Correspondence

Roberto Cortès, MD, Editor

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Melorheostotic skin changes

Case report

A 9-year-old Turkish girl complained of thickened skin over the left thigh and buttock. Hyperpigmented macules on the left buttock had been present since birth, and these lesions had gradually enlarged and thickened. New lesions had appeared gradually over recent years.

On physical examination, there were hyperpigmented and indurated plaques over the inner left thigh and buttock. Hyperpigmented macular lesions were present on the posterior part of the left thigh and inner part of the right thigh (Fig. 1). Hyperpigmented oval macular lesions with hypertrichosis were observed symmetrically on the anterior-medial surface of the lower legs (Fig. 2). Hypertrichosis was limited to the hyperpigmented macular lesions. There was no record of any similar condition in the patient's family history. Accessory tragi were seen on the upper side of the left tragus. The patient and her brother had suffered from deafness since birth.

Skin biopsy of the thickened area was performed. Dermatopathologic examination revealed increased melanin pigmentation in the basal layer of the epidermis. There was an increased amount of collagen within the reticular dermis. Sclerotic collagen bundles extended through the subcutaneous fat. These changes were suggestive of morphea, and therefore the papillary dermis was not involved.

Radiographs of both lower legs and pelvis were normal. Bone scintigraphy was performed 3 h after intravenous injection of 10 mCi ^{99m}Tc-methylene diphosphonate (^{99m}Tc-MDP, Tecnegas MDP, Mallinkrodt Medical, The Netherlands). Anterior and posterior whole-body images (256 × 256 word matrix, present counts of 500,000) were obtained with a gamma camera (Toshiba GCA 601e) using a parallel hole, medium energy, all-purpose collimator. Scintigraphic images were interpreted visually to assess any abnormalities. Diffuse, superficial, mildly increased tracer activity on the anterior aspect of the left thigh and diffuse intance uptake in the lower lumbar spine and sacroiliac fields appeared.

Discussion

Melorheostosis, a rare bone dysplasia, does not manifest itself until late childhood or early adolescence. It usually progresses into adult life. Melorheostosis with circumscriptum scleroderma, sclerodermatous skin changes with hypertrichosis, and strawberry marks has been described.

In this case, hyperpigmented and indurated plaques were histopathologically suggestive of morphea. In addition to these lesions, our patient had hyperpigmented macular lesions with hypertrichosis and the bone changes of melorheostosis. All of these signs enabled us to make a diagnosis of melorheostotic skin changes.

Melorheostosis is a rare monostotic or polyostotic bone disease of unknown etiology. It is characterized by a slowly progressing linear hyperostosis, fibrosis of the skin and subcutaneous tissue, contractures of the joints, and aching pain. The lower extremities are more commonly involved than the upper extremities.[1]

Melorheostosis is not a hereditary condition. It is most likely a congenital entity that does not manifest itself until late childhood or early adolescence, with "insidious onset." It progresses into adult life. It typically presents with irregular thickening of the cortex, generally on the side of the bone. A distinctive feature of hyperostotic bone is that its appearance resembles the wax dripping down the side of a candle.[2]

Melorheostosis with several skin changes has been described. Maroteaux and Pascoud described a form of melorheostosis combining osteopecilia and circumscriptum scleroderma.[3] Melorheostosis with fibrosis of the skin and

