

CASE REPORT

Bullous Erythema Nodosum Leprosum masquerading as systemic onset juvenile idiopathic arthritis: A case report

SWETALINA PRADHAN*, BIBHU PRASAD NAYAK*,
TANMAY PADHI* & MADHUSMITA SETHY*

* *Veer Surendra Sai Medical College, Burla, Sambalpur,
Odisha, India*

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Introduction

ENL or Type 2 lepra reaction is commonly seen in patients of lepromatous leprosy with high bacillary index and usually starts after institution of multi drug therapy.¹ Although it commonly presents with tender nodular lesions, various forms like vesicular, pustular, necrotic or bullous have been reported in adults.

Case report

A 10 year old male child presented with a history of fever, swollen and tender bilateral elbow and interphalangeal joints for 2 days (Figure 1a and b) and intermittent fever and joint pain for the past 2 months.

General and systemic physical examination revealed pallor and cervical and epitrochlear lymphadenopathy, mild hepatosplenomegaly and diffuse erythema over his face (Figure 1b).

The relevant laboratory findings were Hb- 8mg%, TLC-15,000/mm,³ TPC-3-6 lacs/mm,³ ESR-50mm in the 1st hour, positive C-reactive protein and IgM Rheumatoid factor, normocytic normochromic anemia without any atypical cells, normal chest X-ray and negative MP-QBC. On the basis of fever, joint swelling and laboratory markers the patient was provisionally diagnosed with mild grade systemic onset juvenile idiopathic arthritis (SOJIA) and was started with naproxen. Subsequently, a few sterile bullous lesions appeared over his bilateral elbow joints and forearms (Figure 1a and b). Dermatological examination of the patient revealed a few nodules near the elbow joint and sensory loss over his bilateral hand without any hypopigmented patch. The sister of the patient had multiple hypopigmented

Correspondence to: Swetalina Pradhan, Veer Surendra Sai Medical College, Burla, Sambalpur, Odisha, India
(e-mail: dr.swetalinapradhan@gmail.com)

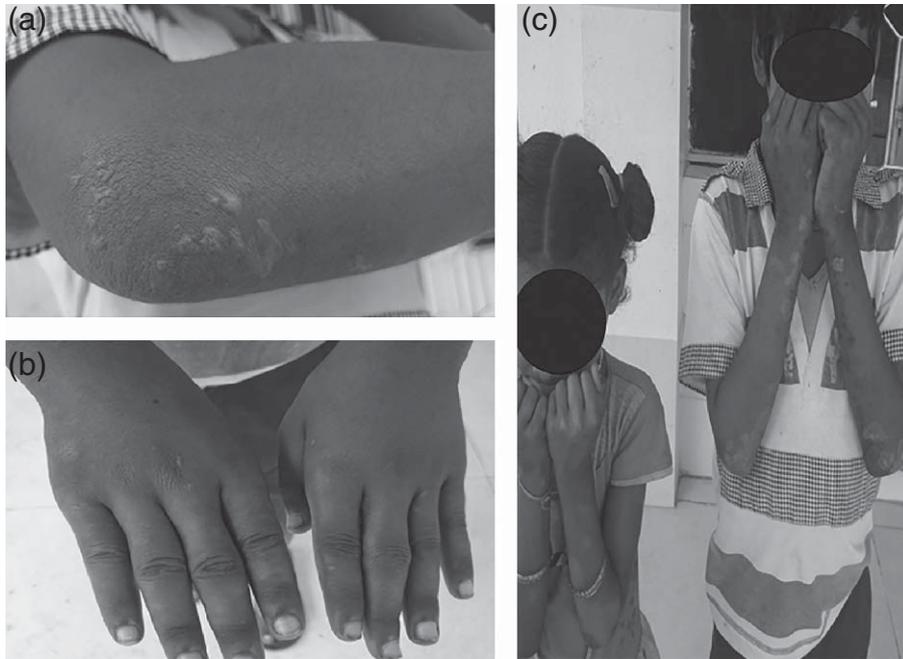


Figure 1.

- a) Erythematous, swollen elbow joint with bullous lesions over it
- b) Swollen interphalangeal joints of bilateral hand
- c) Multiple hypopigmented patches over bilateral hands in the girl child and healed bullous ENL lesions over the bilateral hands in male child

patches all over her body, along with loss of sensation (Figure 1c). The slit skin smear was positive and histopathology of the skin from lesions showed ulcerated stratified squamous epithelium, intraepidermal bulla, foamy macrophages with a few giant cells in the dermis, vasculitis with perivascular neutrophilic and lymphocytic infiltration along with dense infiltration of neutrophils in the dermis and subcutaneous fat (septal panniculitis) consistent with features of erythema nodosum leprosum (Figure 2).

A diagnosis of nodular lepromatous leprosy with bullous ENL was made and oral prednisolone 1mg/Kg/day and WHO MDT MB (Child) was started. The bullous lesions and fever responded dramatically to treatment (Figure 1c).

