

BRIEF REPORTS

Ear Cellulitis and Rash in a Patient With Rheumatoid Arthritis and Diabetes

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Pityriasis lichenoides et varioliformis acuta (PLEVA), also known as Mucha-Habermann disease, is well known to dermatopathologists but might be unfamiliar to many primary care physicians. This brief report describes a case of PLEVA subsequent to cellulitis of the ear pinna, offers a brief review of the disease, and illustrates the importance of the family physician continuing an aggressive workup of a mysterious skin disorder until a clear diagnosis is established.

Case Report

A 56-year-old woman with rheumatoid arthritis, type 2 diabetes mellitus, and hypertension came to the clinic complaining of a 5-day history of left ear redness and swelling. She denied pain, fever, headache, changes in her hearing, or trauma to the affected area. She had had type 2 diabetes mellitus diagnosed 6 months earlier, and rheumatoid arthritis, characterized by diffuse joint involvement, diagnosed 4 years earlier. She had been taking azathioprine for more than 1 year and methotrexate for the past 4 to 6 months. Her complete blood count and findings from a chemistry panel were normal 1 week before her visit.

Her current medications included the following: azathioprine 250 mg daily, methotrexate 7.5 mg weekly, lisinopril 20 mg daily, triamterene-hydrochlorothiazide 37.5/25 mg daily, atenolol 50 mg daily, glipizide 10 mg daily, and salsalate 750 mg twice daily. She had no known drug, food, or environmental allergies. The patient last took a short course of corticosteroids 10 months before this visit. She did not smoke, drink alcohol, or inject drugs. She had not recently traveled and reported no history of blood transfusion, hepatitis,

or suspected exposure to the human immunodeficiency virus. The patient was employed as a clerical worker.

The patient appeared nontoxic. Her temperature was 98.1°F, blood pressure 108/70 mmHg, pulse 72 beats per minute, and respiration 16/min. The left ear pinna was erythematous. The skin appeared slightly thickened and edematous. A tender, circumscribed area of erythema was present at the anterior, posterior, and inferior auricular areas. The ear canal and tympanic membrane appeared within normal limits. Cervical and preauricular adenopathy was noted on the left. The remainder of her directed physical examination was unremarkable except for joint findings consistent with rheumatoid arthritis.

It was believed that the patient had erysipelas. She was given 1 g of cefazolin intramuscularly, and oral cephalexin 500 mg every 6 hours was prescribed. A follow-up appointment was scheduled for 48 hours. The patient was advised to telephone or return immediately if any symptoms worsened or if fever developed.

At her follow-up appointment, the patient remained afebrile and without pain. There was a marked decrease in the erythema surrounding the ear. An area of dry, necrotic-appearing ulceration surrounded by several pin-point necrotic lesions had developed on the posterior pinna. Ciprofloxacin was added to the patient's drug regimen, and she was instructed on antiseptic wound care.

Five days later the patient returned for a follow-up evaluation. She reported full adherence to her prescribed medications. She continued to appear nontoxic and was afebrile. The left ear pinna continued to appear edematous and thickened. The erythema of the pinna and surrounding skin had nearly resolved. The necrotic area on the posterior pinna had enlarged. Crops of reddish-brown papules, some with a slight pustular appearance, had erupted on the face, trunk, and

Submitted, revised, 10 July 1998.

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upper extremities. There was mild itching.

A punch biopsy was obtained from a lesion on the trunk. A complete blood count showed a white blood cell count of 5200/ μ L with 75 percent neutrophils, 3 percent band cells, 10 percent lymphocytes, 5 percent monocytes, 3 percent eosinophils, and 2 percent basophils. Westergren sedimentation rate was 49 mm/h. Results of serum chemistries and urinalysis were within normal limits. Multiple sets of bacterial and fungal blood cultures were obtained (all eventually showed no growth). Tests for cryoglobulins were negative. Complement C3 was normal, and C4 was slightly decreased at 10 mg/dL.

The patient was admitted to the hospital, and intravenous clindamycin and tobramycin were prescribed. Her differential diagnosis included necrotizing cellulitis with disseminated infection, septic emboli, disseminated candidiasis, disseminated varicella zoster, vasculitis, panniculitis, relapsing polychondritis affecting the left ear pinna, and an atypical drug eruption. Antiviral medication was not given. Intravenous methylprednisolone was initially given but discontinued after two doses.

On the following day the patient's condition was essentially unchanged except that some trunk lesions appeared to be developing necrotic centers. She remained afebrile and nontoxic in appearance. The punch biopsy obtained on the day of admission showed interface and superficial lymphocytic perivascular dermatitis suggestive, but not diagnostic, of a drug eruption. No septic emboli, bacteria, or fungi were seen.

The patient remained hospitalized for 4 days. The erythema of her left ear pinna and surrounding skin had completely resolved, but the patient continued to have a rash. Serial complete blood counts were within normal limits. The patient was discharged on oral clindamycin to complete a 10-day course.

A repeat skin punch biopsy with an effort to obtain subcutaneous fat was performed. Review by a dermatopathologist concluded with a diagnosis of PLEVA.

Discussion

Abrupt onset of papulovesicular eruptions that evolve into necrotic lesions characterizes the rare disease PLEVA. The rash typically appears on the trunk and extremities, but the face and scalp are involved in approximately 10 percent of cases.¹ Al-

though pityriasis lichenoides in its chronic form was first described in 1894, Mucha and Habermann later described the acute form.² In 1966 Degos et al³ reported a hyperacute variant now recognized as febrile ulceronecrotic Mucha-Habermann disease (FUMHD). Contemporary authors, referring to the characteristics of the individual lesions, categorize PLEVA as acute or chronic.¹ Although PLEVA is often described as a type of papulosquamous parapsoriatic disease, the best way to categorize the clinical variants of PLEVA remains under discussion.

The cause of PLEVA is not known. Evidence suggests that PLEVA is a hypersensitivity reaction to an infectious agent. There have been reports of an association with endocrinopathy, rheumatoid arthritis, polyarthritides, and other autoimmune diseases.^{2,4} PLEVA occurs in adults and children, more commonly affecting those who are male. FUMHD is rare; in a 1996 review, only 15 patients with this variant had been reported.²

Clearly the patient described in this report had a relatively mild case of PLEVA. It is noteworthy that the patient was on methotrexate at the time of her illness, because methotrexate is proposed as a treatment in severe cases of FUMHD, especially if other therapeutic measures fail.⁵ Meticulous supportive care and attention to superinfection of the skin lesions are important. PLEVA is usually self-limited. Other treatment options include erythromycin given for several months, phototherapy with psoralen ultraviolet A (PUVA) and UV-B, steroids, and dapsone.

The patient declined a prolonged course of erythromycin or any other intervention. No fulminant progression was noted after several months. Regression of many lesions spontaneously occurred. Long-term follow-up and consideration for surveillance biopsies have been recommended because of concern about a potential relation between lymphomatoid papulosis and chronic T-cell lymphoma.⁶

Recommendations

This case shows the need for a careful clinical approach when confronted with an unfamiliar skin disease. Repeating a skin biopsy might be necessary if the first specimen is nondiagnostic. The family physician should routinely discuss perplexing cases with the pathologist who is reviewing the biopsy; confirming the adequacy of the tissue sub-

mitted and explaining the clinical picture is instrumental. PLEVA was not considered in the initial differential diagnosis because the attending physician was unfamiliar with the clinical entity. Three subspecialty consultants, including a dermatologist, offered differential diagnoses that included infectious, connective tissue, and drug eruption causes. Persistence in pursuing a diagnosis of a mysterious rash pays off in the final diagnosis.

References

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