

# Letters to the Editor

thereby compromised (10). Accordingly, he developed a sudden shock from septicemia and died without exhibiting conventional signs such as soft tissue necrosis or necrotizing fasciitis. Septicemia without fever is not rare, as illustrated by this case. Some reports state that fever was not observed in 30% of STSS cases (6).

Septicemia tends to occur in patients with underlying diseases such as hypertension and diabetes mellitus (6). The possibility of STSS should be considered in patients with severe pain in the extremities which is suggestive of necrosis, even in the absence of skin color changes. Therefore, it is necessary to have a high index of suspicion for an infectious etiology and to begin treatment immediately before the diagnosis of disease is confirmed.

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## Spondyloarthropathy and Turner's syndrome

Sirs,

Chromosomal abnormalities have lately been shown to lead to a predisposition for autoimmune diseases. Case reports exist in the literature demonstrating the association of Turner's syndrome (TS) with polyarticular or oligoarticular type of juvenile chronic arthritis (JCA) (1, 2). We present an unusual case of a TS patient who suffered from buttock and low back pain and oligoarticular arthritis, concluding with a delayed diagnosis of arthritis resembling ankylosing spondylitis.

A 30-year-old female presented to our clinic suffering from occasional pain and swelling in both knees and the right ankle, as well as pain in the buttock area since her childhood. Her back pain had an insidious onset and showed progression over the years with no response to simple analgesics anymore. Examination revealed an atypical facial appearance, growth retardation, the absence of secondary sex characteristics along with primary amenorrhoea. She had a short stature, short webbed neck with a low hairline, wide chest with atrophic nipples and juvenile female external genitalia. Neck rotation was limited. She had painful and limited trunk flexion with forward bending mainly performed via hip flexion. Sacroiliac provocation tests were painless but limitation of bilateral hip rotation, shortened hamstring muscles and pain in the right knee on performing passive range of motion were observed. No neurological deficit was detected. Radiological investigation revealed bilateral grade III sacroiliitis and signs of inflammation in both hip joints (Fig. 1), as well as generalized osteopenia.

Routine haemanalysis indicated a normal blood count, slightly elevated liver function tests and high alkaline phosphatase levels. Serologic investigation was negative for C reactive protein, rheumatoid factor, ANA and markers for Hepatitis B, Brucella and Salmonella infections. The erythrocyte sedimentation rate was 34 mm/h. HLA typing was negative for HLA B27. She had low

serum estradiol and elevated FSH and LH levels and normal thyroid function tests. Pelvic ultrasonography demonstrated a hypoplastic uterus and fibrotic gonads. Caryotype analysis was performed and a 45 XO configuration revealed Turner's syndrome. Bone mineral density measurement by the DEXA technique revealed severe osteoporosis of the lumbar vertebrae ( $t = -5.58$  SD). A diagnosis of spondyloarthropathy co-existing with TS was reached and anti-inflammatory medication along with vitamin D and calcium supplementation was started, as well as advising a home exercise program to the patient.

The relationship of TS with arthritis had only been demonstrated through case reports in the literature until Zulian *et al.* (3) published a large series of patients with JCA associated with TS. The prevalence of the two conditions together was shown to be at least 6 times greater than would be expected if they were only randomly associated. Two different patterns of arthritis were present in their series and also in the literature; polyarticular and oligoarticular. Our case is of interest due to the form of presentation of the arthritis. To our knowledge, this is the first description of TS-related arthropathy involving the axial joints along with the peripheral joints.

It has been suggested that chromosomal breakage and rearrangement can induce the specific immunological alterations which are responsible for the onset of arthritis and other autoimmune conditions seen in patients with TS (3). Another possible link



**Fig. 1.** Anteroposterior lumbosacral radiogram demonstrating bilateral Grade III sacroiliitis, hip involvement and generalized osteopenia.

between TS and arthritis may be tied to the serum hormone levels in these patients. Recent studies have shown the effects of sex steroids and neurohypophyseal hormones on immunity and autoimmunity (4). Radiographic osteopenia is a frequent finding in TS (5). It is suggested that this is not an intrinsic feature of TS but results from extreme oestrogen deprivation (6). Our patient had a chronic inflammatory disease affecting the musculoskeletal system which in the end may possibly have exerted an additional effect on the development of severe osteoporosis.

In TS, the diagnosis of arthritis is usually delayed because the joint symptoms are assumed to be related to the syndrome itself and not to an inflammatory arthritis (1). This case highlights the relationship between TS and different rheumatological conditions. Clinicians should therefore be aware of the possibility of an inflammatory process in their patients with Turner's syndrome in order to make an early diagnosis. The possibility of sacroiliitis and spondylitis should also be kept in mind in the follow-up of TS patients.

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## Femoral head osteonecrosis after topical corticosteroid therapy

Sirs,

Femoral head osteonecrosis as an adverse effect of topical corticosteroid therapy has been reported in two cases in the medical literature (1, 2). We encountered recently the case of a 43-year-old man who presented with a 2 month history of left hip pain. He complained of a dull, 'toothache-like' pain in the front of the left hip and thigh. The pain had been gradual in onset and, although at first helped by rest, became almost constant causing an antalgic limp. His medical history showed that he had for the last 10 years been followed for diffuse psoriasis in a dermatology department. He was treated first with methotrexate and topical betamethasone dipropionate. Eight years ago he interrupted his visits to the dermatologists, discontinued methotrexate and used to apply topical betamethasone dipropionate twice daily all over his body. He had no history of trauma or alcohol intake.

Physical examination showed a severe and diffuse psoriasis. Rheumatological examination revealed that he walked with a slight left-sided antalgic gait. He had a discrete limitation in range of motion of internal rotation of the left hip. Laboratory tests showed ESR 10 mm/1st hr, and a white cell count and haemoglobin in the normal range. Serum levels of cholesterol, triglycerides, glucose, and uric acid were in the normal range. Rheumatoid factor was negative as were antinuclear antibodies. Pelvic x-rays and CT scan showed evident osteonecrosis of the left femoral head (Fig. 1). The sacroiliac joints were normal. Treatment consisted first of analgesics and the suppression of weight bearing. One month later, a core decompression of the left hip was carried out.



Fig. 1. CT scan of the left hip showing evident osteonecrosis of the femoral head.

Systemic side effects of topical corticosteroids are rare, but may occur especially in children and elderly patients (3). Possible adverse systemic effects are related directly to such factors as the application site, duration of application, potency, and occlusion of the medication. The application of high-potency corticosteroids should be limited, when possible, to a twice-a-day basis for 3 to 4 weeks. Physicians must also be aware of increased transcutaneous penetration in certain areas of the body with thin epidermis, such as the eyelids, periorbital area, axillas, crural region and genitalia. A patient with an integumentary perturbation, such as an exfoliating dermatitis (e.g. diffuse psoriasis as in our case), has a much greater amount of percutaneous penetration (4,5). Long and frequent application carries the risk of suppressing the hypothalamic-pituitary-adrenal axis and even producing Cushing's syndrome, especially in very young patients (6).

In our patient, the only predisposing factor for femoral head osteonecrosis was topical betamethasone dipropionate, which had been used for about 10 years on a daily basis and applied over a large part of the body surface because of a severe psoriasis.

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