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the blood vessels. The histological features were compatible with nevus lipomatosus [Figures 3a and b].

On magnetic resonance imaging, abnormal areas of fat deposition on the left side of the face, left flank and thigh in the subcutaneous plane with no infiltration of the internal organs and muscles were seen.

The patient was referred to the plastic surgery department for surgical excision.

NLCS is a rare disorder characterized by nevoid accumulation of mature adipocytes in the dermis. It often presents at birth but can appear within the first two decades of life, as in our case.[2] There is no familial or sex predilection.

Two clinical types are recognized. The classical type, reported by Hoffman and Zurhelle, consists of multiple flesh-colored or yellowish sessile lesions with a tendency to coalesce into plaques with a smooth or corrugated surface and zonal distribution and following natural cleavage lines of the skin. They have a predilection for the pelvic girdle and sacral and lumbar regions.

The second type manifests as small solitary nodules mimicking skin tags and occurring over the arms, knees, axillae, ears and scalp. They appear at a later stage of life. Occurrence over atypical sites like the nose, calf and clitoris has also been reported. In our patient, both the plaques were of the classical type, the second one being a giant NLCS, 40 cm × 28 cm in size. The largest size reported so far has been a 20 cm × 30 cm-sized bilateral cerebriform plaque extending from the lower abdomen to the left leg in a 36-year-old Brazilian woman.[3]

NLCS has been reported in association with café au lait macules, leucodermic spots, hypertrichosis over the nevus, comedo-like lesions and angiokeratoma of Fordyce.[4]

The pathogenesis of NLCS is unknown and several theories have been put forth. Proposed pathogenesis include adipose metaplasia during the course of degenerative changes in dermal connective tissue, as stated by Hoffman and Zurhelle and supported by Nikolowsky, and developmental displacement (heterotopia) of adipocytes or development of mature adipocytes by mesenchymal perivascular cells present initially as primitive lipoblasts that subsequently transform into mature adipocytes.[2]

On histopathology, the distinguishing feature is presence of ectopic mature adipocytes in the dermis intermingled with collagen bundles and perivascular infiltration of dermis and subcutis with chronic inflammatory cells.[2]

This condition may be confused with plexiform neurofibroma, connective tissue nevus, vascular malformation, lipomatosis or lipoblastomatosis. The treatment of choice is surgical excision.[5] Patients unwilling for surgery may undergo cryotherapy, which yields partial though satisfactory results.[4]

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Nifedipine-induced acute generalized exanthematous pustulosis in a case of acute glomerulonephritis

Sir,

Acute generalized exanthematous pustulosis (AGEP) is a disease characterized by the rapid occurrence of many sterile nonfollicular pustules usually arising on an edematous erythema, often accompanied by leucocytosis and fever.[1] Drugs are attributed as the most important cause of AGEP, although, rarely, it can be due to infections or idiopathic.[2] Herein, we report a case of AGEP induced by nifedipine in a patient of acute glomerulonephritis.

Nifedipine-induced acute generalized exanthematous pustulosis in a case of acute glomerulonephritis

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A 27-year-old male patient diagnosed as a case of acute glomerulonephritis on furosemide and enalapril for last 3 months was admitted to the medical unit of our institute with severe hypertension and was prescribed nifedipine for control of the hypertension. On the third day of treatment with nifedipine, he developed fever, which was followed by a generalized skin rash consisting of numerous discrete 2-3 mm pustules over an erythematous skin affecting the upper extremity, chest, abdomen and thighs [Figure 1]. The patient’s past medical history was negative for any similar eruptions, psoriasis or any drug allergies. On bacteriological and mycological examination of the pustules, no organisms could be isolated. Tzanck test was negative for any acantholytic cells or multinucleated giant cells. A skin biopsy showed spongiosis with dense subcorneal neutrophilic infiltrate and edematous papillary dermis with perivascular mixed mononuclear cell infiltrate. Based on the clinical features, negative microbiological study and histopathological findings, a diagnosis of AGEP was made and the suspected drug nifedipine was withdrawn and the patient prescribed oral cetrizine and paracetamol. After 2 days of stopping nifedipine, the patient became afebrile and there were no further new eruptions. At the end of 1 week, all the skin lesions were healing with desquamation.

Cutaneous drug reactions occur at a frequency of 1-8% and can be higher for certain classes of drugs.[3] AGEP is a severe cutaneous adverse reaction and has a typical clinical evolution pattern, in that the eruptions occur 24 h-3 weeks (average, 5 days) after administration of the offending drug and quickly resolve on discontinuation of the drug in a 1-2 week time period.[1,2,4,5] A recent multinational case control study, Euro SCAR, has found the highest risk of AGEP due to drugs like antibiotics (macrolides-pristinomycin, aminopenicillin, quinolone, sulfonamide), anticonvulsants, hydroxychloroquine, terbinafine and diltiazem.[5] Other drugs reported to be associated are glucocorticoids, oxicam, nonsteroidal anti inflammatory drugs (NSAIDs), fluconazole, chlorpromazine, mesalazine, allopurinol, enalapril, furosemide, nystatin and hydroxyzine.[1-12] The Euro SCAR study did not find any association of AGEP with psoriasis or infective causes, although earlier studies have shown a higher incidence of AGEP in psoriasis and even some have considered it as an entity no different from pustular psoriasis.[6] However, the clinical evolution pattern of AGEP is different as compared with pustular psoriasis. AGEP is mediated by T-cells, which produce high levels of CXL8 (interleukin-8) and granulocyte monocyte CSF.[5] CXCL-8 is also produced by keratinocytes in the lesion of AGEP.[3] AGEP can be confused with other pustular cutaneous dermatoses like pustular psoriasis, drug hypersensitivity syndrome with pustules, subcorneal pustular dermatosis, pustular vasculitis, bacterial, viral and fungal infections and, in severe cases, from toxic epidermal necrolysis. The temporal relationship of the development of the eruption with the intake of the suspected drug and its prompt subsidence on withdrawal of the drug along with the histopathological findings and negative microbiological study helps to arrive at a diagnosis. Among the calcium channel blockers, diltiazem has a very high risk of developing the eruption.[1,13,14] Cross reactivity with other calcium channel blockers like nifedipine and verapamil has been demonstrated by patch testing.[13] A knowledge of the possible groups of drugs responsible for the reactions is essential for the clinicians as prompt withdrawal of the offending drug helps resolution of the eruption, which otherwise can be occasionally fatal. Some patients may require the use of systemic corticosteroids.[15] Because AGEP is a T-cell-mediated reaction, it is suppressed by steroids like any other T-cell-mediated reaction; nevertheless, it should be kept in mind that steroids can themselves be a cause of AGEP.[12,9]

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Figure 1: Numerous monomorphic pustules over the forearm
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