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CASE REPORT

A strange lupus-like malar rash with renal involvement: an angioimmunoblastic T-cell lymphoma – A case report

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Key Clinical Message

Cutaneous malar rash and kidney involvement has not previously been reported as presenting symptoms of an angioimmunoblastic T-cell lymphoma (AITL). We report a case of a woman with erythematous rash. A PET-CT revealed a lymphadenopathy and splenomegaly. An inguinal lymph node biopsy showed an AITL. There was clinical improvement after prednisone.

Keywords

Lymphoma, malar rash.

Background

Cutaneous eruptions associated with fever in elderly patients have a wide differential diagnosis. Hematological disorders are a rare cause of dermatological symptoms, and angioimmunoblastic T-cell lymphoma, which is an uncommon non-Hodgkin's lymphoma involving T cells, rarely involves the skin. We report two unusual manifestations of AITL: a malar lupus-like rash and renal involvement that, to our knowledge, have not been reported before.

Case Report

An 80-year-old obese woman with a history of mastectomy for breast cancer was admitted to our hospital in September 2013 for acute dyspnea and intermittent fever of 1-month duration. Initial chest radiography suggested bilateral pleural effusions, and laboratory values showed thrombocytopaenia and deterioration of renal function (plasma creatinine on admission was 2.31 mg/dL). There were no abnormal physical findings. After 10 days of treatment with penicillin–sulbactam and levofloxacin, an erythematous rash appeared on the abdomen and face that resembled the malar rash of lupus (Fig. 1A and B). We promptly discontinued the antibiotics, but the rash persisted.

Negative blood and urine cultures, negative screening tests for EBV, CMV, HIV, HCV, Echo-, Coxackie-, and Adeno-viruses, and normal transthoracic echocardiography made an infectious process unlikely. An abdominal ultrasound showed moderate splenomegaly. Because of the malar rash and fever, we examined the autoimmune profile and found proteinuria, but the rheumatologic markers (ANA, anti-ENA, and ANCA) were negative. Lactic dehydrogenase, beta 2 microglobulin, and uric acid levels were elevated. Angiotensin-converting enzyme (ACE) plasma activity was resulted at the upper limit of the normal. A total body FDG-PET revealed a systemic lymphadenopathy and splenomegaly (Fig. 2), and an inguinal lymph node biopsy showed altered architecture with perifollicular and nodular diffuse effacement and vascular proliferation, together with plasma cells, histiocytes, and immunoblasts. The immunophenotypic results were positive for the following markers: CD3, CD2, CD4, PD1, Bcl6, and CD10. A biopsy of the bone marrow showed diffuse lymphocytic infiltrates. The patient was diagnosed with an angioimmunoblastic T-cell lymphoma (AITL).

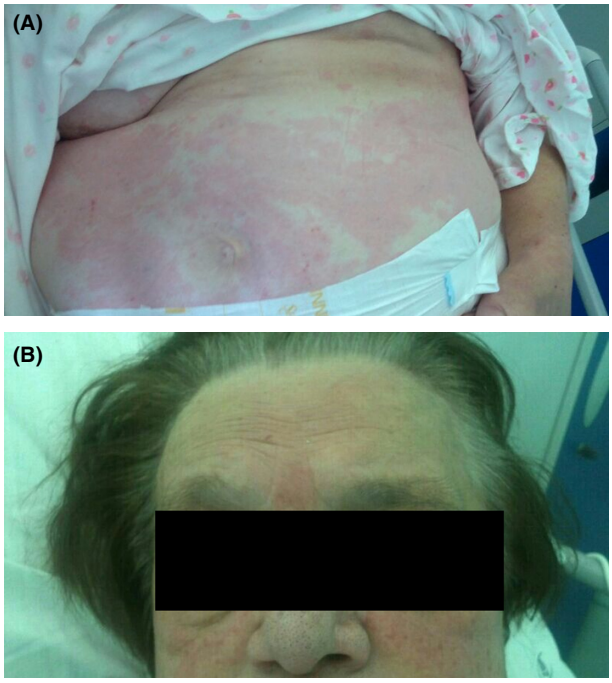

Figure 1. (A and B) Skin manifestations (face and abdomen).

