Picture Story

Idiopathic atrophoderma of Pasini and Pierini in a young boy

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Introduction

Idiopathic atrophoderma of Pasini and Pierini (IAPP) is a rare dermatologic condition of unknown aetiology which is characterized by the presence of dermal atrophy[1]. The condition was initially called `progressive idiopathic atrophoderma' by Pasini who first described it in 1923[2]. The condition was further described by Pierini and Vivoli in 1936[2]. We present a 12-year-old boy with the classical features of IAPP.

Case report

A 12-year-old boy presented with two hyperpigmented lesions on his back. The lesions had started as small discoloured areas 2 years ago and gradually enlarged in size. Except for the colour change of the skin, he did not have any discomfort or pain. On examination, he had two lesions, one circular in the mid-spine area about 7cm in diameter and another oval in the lumbar area 9 x 5cm. The lesions were non-tender, uniformly hyperpigmented, with a slight depression of the skin with an abrupt edge and normal sensation was present (Figure 1).

He did not have arthritis or uveitis. The haemoglobin level was 13g/dL (normal range 13-17g/dL). The total white cell count was 8,700/ cu mm (normal range 4,000-11,000 /cu mm) with 69% neutrophils (normal range 45-70%) and 25% lymphocytes (normal range 25-40%). Platelet count was 252,000/cu mm (normal range 200,000 to 490,000 /cu mm) and the erythrocyte sedimentation rate was 10mm in the first hour (normal range 4-10mm in the first hour). Antinuclear antibody test was negative.

Based on the characteristic nature of the skin lesions, he was diagnosed to have atrophoderma of Pasini and Pierini and was treated with topical imiquimod 5% cream twice a day, oral hydroxychloroquine 200 mg once a day and oral methotrexate 15 mg once a week for a year. There was no change in his skin lesions after 1 year. There were no new lesions.

Discussion

Lesions of IAPP are typically circular or oval hyperpigmented areas that are slightly depressed below the surrounding skin, described as `cliff drop border'[3].They are usually located on the backs of adolescents or young adults[3]. IAPP is more common in the Caucasian race and in females[4]. Some reports in the past have linked IAPP to *Borrelia burgdorferi* infection and raised antibody titres were noted in some patients[5]. On the other hand, IAPP is considered as a subtype of morphoea by some authors, as an atrophic non indurated variant of morphoea without the typical sclerosis[6,7,8]. Classical morphoea often starts as a localised patch of erythema, induration with erythematous halo or altered skin pigment[9].The lesions are characterized by skin thickening and excessive accumulation of collagen and may involve atrophy of subcutaneous or deeper tissue[10]. Histopathology in early morphoea may reveal perivascular infiltration with mononuclear cells and in advanced stages the entire dermis may be replaced by collagen[10]. Distinct features of IAPP compared to morphoea were identified by Canizares O, *et al*[2] in 1958 and the term 'Idiopathic Atrophoderma of Pasini & Pierini' was coined. The diagnosis is usually based on the history and characteristic clinical appearance of the lesions, as histopathological changes are minimal and non diagnostic[1]. When biopsy is performed, changes may include reduced thickness of the affected dermis, with atrophy and hyalinization in the collagen[1]. No treatment has been found to be uniformly effective[1]. Differential diagnoses include post-inflammatory pigmentation, lichen sclerosis and anetoderma[4].

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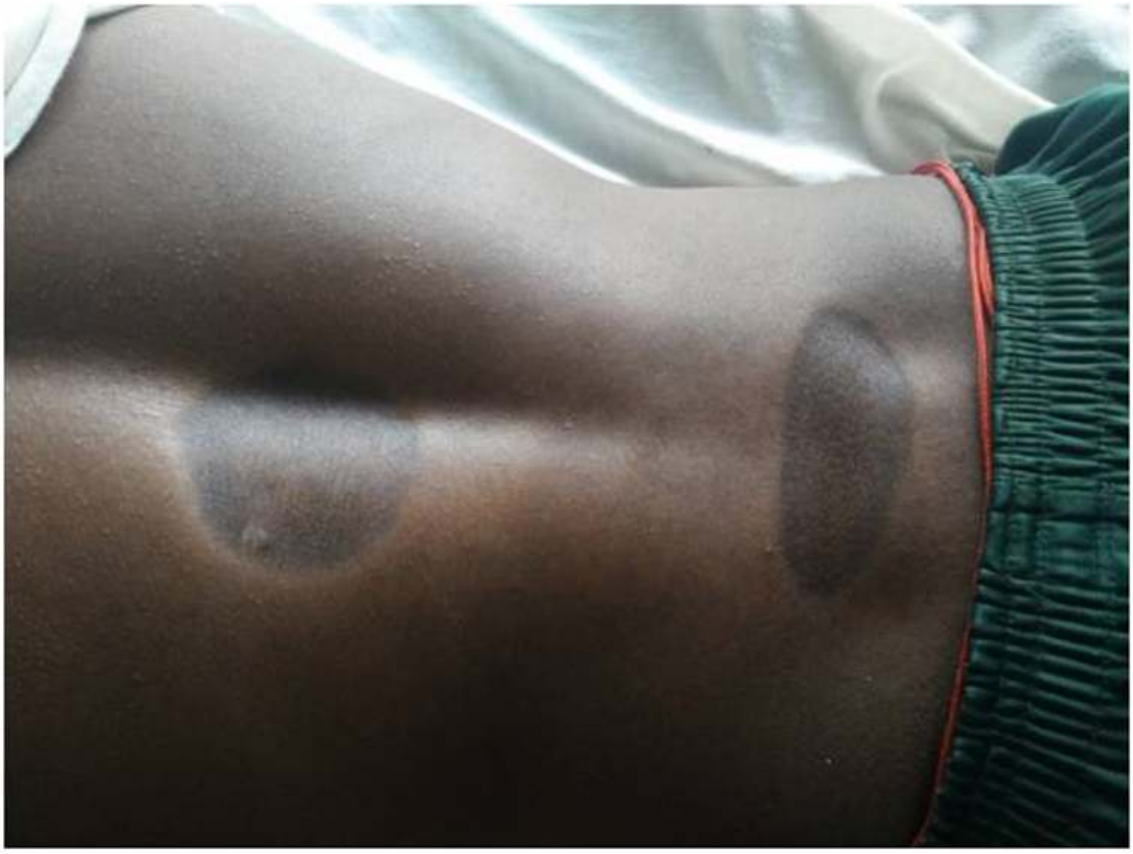

Figure 1: ***Hyperpigmented lesions on back with slight depression of skin and abrupt edges***

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