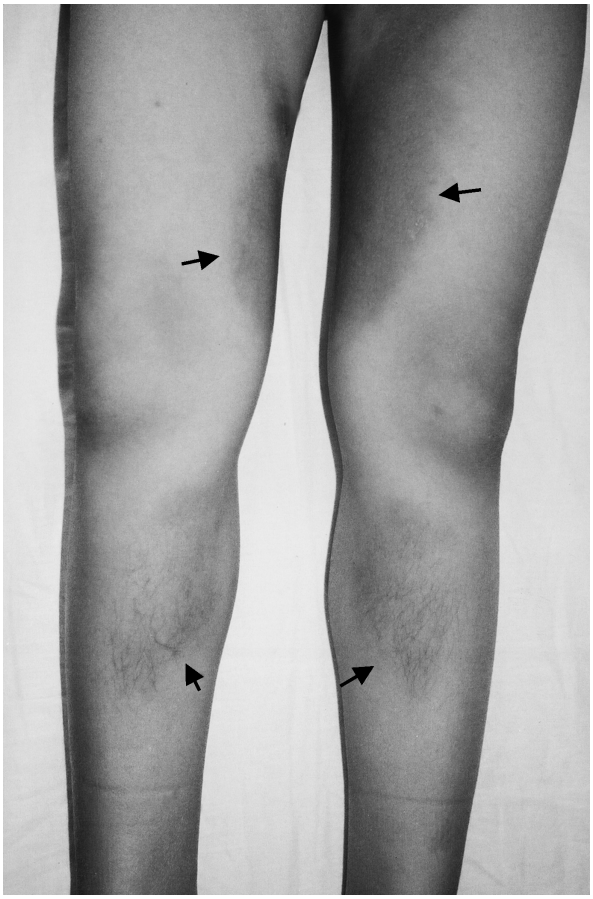

Figure 1 Hyperpigmented, indurated areas; hyperpigmented areas with or without hypertrichosis on the lower legs

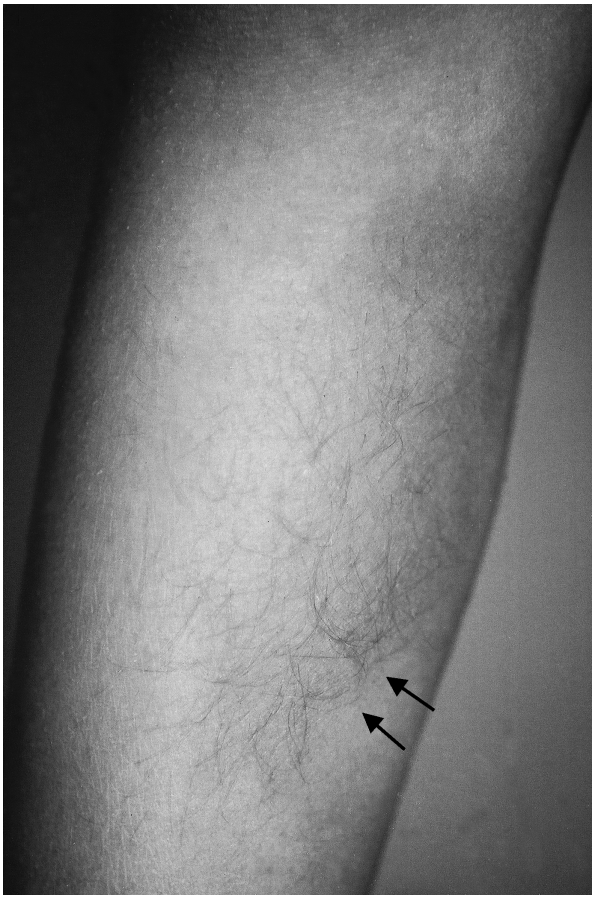

Figure 2 Hyperpigmentation with hypertrichosis

subcutaneous tissues has been reported by Steffens and Koob.[1] Fikry *et al.*[4] also described melorheostosis of a lower limb with calcification of the soft tissue.

Takeda *et al.*[5] reported a case of melorheostosis with linear sclerodermatous skin changes. Their case complained of pain in the left elbow joint and thickened skin over the left upper limb. Histopathology of the thickened area demonstrated a normal epidermis and a proliferation of normal appearing collagen fibers in the subcutaneous fat with no inflammatory changes. Miyachi described linear melorheostotic scleroderma with hypertrichosis.[6]

Numerous soft tissue and vascular anomalies have been reported in patients with melorheostosis.[7] A case of melorheostosis with minimal change nephrotic syndrome, mesenteric fibromatosis, and capillary hemangiomas was reported by Roger *et al.*[8] Chauc described Raynaud disease associated with melorheostosis.[3]

Moreno-Alvarez *et al.*[9] reported a case of a 9-year-old boy with linear scleroderma and melorheostosis of the iliac bone. Yamamato *et al.*[10] observed two painless masses occurring in the subcutis of the posterior aspect of the right forearm over excrescences of the underlying ulna due to melorheostotic deformity. Histopathologically, proliferation and occasional eosinophilic degeneration of the collagen fibers were seen.