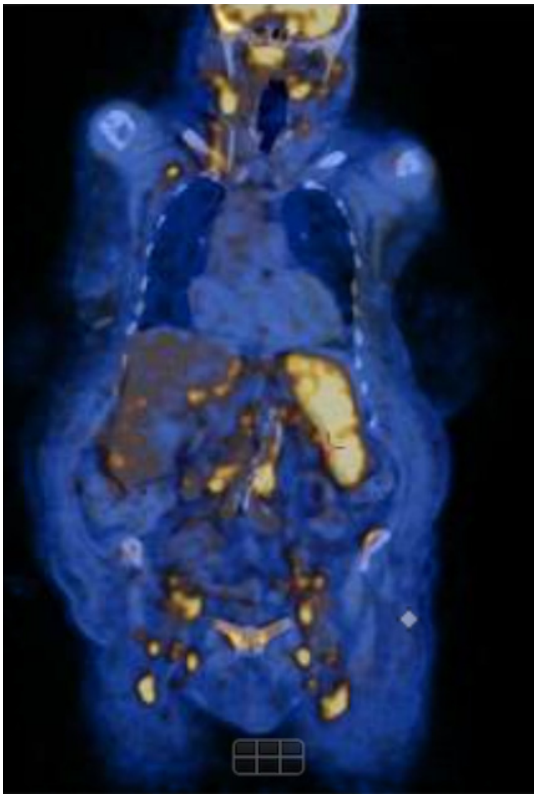

Figure 2. PET-CT showing lymphadenopathy and splenomegaly.

Treatment with 50 mg of prednisone daily was followed by clinical improvement; erythema, fever, anemia, and renal dysfunction resolved after a few days. After 1 month of treatment was completed with cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP), the patient remained in complete remission, with a follow-up of 1 year.

Discussion

Angioimmunoblastic T-cell lymphoma is a rare non-Hodgkin's lymphoma that involves T cells. It is characterized by generalized lymphadenopathy, fever, weight loss, night sweats, pruritus, and autoimmune manifestations, especially a skin rash on the trunk, abdomen, or, rarely, the arms. To the best of our knowledge, there are a few reports [1–3] of this non-Hodgkin's lymphoma presenting with either a lupus-like malar rash or with renal involvement, but no patients have presented with both. Cutaneous eruptions are usually caused by either drugs or viral infections. For example, up to 5% of patients receiving penicillins, sulfonamides, captopril, phenytoin, or gold will develop a maculopapular eruption. Accompanying features may include pruritus, fever, eosinophilia, and transient lymphadenopathy. Similar maculopapular eruptions are normally seen in the classic childhood viral exanthems, including measles, and in infections caused by Epstein–Barr Virus, Echovirus, Coxackie virus, and Adenovirus. The early lesions of Rickettsial and meningococcal infections can include erythematous macules and papules before they become purpuric. Maculopapular eruptions are associated with early HIV infection, early secondary syphilis, typhoid fever, and acute graft–versus-host disease. Rheumatologic diseases are also often accompanied by a skin rash. For example, a malar rash can be seen in 90% of patients with systemic lupus erythematosus, and its multiorgan component affects the peripheral joints and kidneys, leading to proteinuria and deteriorating organ function.

The cutaneous features of angioimmunoblastic T-cell lymphoma most commonly comprise a maculopapular eruption on the trunk and abdomen, but purpura, plaques, papulovesicular lesions, nodules, and erythroderma have also been reported. Forty-four percent of patients have a nonspecific maculopapular dermatitis, which precedes other clinical symptoms by at least several weeks [4, 5], suggesting that AITL should be included in the differential diagnosis of any maculopapular eruption of unknown etiology that is accompanied by lymphadenopathy. The histological findings of AITL in lymph nodes are characteristic, while those in the skin may be very subtle, comprising only mild lymphoid infiltrates. The rash in our case of AITL with cutaneous and renal involvement

mimicked a toxic erythema, drawing attention to the need to discriminate between a monoclonal proliferative disease and autoimmune or other skin rashes (Table 1).

