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Case Report Open Access

Erythema Gyratum Repens Secondary to Acute Myeloblastic Leukemia

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Abstract

Erythema gyratum repens is a rare dermatologic manifestation of systemic diseases mainly secondary to neoplastic conditions. We report a case of an 80 year-old patient who was admitted to the hospital for prolonged fever, general status alteration, and pruritic skin lesion. He was known to have lymphoma in the past and current myelodysplastic syndrome. Physical examination revealed multiple, annular, rapidly growing erythematous plaques over the right thigh. Bone marrow aspirate and biopsy revealed Acute Myeloblastic Leukemia (AML). Diagnosis of Erythema Gyratum Repens (EGR) was made on clinical ground and it was secondary to AML after excluding others possible diagnosis.

Keywords: Erythema gyratum repens; Acute myeloblastic leukemia; Paraneoplastic skin rash

Introduction

Erythema gyratum repens (EGR) was first described in 1952 by Gammel in a woman with breast cancer [1]. EGR is a rare clinical entity; concentric erythematous bands with a wood grain appearance and scaling edge characterize the rash. The pathophysiology of erythema gyratum repens is unknown, although an immune response is postulated. The cutaneous eruption may precede, occur with, or follow the diagnosis of malignancy. In 80% of cases, EGR is related to underlying malignancies [2].

The diagnosis is based mainly on clinical ground and characteristic skin lesion. Although no specific microscopic features on skin biopsy, focal parakeratosis, mild hyperkeratosis, acanthosis, spongiosis and a superficial perivascular lymphocytic infiltrate might be seen. Biopsy is useful to rule out other possible mimicking annular diseases [3]. Treatment resides primarily by treating the underlying conditions. We report a case of EGR in a patient who presented with prolonged fever, pruritic skin rash secondary to acute myeloblastic leukemia.

Case Presentation

An 80 year-old Caucasian man was admitted to the hospital for prolonged fever, weight loss, and general status alteration. He noted a pruritic skin lesion, around 2 cm in diameter appeared two days before admission. As past medical history, he had been treated for Hodgkin lymphoma in 1994 and non-Hodgkin lymphoma in 2004. One year ago, he developed Myelodysplastic Syndrome. Past social history he is an ex-smoker. Allergies: No known drugs or food allergies. Home Medications: Paracetamol PRN. No recent travel. No history of contact with sick persons.

On physical examination, vital signs: HR: 96/min BP: 100/60 mmHg Temp: 38.8°C SpO_2: 97% RR: 18/min. The patient was conscious, cooperates, and oriented. Heart, Pulmonary, and abdominal exams were unremarkable. No palpable lymph nodes (Cervical, axillary, Supraclavicular, inguinal, and femoral). Musculoskeletal exam: normal. Cutaneous examination on admission and after 48 h of hospitalization revealed multiple, annular, rapidly growing erythematous plaques over the right thigh (Figures 1 and 2).

Laboratory exam showed WBC: 18000 x 10^9/L, ESR 65 mm/h, CRP 40 mg/L, Hemoglobin: 10 g/dl Hematocrit 30%, Platelets: 65000 x 10^9/L. Coagulation studies: Normal. BUN/Creatinine/Liver Function tests: normal. Multiple Blood cultures and urine culture were sterile. Viral and Bacterial Serology (HIV, HBV, HCV, HSV1-2, Lyme disease): Negative. Autoimmune antibodies panel: Negative. Quantiferon and PPD skin test: Negative. We did a thoraco-abdominopelvic CT scan, gastroscopy and colonoscopy were normal. Transthoracic echocardiogram revealed no valvular vegetations or arguments in favor of endocarditis.

We did a work up for bicytopenia (Anemia and thrombocytopenia). Peripheral blood smear showed 40% of circulating Blast cells. Bone marrow aspiration and biopsy performed which confirmed the diagnosis of acute myeloblastic leukemia (AML) with maturation, M2 Subtype. Based on the clinical ground and the characteristic aspect of rapidly migrating concentric skin bands, we made the diagnosis of Erythema Gyratum Repens (EGR). After excluding solid organs malignancies, infections, and autoimmune diseases, we concluded that EGR was secondary to AML. Treatment by chemotherapy for AML was considered.

