Connective Tissue Lipoatrophic Panniculitis

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Abstract
A 23 years old girl with a past history of sero-positive oligoarticular juvenile idiopathic arthritis developed nodules and depressed scars over face and limbs. Biopsy from the lesion revealed mixed panniculitis. She responded to steroids and hydroxychloroquine but has residual lipoatrophic scars needing reconstructive surgery.

INTRODUCTION

Lipoatrophic panniculitis is a rare disorder. The one described in association with autoimmune disease, has been labelled as connective tissue panniculitis. It often leaves residual scars that require surgical reconstruction. We discuss here such a case juvenile oligoarticular idiopathic arthritis.

CASE REPORT

MT a 23 year old female, presented to us with two years history of multiple painful swellings all over the body. These were not associated with any constitutional symptoms and would heal leaving depressed scars. They would recur every 4-6 weeks, each episode lasting 2-3 weeks. She had a past history of oligoarticular arthritis involving right middle finger (PIP) and right elbow with a positive rheumatoid factor since August 1992. She was diagnosed as juvenile rheumatoid arthritis (now called juvenile idiopathic arthritis) and was treated with anti-inflammatory drugs and steroids intermittently for 2 years. Since then she has been having off and on pain in the fingers, elbows and feet without any noticeable swelling.

General physical examination revealed multiple large erythematous nontender nodules over the forearms. There were few depressed scars over the upper arms and both the cheeks (Fig. 1). There were fading tender nodules over the legs. The joints were normal and systemic examination including eyes was normal. Investigations showed Hb of 11 gm/dl, TLC 3700/mm³, P55%, L 35%, platelets 1.9 lac/mm³. ESR 25 mm in 1st hr, rheumatoid factor positive (128 IU/L) and CRP positive at 12 mg/L. ANA was negative, serum calcium 9.0 mg/dl, blood sugar (fasting) was 84 mg/dl, thyroid function tests were normal. X-ray chest and X-ray of both hands were normal. Mantoux test was negative. Biopsy taken from the right leg nodule showed fibroadipose tissue with chronic inflammatory reaction involving the septae and fat lobules with focal areas of fat necrosis (Fig. 2). The inflammatory cells consisted of lymphocytes, many foamy histiocytes, few plasma cells and occasional neutrophils, a picture suggestive of mixed panniculitis. A diagnosis of lipoatrophic panniculitis was made.

The patient was put on 0.5 mg/kg/day of prednisolone for 8 weeks which was then tapered off over the next 16 weeks, hydroxychloroquine 200 mg BD was also started and continued for six months after a baseline eye examination. She improved clinically with disappearance of the nodules.

DISCUSSION

Lipoatrophic panniculitis includes all types of panniculitis in which atrophy is the predominant feature. Three described
cases of panniculitis are associated with diabetes mellitus. Hashimoto’s thyroiditis and juvenile rheumatoid arthritis have suggested an autoimmune aetiology\(^1\) and possibly auto-antibodies to fat cells.\(^2\) It can be primary or secondary to erythema nodosum or erythema induratum. Primary lipoatrophic panniculitis occurs in both adults and in children either as an acute benign condition or as a chronic recurrent disease.\(^1\) Mixed panniculitis with fat atrophy, suggestive clinical picture and positive immunological markers indicate a diagnosis of connective tissue disease-related panniculitis;\(^3;4\) the latter can be thought of as a subtype of a broader group of lipoatrophic panniculitis.\(^4\) This is a rare condition described for the first time in 1980 by Peters and Winkelmann, what was then called ‘atrophic connective tissue disease panniculitis’. Six cases were described, three each were adults and children and all were females. All the adults had inflammation of fat of trunk and limbs leading to lipoatrophy. Two of the patients were ANA positive and one was SS-B antibody positive. Histopathology of the lesions showed lymphocytic panniculitis. These patients responded to anti-malarial therapy. The children had more localised disease and were ANA-negative.\(^3\) Although our patient was ANA-negative, the association with a positive rheumatoid factor suggests that she had a connective tissue disease related panniculitis.

In adults, the response to therapy may be unsatisfactory due to the severity and chronicity of the disease. Corticosteroids in high dosages have been reported to be effective. Reconstructive surgery is often needed. This can be done with the patient’s own soft tissue or with an alloplastic material such as silicon.\(^5\)

Our patient did respond to a combination of steroids and anti-malarials. Re-constrictive surgery to improve the lipoatrophy over the face and the right upper arm is being contemplated.

**References**