Table 1 Skin manifestations and other conditions associated with melorheostosis

Linear scleroderma with or without hypertrichosis[6,9]
Capillary hemangiomas[3]
Desmoid tumor[11]
Calcification of soft tissue[4]
Thickened skin[5]
Painless masses[10]
Atypical decubital fibroplasia[10]
Raynaud disease[3]
Minimal change nephrotic syndrome[8]
Mesenteric fibromatosis[8]
Osteopecilia[3]
Renal artery stenosis[7]

Melorheostosis may be associated with many conditions and skin disorders (Table 1).[3–11] In cases of linear sclerodermatous

skin changes with hypertrichosis and hyperpigmented macules with hypertrichosis, melorheostosis should be considered.

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X-linked ichthyosis and undescended testes

X-linked ichthyosis (XLI) is relatively common genetic disorder of keratinization that was first described by Cockayne[1] but was defined as a distinct entity by Wells and Kerr in 1965.[2,3] It is caused by a deficiency of the steroid sulfatase (STS) enzyme. The disease was characterized clinically by generalized scaling of the skin with large, dark-brown polygonal scales, more prominent on the extensor aspects of the limbs.[3] Extracutaneous manifestations include corneal opacity and cryptorchidism. We present one patient with XLI who was discovered to have undescended testes to emphasize the importance of testicular examination for all patients with this disease.

Case report

A previously healthy 5-year-old Saudi boy, whose birth had been by full-term spontaneous vaginal delivery, was admitted to the pediatric ward with a 2-week history of fever and malaise with the clinical impression of infectious mononucleosis. A dermatology consultation was requested because of skin changes present for the last 2 months. The skin changes affected mainly the trunk and extremities with no associated symptoms. The mother and father were first-degree cousins. They had three other boys and a girl. There was a history of similar skin changes in one 10-year-old brother. This brother was born after a difficult, prolonged labor. Skin examination revealed diffuse scaling with small brownish polygonal scales on the trunk (Figs 1 and 2) and extremities, more pronounced on the extensor surfaces of the extremities. Preauricular fine brown scales gave a dirty or unwashed appearance. Flexural

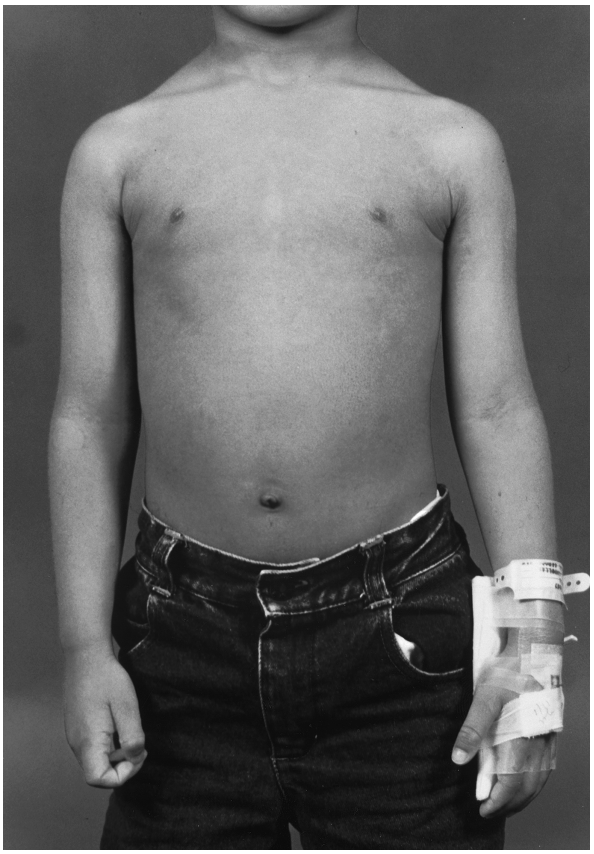

Figure 1 Diffuse scaling with small, brownish, polygonal scales on the trunk

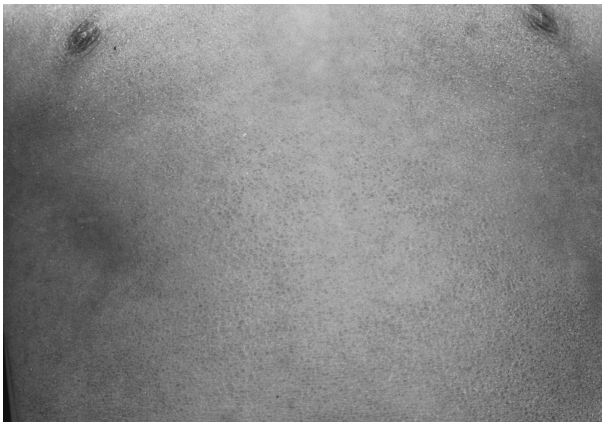

Figure 2 Close-up of the truncal skin showing the small, brownish polygonal scales

