Methotrexate in autoimmune urticaria

Sir,

Chronic idiopathic urticaria may be autoimmune in origin (autoimmune urticaria), caused by functional autoantibodies that activate mast cells and basophils through cross linking the high affinity IgE receptor (FcεRIα) to secrete histamine.[1] An incidence of 30% to 50% has been reported by various investigators.[1] While there are no clinical features that distinguish ordinary urticaria from autoimmune urticaria, a simple test, the autologous serum skin test, can be used to detect functional autoantibodies. Patients with autoimmune urticaria whose disease is pursuing a severe disabling and recalcitrant course have been treated with immunosuppressive therapy, including cyclosporine.[2] However, its high cost makes this drug an impractical option in India. Gach et al successfully tried methotrexate in two patients without detectable autoantibodies and in whom steroids and antihistamines were not effective.[3] However, there are no randomized controlled studies of the use of methotrexate in patients with autoimmune urticaria.[4] We report our preliminary experience of using methotrexate in four patients with autoimmune urticaria.

We tested 45 patients (age ranging from 15 to 55 years) with chronic idiopathic urticaria with the autologous serum skin test for autoantibodies. Twelve of them showed a positive result, including four (3 females and 1 male) who were recalcitrant to treatment with oral antihistamines (fexofenadine, cetirizine, hydroxyzine). After performing baseline investigations (complete blood count, random blood sugar, SGPT, and urine examination), we tried methotrexate in these patients with autoimmune urticaria in a dose of 2.5 mg orally twice a day on Saturday and Sunday of every week. Informed consent was taken before starting methotrexate. In addition, cetirizine 10 mg and folic acid 1.5 mg were given daily. All four patients showed a remarkable effect in the form of reduction in whealing and itching in one month. Investigations were repeated after one month for monitoring of side effects. Treatment with methotrexate was continued for 2 months and later only cetirizine was continued. One patient developed a relapse within two weeks of stopping methotrexate and was again started on methotrexate.

All the four patients had troublesome urticaria that was difficult to control with antihistamines alone. After a course of methotrexate the urticaria was controllable with cetirizine in three patients. In India methotrexate has the potential of being a viable option for the treatment of resistant autoimmune urticaria as it is cost effective and most dermatologists have the experience of using it for psoriasis. A larger controlled study needs to be undertaken to confirm these preliminary findings.

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Unilateral angiokeratoma of fordyce

Sir,

Angiokeratoma of Fordyce is a localized form of angiokeratoma affecting the scrotum and is probably the commonest of the angiokeratomas. We report a rare case of angiokeratoma of Fordyce with unilateral occurrence without any underlying disorder. Such a case has been reported only once in the literature recently, in association with a unilateral varicocele.[1]

A 32 year old male rickshaw driver presented with

multiple asymptomatic reddish purple colored raised lesions on the left side of the scrotum since 2 years with progressive increase in number. There was no history of bleeding from the site or history of swelling on the left side of the scrotum.

On examination, he had multiple, discrete, soft, smooth surfaced purplish red papules present unilaterally on anterolateral aspect of left side of the scrotum (Figure 1). Palpation ruled out varicocele, epididymal mass or inguinal hernia. Skin biopsy from a purplish papule showed mild hyperkeratosis with large, numerous, dilated and congested capillaries in an expanded papillary dermis (Figure 2). USG abdomen, pelvis and scrotum were done to rule out any vascular anomaly. On the basis of clinical features and histopathology, the patient was diagnosed as angiokeratoma of Fordyce with unilateral presentation as an unusual finding. We treated this patient with radiofrequency cauterization.

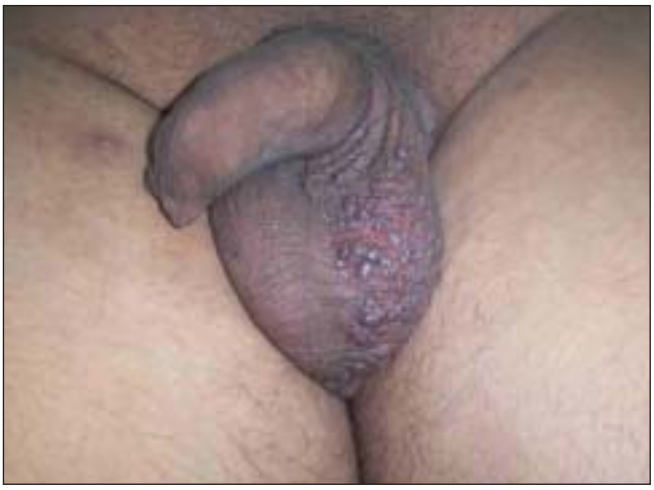

Figure 1: Angiokeratoma of Fordyce: unilateral lesions

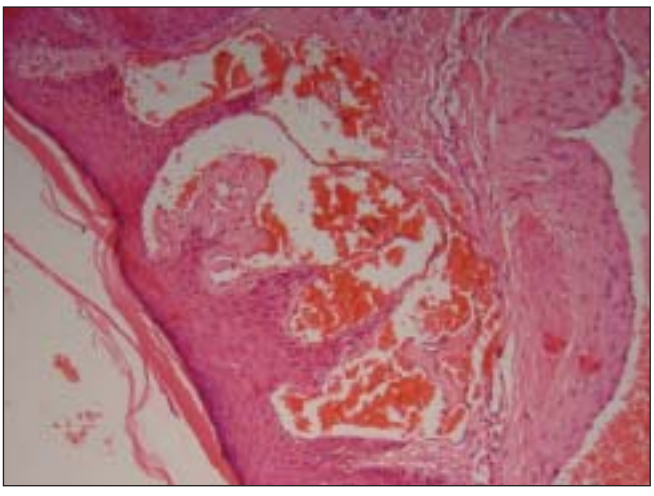

Figure 2: Dilated capillaries within enlarged dermal papillae. Epidermis shows mild hyperkeratosis (H/E, X200)

