Papuloerythroderma Secondary to Granulocytic Sarcoma of Lymph Node

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Abstract

Papuloerythroderma (PE) is a rare cutaneous condition characterized by intensely pruritic, papular eruptions sparing major skin folds. These lesions later coalesce to form plaques and potentially evolve into erythroderma. The characteristic sparing of large skin folds in PE is known as the deck chair sign. Various associations for PE have been identified. We are here reporting a case of PE secondary to granulocytic sarcoma of lymph node. To the best of our knowledge, PE associated with granulocytic sarcoma has not been reported before.

Keywords: Deck chair sign, granulocytic sarcoma, papuloerythroderma

Introduction

Papuloerythroderma (PE) has been reported as early as 1979 when two cases of diffuse erythroderma presenting with lichenoid papular eruption were reported.[1] The term PE was, however, used for the first time in 1984 by Ofuji, who described the term papuloerythroderma for a condition characterized by generalized pruritic lichenoid lesions with relative sparing of major skin folds (also known as deck chair sign).[2,3] A meta-analysis of 170 cases was done by Torchia et al., in 2010, and on the basis of their observations, they classified PE as primary PE, secondary PE, PE like T-cell lymphoma, and pseudo-PE. Here, we are reporting a case of papuloerythroderma secondary to underlying granulocytic sarcoma of lymph nodes.

Case Report

A 45-year-old male patient came to the dermatology outpatient department with multiple, pruritic, raised reddish lesions involving face, upper limb, trunk, and lower limbs along with swelling over the submental and lateral side of the neck since the past 3 months [Figures 1-3]. He also complained of difficulty in deglutition. There was history of recurrent low-grade fever. He had significant weight loss over a period of the past 2 months. The patient had a negative history of hematemesis, melena, vomiting, abdominal pain, cough, hemoptysis, any seasonal variation, or any drug intake. Family and personal history were not significant.

Cutaneous examination revealed multiple erythematous confluent papules with fine scaling, characteristically sparing skin folds of neck and abdomen, known as deck chair sign [Figure 1]. Cervical, axillary, and inguinal lymph nodes were bilaterally enlarged, firm, nontender, and mobile. Systemic examination showed hepatosplenomegaly. Routine hematological investigation had eosinophilia (40%), and serum immunoglobulin E (IgE) level of the patient was increased (>2000 IU/ml). Rest biochemical parameters were within normal limits.

Ultrasonography of the abdomen showed enlarged liver (16.9 cm), enlarged spleen (15 cm), and multiple enlarged lymph nodes in para-aortic, peripancreatic and bilateral inguinal regions. Ultrasonography of the neck showed multiple enlarged lymph nodes in level IA, B/L level IB, II, III, IV, V and bilateral three to four lymph nodes in supraclavicular region. A punch biopsy was sent from the most characteristic cutaneous lesion and the histopathological examination

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showed extensive collagenization in dermis along with eosinophils and occasional premature granulocytic infiltration surrounding blood vessels [Figures 4-6].

Biopsy of the cervical lymph node was suggestive of infiltrate with atypical large cells. Immunohistochemistry of the same showed CD43; myeloperoxidase; leukocyte common antigen positivity in all cells [Figure 7a-c], CD68, KP1; CD68 PGM 1; CD117 positivity in some cells and CD20, CD2, CD5 negativity which confirmed the diagnosis of granulocytic sarcoma.

Bone marrow biopsy showed normocellular marrow with increased eosinophil count with normal cytogenetic test.
On the basis of characteristic clinical findings and diagnostic criteria described by Torchia et al., the diagnosis of PE secondary to granulocytic sarcoma was made. The patient was referred to Oncology Department. Further investigation in the form BCR-ABL gene and FLT3 gene mutation, which are for prognosis of sarcoma, came negative. The patient was referred to onco-physician where he was given three cycles of intravenous doxorubicin and cytosine arabinoside. The patient improved dramatically after first induction phase only having postinflammatory hyperpigmentation [Figure 8].

**Discussion**

Four cases of PE were first described by Professor Shigeo Ofuji, Kyoto University, Japan, in 1984. An annual incidence of 1.5/million over 10 years’ observation was found by Bech-Thomsen and Thomsen, most of which were reported from Japan. PE is an intensely pruritic skin condition with characteristic clinical features. In 1986, Farthing et al.
described the typical sparing of the major body folds in PE and pressure areas of the skin lesions as “deck chair sign.” Its pathogenesis is still not known. It is believed that it could be a hypereosinophilic syndrome or paraneoplastic dermatoses. There has been an association of PE with malignancy in 21.76% of patients, atopic dermatitis in 9.23% of patients, and drug-induced hypersensitivity in <5% of patients.

A cutaneous biopsy is done to rule out any cutaneous malignancy. The most common malignancies associated with PE are T-cell lymphoma and gastric carcinoma. Torchia et al. described criteria for papuloerythroderma of ofuji (PEO) in 2009, which included necessary and minor criteria. These criteria are as follows:

**Necessary Criteria**
1. Erythroderma-like eruption with flat-topped papules, red-to-brown in color and with a cobblestone appearance
2. Sparing of the skin folds and creases (deck chair sign)
3. Itch
4. Histopathological exclusion of cutaneous T-cell lymphoma (CTCL) and other recognized skin diseases
5. Workup to exclude malignancy, infection, drugs and/or atopy as a causal factor.

**Minor criteria**
1. Age >55 years
2. Male gender
3. Peripheral and/or tissue eosinophilia
4. Peripheral lymphopenia
5. Increased serum IgE.

Depending on the fulfilling of the above criteria, PE is classified as:
- PEO or primary idiopathic PE (Criteria 1–5)
- Secondary PE (Criteria 1–4)
- PE-like CTCL (Criteria 1–3 and 5).

In our case, the patient had necessary criteria of erythroderma such as eruptions, deck chair sign, severe pruritus, and no evidence of CTCL on histopathology. Minor criteria which were fulfilled include gender, peripheral eosinophilia, and raised serum IgE level. Based on proposed criteria, the case was diagnosed as PE secondary to granulocytic sarcoma. There were eosinophils present in the dermis with occasional premature granulocytic cells in perivascular area which gave us a clue toward an underlying malignancy. The patient was referred to the oncology department for further management. The patient improved after starting therapy for granulocytic sarcoma.

**Conclusion**
Although the cause of PE remains obscured in majority of the patients and with its rare occurrence in Indians, a typical clinical presentation should clinch the diagnosis of PE and an active search for underlying malignancy should be made for better patient management.

**Declaration of patient consent**
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**
There are no conflicts of interest.

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