areas were relatively unaffected. Testicular examination revealed the left testis to be undescended. On the basis of the clinical findings, a diagnosis of XLI was made.

Skin biopsy showed hyperkeratotic stratum cornium with a normal granular layer, which is typical of XLI. Slit lamp examination of the eyes revealed no corneal opacities and normal eyes. Topical emollients containing 10% urea and lactic acid led to an impressive improvement in a few days and the child recovered completely from his viral illness.

Discussion

X-linked ichthyosis is the second most common type of ichthyosis after vulgaris.[4] The incidence of XLI is estimated to be between 1 : 2000 and 1 : 6000 male live births with no significant racial or geographic differences.[3] The disease is transmitted by carrier females as an X-linked recessive trait and usually affects males only.[3] A few female patients with the disease have, however, been reported.[4] The disease is caused by a deficiency of steroid sulfatase (STS) enzyme,[6,7] which is widely distributed throughout human adult and fetal tissues. The *STS* gene is located on the distal part of the short arm of the X chromosome (Xp 22.3).[3,5,8] Ninety per cent of patients with XLI show a complete deletion of the *STS* gene and the other 10% show only partial deletion or point mutation.[3,5] Deletion of this gene can occasionally extend to involve neighboring genes, causing a contiguous gene defect.[3,5,8,9] Therefore, XLI may be associated with Kallman's syndrome (hypogonadotrophic hypogonadism and anosmia)[3,5] and X-linked hondrodysplasia punctata.[3,10]

Cutaneous involvement in XLI is characterized clinically by early onset of generalized ichthyosiform hyperkeratotic[5] dark-brown polygonal scales with more involvement of the trunk and the extremities.[3] The flexures are usually less affected.[3] The face is usually free from scales except in the preauricular areas, which gives the classic "unwashed appearance" which is considered by many to be pathognomic.[11] The palms and soles are very rarely affected.[12,13] The hair and nails are normal.[14] Usually the disease improves during the summer and becomes worse during dry, cold weather.[3]

Asymptomatic corneal opacities in the posterior capsule of Descemet's membrane or the corneal stroma, not affecting the visual acuity, are the most common extracutaneous findings.[15,16] They are more frequent during the second and third decades of life[17,18] and are found in 10–50% of affected males and carrier females.[19] These opacities were found to be deposits of cholesterol sulfate crystals.

Cryptorchidism occurs in 10–20% of XLI patients, whereas the incidence in the normal population is 1%.[9] Moreover, independently of cryptorchidism, these patients are at increased risk of testicular cancer.[20] This anomaly could be explained either by a deficit in the STS enzyme or perhaps by a genetic disturbance located on the short arm of chromosome X close to the *STS* gene.[21,22] Testicular examination for male patients with XLI is mandatory and should not be omitted, as failure to detect undescended testicles may lead to potentially serious complications.

Steroid sulfatase deficiency during pregnancy in carrier females leads to an overall decrease in the levels of estrogen, causing poor cervical dilatation and prolonged labor not infrequently necessitating a Caesarean section.[3,4]