Scrotal angiokeratomas were first described by John Addison Fordyce in 1896.[2] Angiokeratomas are typically asymptomatic, 2 to 5 mm, blue-to-red papules with slightly keratotic surface. Commonest site is the scrotum with occasional affection of the shaft and glans of penis, and rarely leg, crural area and bulbar conjunctiva.[3,5] Histopathologically the lesions show one or more dilated subepidermal blood vessels within a widened dermal papilla. The epidermal changes in all forms of angiokeratoma are secondary to friction. The principle morbidity comes from bleeding, anxiety, and sometimes over-treatment due to misdiagnosis. Usually, they do not require treatment. If treatment is needed, laser, electrocoagulation, excision, or cryotherapy may be used.

Exact pathophysiology of angiokeratomas remains unknown, although it has been proposed that increased venous pressure may contribute to their formation.[4] There are many reports of angiokeratomas occurring in the presence of a varicocele or other conditions of increased venous pressure (e.g. hernias, epididymal tumors, urinary tract tumors).[5] On the other hand, in the majority of cases, no cause for increased venous pressure was found.[6] However, it is possible that in these cases, the increase in venous pressure may be so mild as to be undetectable by the ordinary methods leading only to capillary dilatation at a site that has probably the lowest tissue pressure.

In our case, examination and investigations ruled out any overt cause for increased venous pressure on the affected side. However, a small or microscopic vascular malformation may have led to unilateral but subclinical increase in venous pressure, thereby resulting in the ectasia.

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Klippel - Trenaunay syndrome: Association with absence of ipsilateral testis

Sir,

We wish to report a case of Klippel-Trenaunay Syndrome (KTS) with absence of ipsilateral testis, a hitherto unreported association. The KTS, first described by Klippel and Trenaunay in the year 1900, classically consists of a triad of port-wine stain of limb, varicose veins and hypertrophy of soft tissues and bone of the same side. However, the current definition includes port-wine stain and increased limb size, irrespective of the presence of bony overgrowth and varicosity of veins. Though the specific genetic abnormality causing the disease has not been identified till date, a para-dominant mode of inheritance is speculated.[1] Port-wine stain or macular telangiectatic vascular nevus is the earliest, most common and characteristic clinical lesion. It usually involves the lower limb causing hypertrophy of bone and soft tissue, characterized by an increase in the length and girth.

A 16 year old male presented with multiple erythematous macules and plaques involving posterior aspect of left lower limb and left lower abdomen. The macules were present since birth and the plaques, appearing as masses of soft tissue hypertrophy, were noticed by him since last 5 years. There was a history of bleeding from the plaques with the slightest trauma. He also had scoliosis and an abnormal gait.

On examination, he was found to have lengthening of the left lower limb and varicosities of veins over the same side (Figure 1). The areas of soft tissue hypertrophy were nontender but there was a history

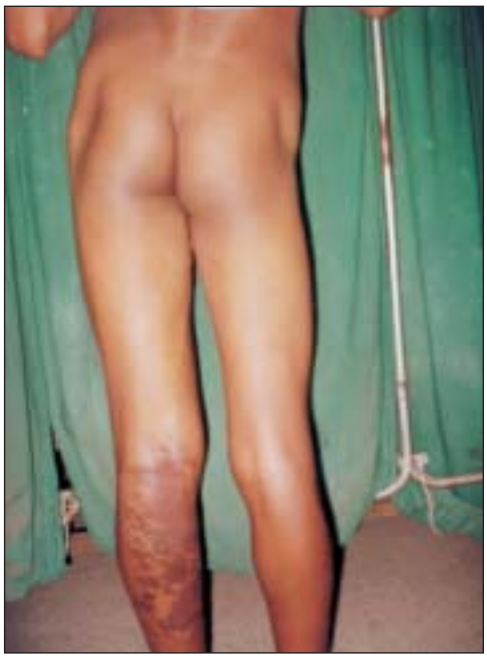

Figure 1: Port wine stain, dilated veins, soft tissue hypertrophy and scoliosis

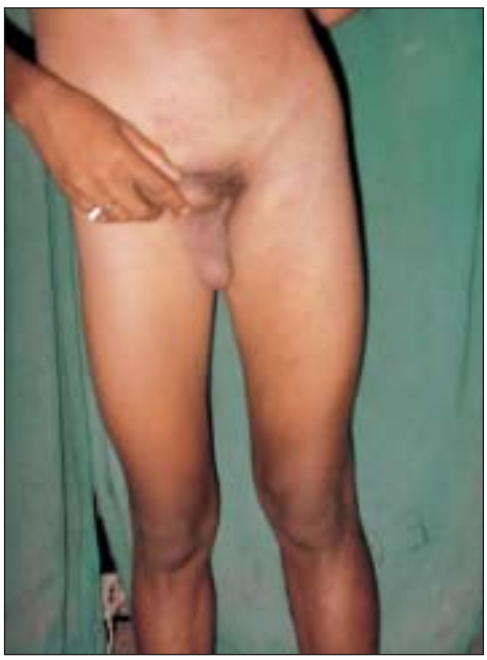

Figure 2: Absence of ipsilateral testis