Discussion

ENL is an immune complex reaction which manifests as crops of numerous evanescent, erythematous, tender nodules and plaques over the extremities, trunk, face and other parts of body. It has various morphological patterns - nodular, vesicular, pustular, bullous and necrotic - with the nodular pattern being the most common.^{2,3} Due to immune complex mediated damage there can also be lymphadenitis, neuritis, iridocyclitis, arthritis, synovitis, myositis, epididymo-orchitis, glomerulonephritis and involvement of the liver and spleen leading to hepatosplenomegaly.^{4,5} It can present with only joint involvement for which it may be confused with systemic conditions like systemic onset juvenile idiopathic arthritis

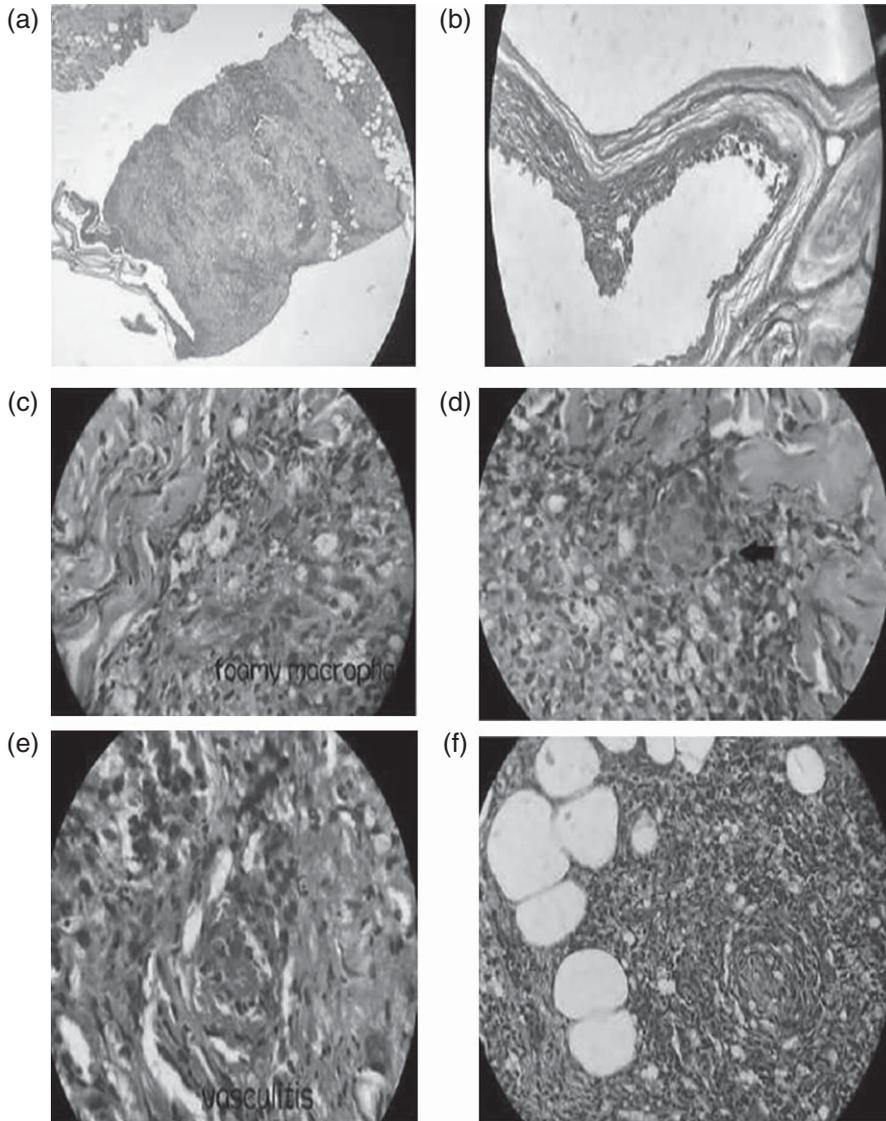


Figure 2.

- a) Scanner view showing Intraepidermal bullae and dense neutrophilic infiltration in dermis and subcutis
- b) 40X view showing intraepidermal bulla
- c) 40X view showing foamy macrophages in dermis
- d) 40X view showing giant cells in dermis
- e) 40X view showing vasculitis
- f) 40X view showing septal panniculitis

(SOJIA), rheumatic fever in the pediatric population and also IgM RA factor may come positive in a few cases.

In our case, the patient presented with fever, swollen tender joints, lymphadenopathy, hepatosplenomegaly and was provisionally diagnosed as SOJIA but after the appearance of a

cutaneous lesion and detailed sensory and peripheral nerve examination and investigations like slit skin smear and histopathology, the diagnosis changed to LL with bullous ENL. There was a family history of contact suggesting ongoing transmission among family members. Besides the diagnosis of SOIJA, in bullous ENL other bullous disorders like pemphigus vulgaris, bullous drug eruption or bullous pemphigoid should be ruled out when there is involvement of oral mucosa and acantholytic cells are found on a Tzanck test.

In case of children presenting with an atypical form of ENL like multiple bullous lesions, without any anaesthetic patch and history of intake of MDT, ENL can masquerade as systemic diseases like SOIJA, and in all such cases detailed sensory and peripheral nerve examination should be done initially followed by specific investigations and family history of contact should be sought in all such cases. The case is interesting both for its rare variant of ENL and presentation in children.

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