The diagnosis of XLI is usually made clinically and can be confirmed in many ways. Use of direct biochemical techniques demonstrating STS deficiency in one of the locations in which it is usually found (placenta, skin fibroblasts, leukocytes and keratinocytes[3,7]) is the most accurate means of confirming the diagnosis, but these techniques are not widely available. Demonstration of an increase in one of the substrates of STS, for example dihydroepiandrosterone sulfate (DHEAS) and cholesterol sulfate, is commonly used.[3] In serum, cholesterol sulfate, which is negatively charged, is carried by low-density lipoprotein (LDL) particles, leading to their rapid migration towards the positive pole during electrophoresis.[3,9] The most recent diagnostic methods are based on direct analysis of genetic material.[3] The use of laboratory techniques such as Southern blotting,[9,23] fluorescence *in situ* hybridization (FISH) and polymerase chain reaction (PCR)[5,9,24] has allowed the molecular detection of *STS* gene deletions. Prenatal diagnosis, utilizing non-invasive techniques demonstrating low estriol levels in maternal urine and plasma, is preferred. Invasive techniques such as amniocentesis to measure sulfated steroids in the amniotic fluid or skin biopsy through fetoscopy can also confirm the diagnosis.[3]

All treatments of XLI attempt to diminish the abnormal cohesion of corneocytes by facilitating their separation. Most patients benefit from topical keratolytics, emollients, and hydrating agents, and they rarely, if ever, require systemic agents.

We present this case to stress the importance of testicular examination whenever a patient with XLI newly presents,

as this may potentially prevent deleterious complications in this relatively common disorder.

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Calcipotriol ointment versus clobetasol ointment in localized vitiligo: an open, comparative clinical trial

Vitiligo is an acquired pigmentary disorder affecting 1% of the general population. There are many therapeutic agents for the treatment of localized vitiligo. Recently, topical calcipotriol ointment has been used for vitiligo either as a combined therapy or a monotherapy. The aim of this open, comparative clinical trial was to evaluate the efficacy and safety of calcipotriol ointment compared with clobetasol ointment.

Fifty-one patients with a definite clinical diagnosis of vitiligo were enrolled in this clinical trial. Five patients in the CAL (calcipotriol) group and four patients in the CLO (clobetasol) group withdrew from the study. Twenty-two patients in the CAL group and 20 patients in the CLO group were included in our study. Before starting therapy, all patients were evaluated clinically to record the duration of the disease, and the areas (cm^2) and the sites of the lesions. Lesions were not to exceed 10% of the body. Patients were also examined by Wood lights before and after treatment. The patients were instructed to apply calcipotriol ointment (50 μg/g) or clobetasol ointment (0.05%) twice daily for 4 months. Patients were assessed every months. Improvement was recorded as no change, minimal (0–25%), moderate (25–50%), marked (50–75%) and complete response (100%) depending upon the existing lesion's extent of repigmentation. The treatment was stopped when all the pigmented areas regained pigment.

Results

All the patients in each group were evaluated at the end of the therapy. Basic data including the age and sex of the patients,

Table 1 Demographic data of patients

	Calcipotriol (n = 22)	Clobetasol (n = 20)
Age	33.6	32.8
Sex		
Male	16	13
Female	6	7
Duration of disease		
< 1 years	15	14
1–3 years	2	3
> 3 years	5	3

Table 2 Clinical results

	Response	Calcipotriol (n)	Clobetasol (n)
Complete improvement	(100%)	0	4
Marked improvement	(75–100%)	0	4
Moderate improvement	(50–75%)	4	5
Minimal improvement	(25–50%)	8	4
No change	(0–25%)	10	3

and duration of the disease are showed in Table 1. The results of the study are showed in Table 2. Minimal response and no change results were 18 (82%) and 7 (35%) for the CLA and the CLO groups, respectively. Moderate, marked and complete response were 4 (18%) and 13 (65%) for the CLA and the CLO groups, respectively. There was significant statistical differences between the two drugs ($P > 0.05$). Mean lesional areas of 10.1 cm^2 to 8.3 cm^2 for the CAL group, and of 7.4 cm^2 to 4.0 cm^2 for the CLO group were detected. All patients tolerated well, but four patients in the CAL group complained of mild irritation and erythema. Symptoms were mild and transient. Seven patients in the CLO group complained of local erytema, acneiform papules, telangiectasia of the skin, but all were transient. There was no difference in the CAL and CLO groups regarding sex.