Patients with AITL may be treated with high steroid doses to relieve symptoms caused by the immune response to the neoplastic cells, such as joint inflammation or pain and skin rash. Recommended first-line therapy for treatment of the cancer is either a clinical trial or chemotherapy with a number of different drugs, such as CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) [6] and radiation therapy. Sometimes higher doses of chemotherapy followed by a stem cell transplant may be required for those who relapse or for whom combination chemotherapy was not effective.

Treatment with 50 mg of prednisone daily was followed by clinical improvement; erythema, fever, anemia, and renal dysfunction resolved after a few days. After 1 month of treatment was completed with cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP), the patient remained in complete remission, with a follow-up of 1 year.

Conclusion

To our knowledge, this is the first case report of AITL presenting with a malar lupus-like rash and renal involvement. The diagnosis was based on the histopathology of a lymph node biopsy.

Table 1. Differential diagnosis of various types of rash.

Disease	Etiology	Description	Clinical syndrome
Measles (Rubeola)	Paramyxovirus	Maculopapular erythematous rash that begins several days after the fever starts	Fever, cough, runny nose, red eyes
German measles (rubella)	Togavirus	Rash beginning on the face which spreads to the rest of the body	Cervical lymphadenopathy
Erythema infectiosum (fifth disease)	Human parvovirus B19	Bright red slapped cheek with diffuse lacy reticular rash	Rash and fever
Exanthem subitum (roseola)	Human herpes virus 6	Maculopapular eruption (no face involvement)	Mild fever and arthritis
Primary HIV infection	HIV	Nonspecific	Adenopathy, arthralgias
Infectious mononucleosis	EBV	Diffuse maculopapular eruption, urticaria, periorbital edema	Lymphocytosis, hepatosplenomegacervical lymphadenopathy
Other viral exanthems	Echoviruses 2, 4, 9, 11, 16, 19, 25	Skin findings mimicking rubella or measles	Nonspecific viral syndromes
Drug-induced eruption	Drugs (antibiotics, anticonvulsionants, diuretics, etc.	Pruritic, Bright red macules and papules on trunks and extremities, sometimes confluent	Fever and eosinophilia
Typus		Maculopapular eruption	Headache, myalgias
Rickettsial spotted fevers	Rickettsia	Proximal extremities maculopapular eruption	Headache, myalgias and regional adenopathy
Ehrlichiosis	Ehrlichia	Maculopapular eruption	Headache, myalgias and leukopenia
Leptospirosis	Leptospira interrogans	Maculopapular eruption, conjunctivitis, scleral hemorrhage	Myalgias, meningitis
Lyme disease	Borrelia burgdorferi	Erythematous annular lesion with central clearing	Headache, myalgias, photophobia
Typhoid fever	Salmonella Typhi	Erythematous macules usually on trunks (rose spots)	Abdominal pain and diarrhea
Rat-bite fever	Spirillum minus	Violaceous or red-brown central rash	Adenopathy and fever
Relapsing fever	Borrelia	Central rash with petechiae	Recurrent fever, Headache, myalgias, hepatosplenomegaly
Erythema marginatum (rheumatic fever)	Group A Streptococcus	Erythematous annular papules and plaques	Fever, myalgias, arthralgias
Systemic lupus erythematosus	Autoimmune disease	Rash malar, photosensitive dermatitis, generalized maculopapular rash, discoid rash	Fever, autoimmune manifestations, anemia
Non-Hodgkin	Hematological disease	Various skin manifestations	Intermittent fever, itch, night sweats, cutaneous manifestations and malar rash

Conflict of Interest

None declared.

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