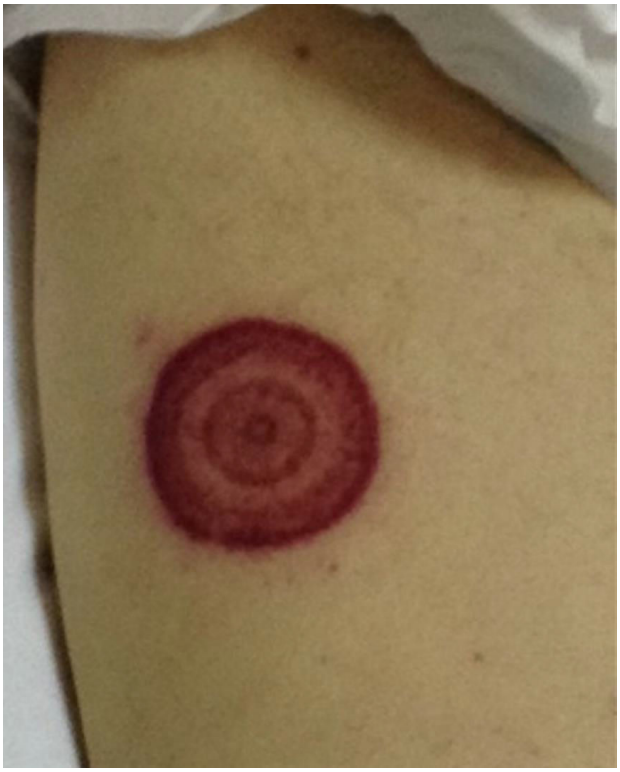

Figure 1: Well-demarcated concentric bands skin lesion on the admission.

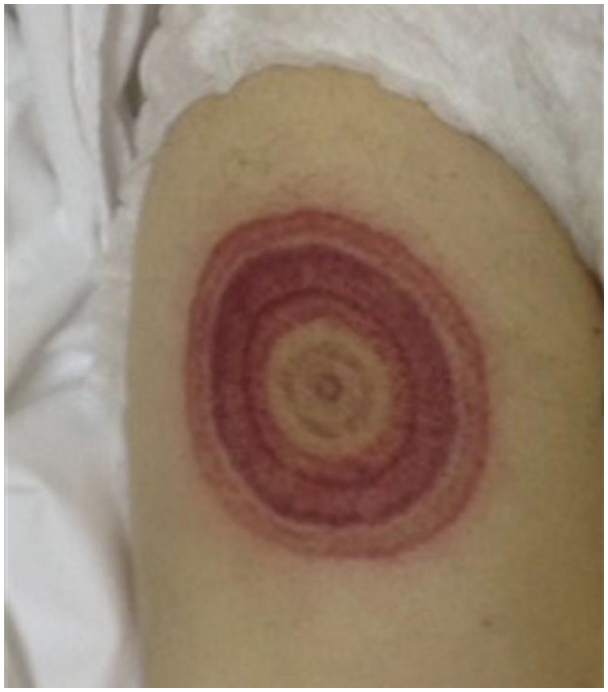

Figure 2: Rapidly growing serpiginous skin lesion on second day of hospitalization.

Discussion

Erythema Gyratum Repens typically is described as rapidly evolving, serpiginous band of erythema lined by edge of scales [1]. It affects the skin in a centrifugal pattern, mainly the trunk and extremities sparing hands, feet and face. Sometimes, rash is rapidly growing by 1 cm per day along with concentric aspect and pruritus may be a prominent symptom. It has been mainly reported in whites, with a 2:1 male to female ratio with an average age of onset of 63 years [2]. EGR in 80% of cases is related to underlying malignancy most notably lung cancer [4,5]. However, EGR in 20% is associated with non-neoplastic diseases such as pulmonary tuberculosis, autoimmune or connective tissue conditions [6]. Eubanks et al. [2] described that erythema gyratum repens in 82% of cases is secondary to internal malignancy and it is most commonly associated with bronchial, esophageal, and breast cancer. Rongioletti et al. did a systematic review of the literature and personal experience [3] regarding the underlying conditions that leads to EGR: they identified 83 cases of true EGR that were associated to malignancy in 70% of patients and 30% of cases were non-paraneoplastic in origin. Bronchial cancer was the most associated neoplasm. Interestingly, they reported one case of multiple myeloma and one case of lymphoma as underlying malignancies of EGR.

The diseases associated with non-paraneoplastic EGR were connective tissue diseases, systemic infections, hyereosinophilic syndrome and idiopathic causes. However, some diseases could mimic erythema gyratum repens such as: Neutrophilic dermatosis, bullous dermatoses, and systemic lupus erythematous gyratum repens [3]. In addition, there is some types of gyrate erythema could be misdiagnosed as EGR such as: erythema chronicum migrans, which is a slowly evolving lesion and it is related to infectious process. Erythema annulare centrifugum it evolves over days to week and it is mainly secondary to superficial fungal infection. Erythema marginatum rheumaticum it evolves rapidly over hours then disappears and it is related to rheumatic fever. The management of EGR consists in the treatment of underlying disorder [2]. Different dermatologic therapies such as topical steroids, vitamin A and immunosuppressive therapies like azathioprine have been used to treat EGR but have failed to relieve skin manifestations [5]. Systemic steroids are frequently ineffective and improvement often correlates with successful treatment of associated condition. This present case of erythema gyratum repens related to acute myeloblastic leukemia is the first case reported in the literature, despite that there are two cases described secondary to hematologic malignancies (multiple myeloma and lymphoma).

Conclusion

To the best of our knowledge, our patient's case is the first case of EGR secondary to Acute Myeloblastic Leukemia. Clinicians should be aware of the clinical aspect of EGR and maintain high index of suspicion, knowing that in major cases EGR is related to serious systemic pathology, usually malignancy. Although non-paraneoplastic EGR should be considered in the differential diagnosis, screening for

internal malignancy still remains a mandatory part of the diagnosis process.

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