Discussion

Vitiligo is a progressive disease which affects 1–2% of the population, and results in the loss of skin and hair pigmentation. Although many topical methods have recently been tried, vitiligo remains one of the most difficult dermatological diseases to cure. Topical corticosteroids are the most commonly used therapeutic agents, but since 1970 several investigators have reported widely varying results with various topical steroids in vitiligo.[1–3] Clayton reported that 0.05% clobetasol propionate was significantly superior to a placebo in the treatment of vitiligo, and 12 out of 23 patients showed between 15 and 75% repigmentation.[1] Kumari was intermittently used as a topical application of clobetasol propionate in vitiligo. A repigmentation of 90–100% was achieved in more than 85% of patients with vitiligo of the face and more than 40% of patients with vitiligo of other body parts.[2] Khalid *et al.* found that 0.05% clobetasol propionate cream was effective in segmental vitiligo. Results were evaluated in 38 patients, and more than 50% repigmentation of the vitiliginous areas was observed in 13 patients (34.2%).[3] Recently, topical calcipotriol has been used for vitiligo as a combined or monotherapy.[4–7] Many investigators have suggested that calcipotriol may act either by 1.25 dihydroxyvitamin D3 receptors on melanocytes or by the modification of defective calcium transport in vitiliginous melanocytes. *In-vitro* experiments have demonstrated that vitamin D3 may enhance tyrosinase activity and melanin production.[4,5] But the exact role of calcipotriol in melanogenesis is not yet understood. Parsad *et al.* applied topical calcipotriol 50 μg/g to 21 patients, aged 5–17 years, with vitiligo. Patients exposed themselves to sunlight the next day for 10–15 min. At the end of the therapy, marked improvement and complete repigmentation was seen in 10 of the 18 patients. Four patients showed moderate improvement while the remaining four patients showed minimal improvement.[6] Ameen *et al.* declared that calcipotriol ointment was an effective and safe treatment for vitiligo. They used calcipotriol ointment twice daily in 11 patients with vitiligo for 2–9 months. Ten out of 11 patients showed between 30 and 100% improvement.[7] In the present study, we found that calcipotriol ointment showed a minimal therapeutic effect in localized vitiligo. The majority of patients in the CLO group showed more than 70% repigmentation presented 12 months onset of the disease. We conclude that autoimmune mechanisms in vitiligo are active during the first year of the disease. Calcipotriol and PUVA combined is a highly effective and efficient treatment, and may be used to shorten PUVA therapy when treating vitiligo.

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Comment on "Is a single dose of ivermectin sufficient in crusted scabies?"[1,2]

Even if the authors have chosen the application of two doses of ivermectin, a single dose could also clear scabies …

Previous reports on the efficacy of ivermectin in the treatment of scabies have given contradictory results owing to the incomparable regimes used. The main factor may be the difference in time between treatment and control/cure. Microscopic control should not be examined less than 2 weeks after therapy to allow all mites and mite products to debride. Secondly, when classifying cure by the decrease in pruritus, sufficient rehydrating skin care should be given because xerosis of the skin occurs due to topical antiscabiosa (which may have been applied before ivermectin treatment).

Standardized protocols should be established for the evaluation of the efficacy of ivermectin in the treatment of scabies.

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Hyper-IgE syndrome revisited

In the October 2001 issue of the *International Journal of Dermatology*, Shermer *et al.*[1] reported two patients with the "hyper-IgE syndrome" and reviewed the literature. As I was involved in early studies of this rare disorder and in differentiating it from cases of recurrent infection of atopic dermatitis,[2–4] this report has provided me with the opportunity to share my thoughts based on the examination of "many" patients over the years.

The best name for the disease is "hyperimmunoglobulin E syndrome with recurrent infection."[5] This cumbersome name avoids confusions inherent in the terms Job syndrome (implies coexisting red hair and hyperextendable joints), Hill–Quie syndrome (implies eczema), and even Buckley syndrome (implicates young boys). All patients have very high serum IgE levels associated with severe and recurrent infections involving the skin and other organs, especially the lungs. The disorder is rare and individual elements of the syndrome are variable and inconstant, leading to disagreement on who is and is not affected.[6] Here are my thoughts.

1 Infections. These are usually abscesses, often containing *Staphylococcus aureus*. They tend to be relatively nonpainful and cold, or at least cool to the touch, and may be long lasting. Patients with recurrent furunculosis in the absence of severe infections of the skin and other organs do not have this syndrome, even though patients with the syndrome often have furuncles. In addition to abscesses, patients invariably have severe and recurrent infections of other organs, especially the lungs and upper respiratory tract. Lung abscesses, pneumonias, bronchiectasis, pneumatoceles, sinusitis, discharging otitis media, and septic joints are examples of such infections and their sequelae. *S. aureus* is the major pathogen, but other encapsulated bacteria may also be present.

