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BENIGN CUTANEOUS MEDIUM VESSEL VASCULITIS (CUTANEOUS PERIARTERITIS NODOSA)

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A case of cutaneous periarteritis nodosa had swelling and intense pain on right lower leg followed by bullae. Incision of bullae resulted in chronic ulcers with peripheral, indurated, violaceus discoloration, livido reticularis and multiple nodules.

Key words : Periarteritis nodosa, Cutaneous Vasculitis.

Benign cutaneous periarteritis nodosa is a distinct clinical entity in which cutaneous lesions predominate and there is no visceral involvement. It was first established as a definite entity by Lyell and Church in 1954.[1] Ruiter,[2] Perry[3] and Golding[4] also agreed that systemic and cutaneous periarteritis nodosa were two distinct entities. Some authors believe that cutaneous changes become an outstanding feature if systemic involvement is less and two forms of the same disease depend on the duration and severity of systemic involvement.[4] Clinically, crops of nodules, livido, ulceration, incipient gangrene, myositis, peripheral neuritis, arthralgia, fever and anaemia are important characteristics.[5-9] Benign cutaneous periarteritis nodosa is a very rare disease.

Case Report

A 60-year male developed swelling and pain in the right leg four months back which was followed by bullae within 2-3 days. Four incisions were given on the right leg which developed into non-healing ulcers. There was no history of fever, intermittent claudication, trauma, intercurrent infection, diabetes mellitus, intake of drugs, hypertension, allergy and aneasthesia. Examination revealed oedema of the right leg below the knee with an indurated dusky violaceus discoloration upto the toes with a few scales, crusts and reticulated vascular pattern. The biggest ulcer was 10 cm × 7 cm on the anterior surface of the right leg with a sharp straight margin and a thick edge and pus oozed from underneath the edge (Fig. 1). Floor of the ulcer had pinkish granulation tissue with a minimal discharge. Other three ulcers were roughly 3 cm × 2 cm in size, on the front and back of right leg, and the dorsum of right foot respectively. Multiple, erythematous, violaceus, non-tender, firm nodules and papules were present around the big ulcer, most prominently above its upper margin (Fig. 1). Temperature

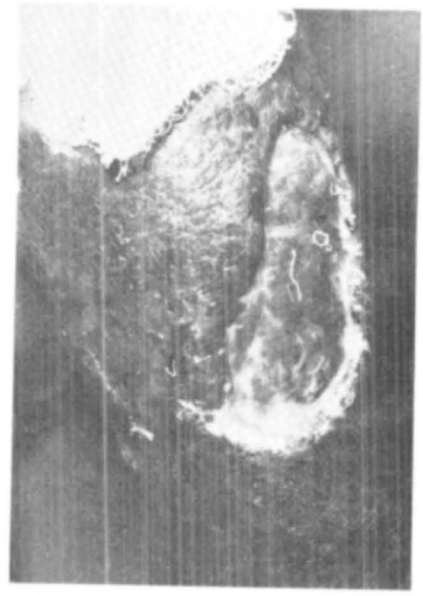

Fig. 1. A large ulcer with sharp straight margins and thickening of peripheral skin.

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was slightly raised over the right leg. Posterior tibial and dorsalis pedis arteries were pulsatile.

Routine investigations of urine, stools and blood were normal. ESR was 30 mm. Fasting blood sugar was 80 mg% and blood urea 26 mg. X-ray leg showed irregularity of the anterior and posterior aspects of the leg, soft tissue swelling with loss of muscle planes and osteomyelitis fibula. Biopsy from the margin of the ulcer revealed a number of vessels in deep dermis showing segmental and complete degeneration of vessel wall with deposition of fibrinoid material. Infiltrate was present within and around the arteries (Fig. 2). Some vessels showed intimal proliferation, thrombosis with occlusion of the lumen, and mixed cellular infiltration into the tissue surrounding the arteries (Fig. 3).

Comments

Immune complex deposits and cell mediated immunity play an important role in the pathogenesis of vasculitis. Polyarteritis or periarteritis nodosa group of systemic necrotising vasculitis belongs to the medium arterial vasculitis group.[10,11] Cutaneous periarteritis nodosa has a good prognosis as spontaneous remission occurs after some interval.

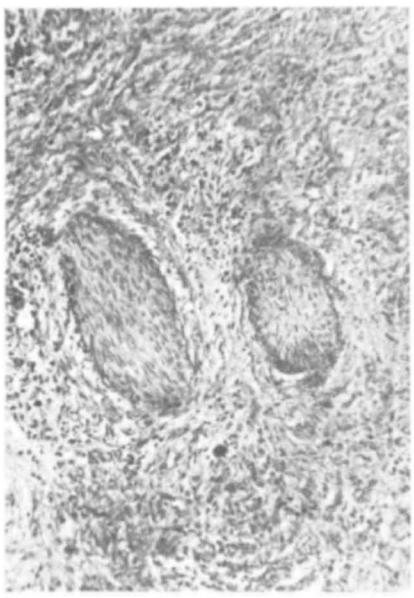

Fig. 2. Panarteritis of blood vessel with partial destruction of its wall and deposition of fibrinoid material. Inflammatory cells invade the vessel wall and the surrounding tissue.

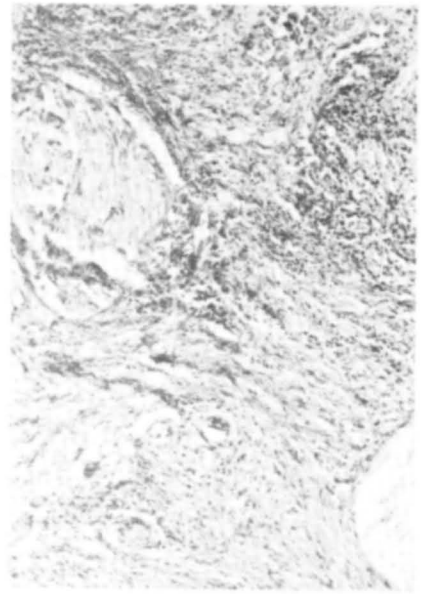

Fig. 3. Two vessels in the deep dermis with intimal proliferation, thrombosis and occlusion of their lumen.

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