF-1 in their offspring. Considering this, we may better serve our patients by restricting the diagnosis of NF-5 to cases that comply with Riccardi's criteria. For the patient this may prevent unjustified stress and, possibly, unnecessary investigations.

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Cutaneous Vasculitis with Lesions Mimicking Degos' Disease and Revealing Crohn's Disease

Sir,

Extra-intestinal manifestations of Crohn's disease (CD) are diverse and may antedate, occur with, or postdate the onset of the bowel disease itself. Skin findings, such as erythema nodosum and pyoderma gangrenosum, are not infrequent, whereas vasculitis is rare (1, 2). We report a case of leukocytoclastic vasculitis revealing CD. The patient also had atrophic and papular lesions mimicking Degos' disease.

CASE REPORT

A 33-year-old woman presented with a complaint of skin lesions, of 16 months' duration. Past medical history included only a single episode of bloody diarrhoea and lower abdominal pain, without neurologic symptoms. This episode had occurred at the onset of the cutaneous disorder. On initial examination, about 10 erythematous papules, 3–8 mm in size, were scattered randomly on her trunk and extremities. The lesions were asymptomatic and presented initially as obvious circumferential full thickness necrosis. They developed into atrophic, porcelain-white scars, ringed by an erythematous telangiectatic border. A few of them had an easily removable adherent scale centrally (Fig. 1). In addition, on both lower legs, physical examination revealed papular petechiae. Other physical findings were within normal limits. Initial usual laboratory tests showed the following values: erythrocyte sedimentation rate, 37 mm/h; WBC count, 10,900/mm^3; hemoglobin, 9,2 mmol/l; platelet count, 307,000/mm^3. Prothrombin and partial thromboplastin times, the total protein level, the globulin fraction, the albumin fraction and complement levels were within normal limits. Urinalysis was normal. The search for cryoglobulin, cryofibrinogen, anti-phospholipid antibodies, anti-nuclear antibodies and anti-DNA antibodies was negative. In vitro platelet function was not evaluated. Histologic findings in a 4-mm punch biopsy specimen from a palpable small purpuric lesion of a leg were consistent with leukocytoclastic vasculitis. Histologic examination of a papular lesion from the abdomen showed a well-defined wedge-shaped area of coagulative necrosis involving the dermis. Within the necrotic area, slight mucin was seen with alcian blue and colloidal iron stain. At the base, the walls of venules and arterioles showed deposits of fibrin and were infiltrated by mononuclear cells and a few neutrophils, with scattered nuclear fragments (Fig. 2). At the lateral borders, the same pattern of endovasculitis with endothelial cell hypertrophy and fibrin thrombi affected the superficial vessels. These findings were consistent with Degos' disease and prompted us to perform a colonoscopy, despite the paucity of the abdominal symptoms. It showed numerous aphthoid-like ulcers, whose histologic analysis demonstrated features of CD. A mesalazine regimen, 2 g per day, resulted in a progressive improvement of the cutaneous lesions and was maintained

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for 12 months. The patient refused another colonoscopy for control. No relapse has occurred, with a 6-month follow-up.

DISCUSSION

Occurrence of leukocytoclastic vasculitis in patients with CD is a rare but well-known feature (1). Although in these cases, the vasculitis is supposed to be related to CD, specific bowel treatments usually do not result in a significant improvement of the cutaneous changes. In addition to classic purpura, our patient's skin lesions included also necrotic and papular lesions leading to atrophic scars that might suggest the diagnosis of Degos' disease.

Degos' disease, or malignant atrophic papulosis, is a multisystem disease characterized by endovasculitis with thrombosis involving the skin, gastrointestinal tract, central nervous system and, less commonly, other visceral organs. In the lack of characteristic laboratory findings, Degos' disease is diagnosed on the basis of its clinical picture of papules surrounded by a china-white telangiectatic fringe with atrophic scarring (2). They usually do not ulcerate but have a thin rim of epidermis overlying them. The histologic appearance is distinctive, with a wedge-shaped zone of ischemia extending from the epidermis through the dermis, deposits of mucin in the early stage and a widespread necrotizing, mainly lymphocytic, vasculitis that affects the entire cutaneous microvasculature, both venules and arterioles (3). The 3-year survival rate of patients with systemic involvement has been estimated at 50%. Degos' disease is usually not thought to be an auto-immune process and is usually not associated with autoimmune disorders, in particular leukocytoclastic vasculitis.