2 Eruption. There are three types of eruption. The more pathognomonic eruption is curious, with mildly inflammatory papules suggesting folliculitis but usually without much pustulation. The eruption occurs in large numbers over wide body areas, including the face. The second and less common type is clinically indistinguishable from severe, extensive atopic dermatitis, although eczema can be localized and associated with excoriated papules. At least three patients with incontinentia pigmenti have also had the syndrome.[7]

3 *Candida*. It is uncertain whether the high prevalence of *Candida* infections of the mouth and nails is causally related to the syndrome or a complication of the long-term and frequent use of antimicrobials in these patients, but the dysregulated immune system may be a predisposing factor.

4 Serum IgE levels. These polyclonal elevations can be enormous (mean level about 20,000 IU/mL[5]). I personally believe that high serum levels of IgE to staphylococci[8] are relevant and causal, inducing a very perturbed host defense to staphylococcal infections once infection starts. Patients with high levels of IgE may have falsely positive radioallergosorbent tests (RASTs) due to nonspecific binding of IgE to the solid phase, but dilutional studies can resolve this.[8]

5 Eosinophilia. Blood eosinophil counts are high and sometimes grossly so. Biopsy of at least the papular type of eruption may disclose numerous tissue eosinophils as well.

6 Bony changes. Patients commonly have a "coarse face" with frontal bossing, hypertelorism, and wide alar bases. Soft tissue changes from facial infections also give patients a "coarse face." These changes are far more common in patients with the papular eruption variant.

7 Impaired neutrophil chemotaxis. I have spent considerable laboratory time working on this subject and believe that the impairment is episodic, associated with infections, and may be a result rather than a cause of the infections.

8 Urticaria. This is common and often occurs together with infections; patients may appear with long lists of allergies to antimicrobials. In at least some of these patients, the urticaria may be caused by the infection rather than the drugs used to treat it.

9 Familial tendency. Onset in childhood is the norm. This syndrome may affect an adult and one of more of their children, suggesting autosomal dominant inheritance with variable penetrance.

Perhaps the immune dysregulation of atopic dermatitis, incontinentia pigmenti, or other disorders can occasionally lead to an acquired syndrome with features similar to a syndrome of congenital origin (but atopic dermatitis probably has a genetic basis too). Identification of relevant genes and pathogenic factors should help to resolve this. The two patients reported by Shermer *et al.*[1] certainly had high IgE levels and eosinophilia, but lacked many typical features of the hyperimmunoglobulin E syndrome with chronic infection, such as infections in organs other than the skin.

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Letter to editor

Lichen sclerosus et atrophicus arising in urethrostomy scar

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Sir,

A 58-year-old man consulted our department in 1999 complaining of whitish lesions of the perineum, which had been present for 6 months.

Between 1994 and 1997 he had several episodes of acute retention of urine, requiring multiple urethral catheterizations. Urethrography showed posterior urethra stenosis and a rosary of multiple urethral stenoses, but no signs of malignancy. Finally, in February 1997, in view of the recurrence of the urethral structures, a perineal urethrostomy was performed.

On dermatologic examination there were several whitish plaques. These were porcelain-like with a keratotic surface and were situated on both sides of the hypospadic urethral orifice, with purpural erythema of the stomal mucosa (Fig. 1). The glans showed whitish coloring around the urethral meatus with thickening and contraction of the prepuce and secondary phimosis. The patient had no extra genital lesions.

Histopathologic study of the perineal area showed an atrophic epidermis, with compact orthokeratosis and homogenization of the collagen of the papillary dermis, and telangiectatic vessels.

The patient was diagnosed as having lichen sclerosus et atrophicus and treated with topical steroids. Two years later the patient is clinically stable.

Discussion

Lichen sclerosus et atrophicus (LSA) is a chronic inflammatory dermatosis affecting both men and women. It usually presents with involvement of the genitalia.

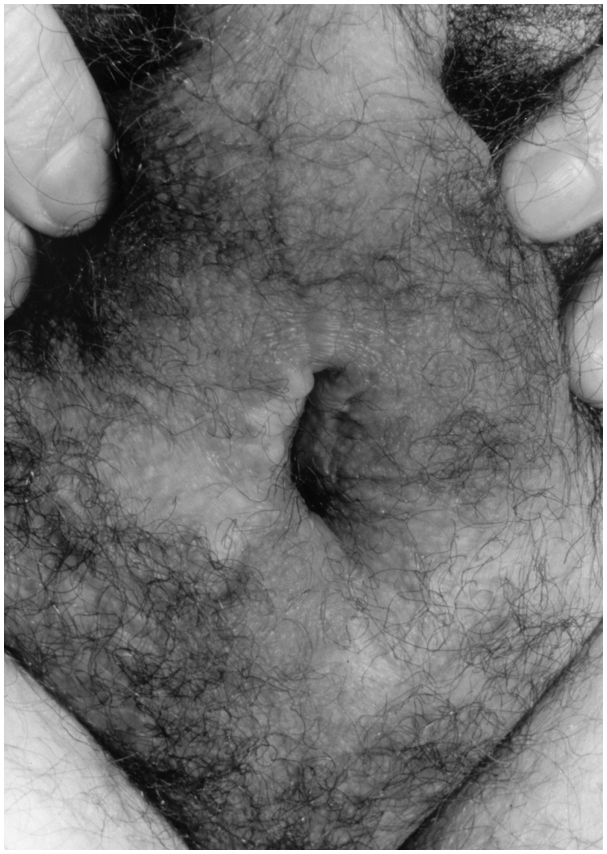

Figure 1 Porcelain-like plaques situated on both sides of the hypospadic urethral orifice

In men the glans and prepuce are most often affected, forming a ring at the free end of the prepuce, known as balanitis xerotica obliterans.[1,2] Although early stages of LSA may be asymptomatic, main symptoms include sudden phimosis, adherence to the glans, painful erection, and urethritis. If the constrictive ring adheres to the glans, it may cause stenosis of the urethral meatus and dysuria, urinary obstruction, and urethrorrhagia;[1] with possible damage to the posterior urethra and even to the urinary bladder and kidneys.[2]

Diagnosis of LSA is clinico-pathologic. The presence of a sclerotic white ring at the tip of the prepuce is diagnostic[2] and the histopathologic changes are typical.[3]

Among the factors related to the origin of LSA,[3–5] Koebner's phenomenon may be suggested as a form of development of the disorder in which post-traumatic clinical situations may play a role, such as circumcision, sunburn, radiotherapy, correction of congenital anomalies, surgery for carcinoma of the penis, and cholecystectomy or skin grafts We found only one reference in the literature of LSA developed in urethrostomy scars.[6]

However, the appearance of LSA in regions of old scar tissue should not invariably be interpreted as resulting from the Koebner phenomenon because, as has been well recorded in the literature,[7] there may be histologic findings of LSA with particular characteristics in the absence of clinical signs of LSA occurring away from scar tissue. In such cases, the correct denomination should be "LSA-like change occurring in association with a scar", and thus should be differentiated from true LSA.[7]

In our case, the clinical and histopathologic diagnoses of LSA were made on the cutaneous lesions found around the urethrostomy. However, the cutaneous lesions on the penis and the urethrografic findings, in the absence of any other justifying cause, suggested that it might be a genital LSA that had been present for a long time. This supported the retrospective diagnosis of balanitis xerotica obliterans.

With regard to peristomal cutaneous lesions, we found descriptions of skin disorders such as irritant reactions, particularly from leakage of urine or feces, allergic contact dermatitis, infections, and pre-existing skin diseases, principally psoriasis, seborrhoeic dermatitis, eczema and pyoderma gangrenosum.[8,9] Lichen sclerosus et atrophicus is not a dermatosis that usually develops around a stoma according to the articles that we have consulted. We have found only two reports describing LSA affecting colostomy sites in two patients with vulvar LSA,[4] and only one case in a zone of urethroplasties from multiple skin grafts in a patient with genital LSA.[6]

In conclusion, we consider our patient as a case of genital LSA, developing into the Koebner phenomenon in the periurethrostomy scar. We emphasize that these lesions are only exceptionally found around a stoma.

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