However, rare cases of lesions mimicking Degos' disease, sometimes named symptomatic Degos' disease, have been observed accompanying autoimmune disorders, such as systemic lupus erythematosus, progressive systemic sclerosis and polymyositis (4–7), with a cutaneous vasculitis in one case. Despite an identical histologic picture, these cases are probably distinct from true Degos' disease and should instead be referred to as "Degos'-like lesions". Indeed, these lesions seem rather to be due to the underlying disease than to be cases of association between true Degos' disease and auto-immune disorders. In particular, they do not share the pejorative prognosis of "malignant papulosis" and do not worsen the prognosis of the underlying disease. We believe that our case is the first case of symptomatic Degos' disease associated with CD. We suggest that Degos'-like lesions, characterized by ischemia due to the thrombosis of the deep reticular dermis blood vessels, were probably linked with leukocytoclastic vasculitis, through the same process leading to vasculitis.

In conclusion, occurrence of leukocytoclastic vasculitis in association with minimal abdominal symptoms should prompt the clinician to perform a colonoscopy, and Degos'-like lesions must be added to the list of cutaneous findings associated with CD.

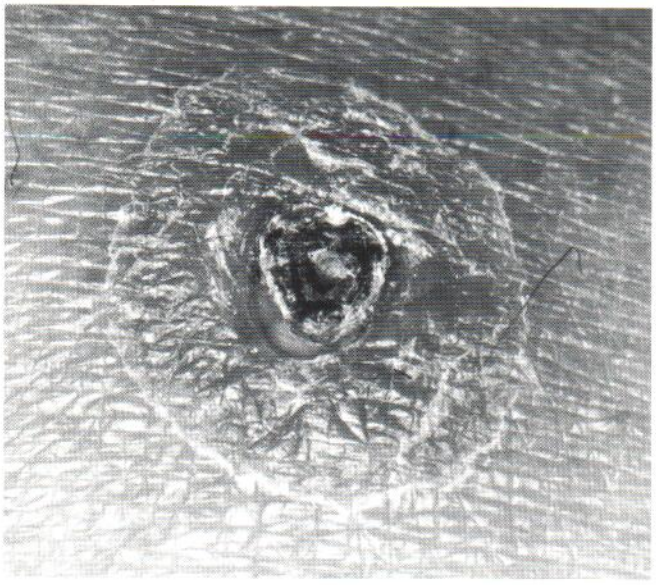

Fig. 1. Necrotic papule with an easily removable adherent scale centrally.

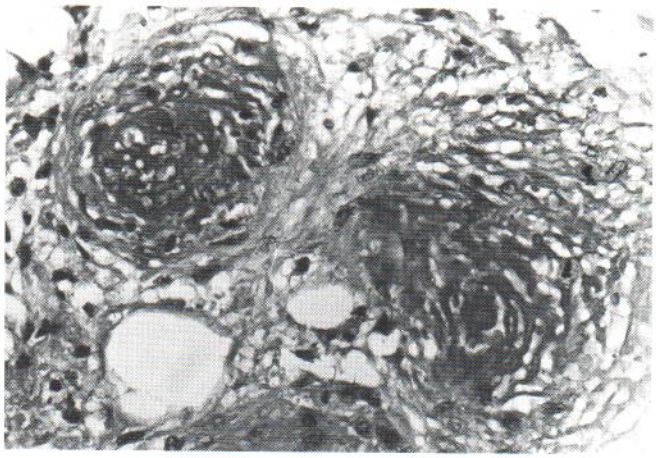

Fig. 2. At the base of the area of necrosis, the walls of venules and arterioles show deposits of fibrin and are infiltrated by mononuclear cells and a few numbers of neutrophils, with scattered nuclear fragments (HES ×400).

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