Ind J Dermatol Venereol Leprol 1993; 59 : 308-10

PARA-NEOPLASTIC STEVENS JOHNSON SYNDROME

Laxmi V Nair, M P Binitha, Najeeba Riyaz

A 14-year-old boy with haemorrhagic crusting of lips and congestion of eyes of a month's duration progressed to Stevens Johnson syndrome. A retro-peritoneal lymphoma was detected after extensive investigation. Stevens Johnson syndrome when persistent and atypical in presentation needs investigation for underlying malignancy.

Key Word : Stevens Johnson syndrome

Introduction

Paraneoplastic erythema multiforme is a well-known phenomenon. But very few cases of Stevens Johnson syndrome due to internal malignancy have been reported.[1] It occurs during the late stage of malignancy, or while the patient is on chemotherapy or radiotherapy.

Case Report

A 14-year-old boy was seen with haemorrhagic crusting of the lips and congestion of the eyes, of a month's duration, which supposedly developed following intake of ampicillin and salbutamol for acute bronchitis.

He was treated symptomatically for a week, but gradual progression of the lesions necessitated systemic steroids in a dose of 40 mg of prednisolone per day. In spite of this, he developed multiple, erythematous papules interspersed with purpuric and iris lesions on the face and acral parts. These soon became necrotic and ulcerated. Haemorrhagic crusting of the nasal vestibules, lid margins and extensive oral erosions was present (Fig.1). General and systemic examination was unremarkable except for marked wasting, pallor and continuous high grade fever.

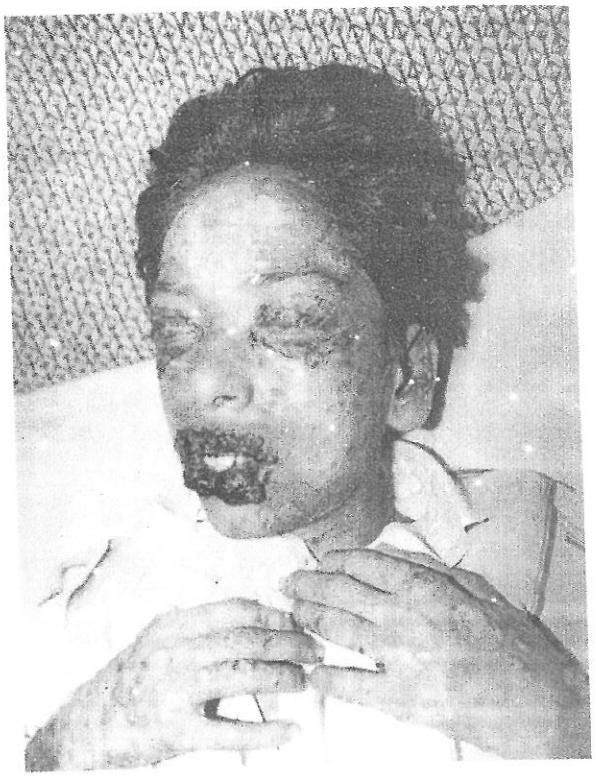

Fig.1. Necrotic papules on the face and acral parts with haemorrhagic crusting of the mucocutaneous junctions

Routine haematologic and urine examination were normal except for slight elevation of ESR and normochromic anemia. The peripheral smear was initially normal, and later showed neutrophilia with toxic granules. Reticulocyte and platelet counts, bone marrow examination, blood urea and

From the Department of Dermatology & Venereology, Calicut Medical College, Kerala-673 008, India.

Address correspondencde to: Dr Laxmi V Nair

sugar, serum electrolytes, creatinine and uric acid were normal. Serum proteins showed a reversal of albumin-globulin ratio. Cultures of blood, urine, sputum and skin lesions showed no bacterial growth. Direct Coomb's test, sickling test and Widal test were negative. Antibodies against HIV and hepatitis B virus were not detected. Anti-nuclear factor, anti-DNA, anti dsDNA antibody, serum immune complexes and complement C_3 and C_4 levels were normal. Serum immunoglobulins showed IgG 17 mg% (normal 1-1.25g%), IgM 14 mg% (50-125) and normal IgA. X-rays of the chest and abdomen were normal. Biopsy from a papule on the forearm showed hyperkeratosis, focal areas of basal cell degeneration and marked spongiosis in the epidermis. There was no evidence of acantholysis. Mononuclear infiltrate was seen in the upper dermis with congested capillaries. The picture was consistent with erythema multiforme. Facilities for immunofluorescent tests were not available.

One and a half months after admission, an ill defined, firm, non-tender mass was palpated in the hypogastrium. Ultrasound scanning of the abdomen showed a well-defined, oblong hypoechoic mass 6x4.5 cm in size, in the left iliac fossa. CT scan revealed a fairly large, circumscribed, solid, soft tissue mass in the left lower quadrant of the abdomen, extending into the pelvis and lying along the postero-left lateral aspect of the bladder, causing compression and displacement of the adjacent bowel loop and the lower portion of the left ureter. The CT finding was suggestive of a retroperitoneal lymphoma. Fine needle aspiration cytology and CT scan guided biopsy were attempted twice without success. Since the patient's general condition deteriorated, an exploratory laparotomy was deferred.

With these findings, a diagnosis of Stevens Johnson syndrome secondary to lymphoma was made. He developed extensive pneumonia, CSOM and keratoconjunctivitis and succumbed to the disease.

Discussion

Though our patient had extensive, persistent, vesicular and necrotic, cutaneous and mucosal lesions resembling paraneoplastic pemphigus, the lack of denudation and the absence of acantholytic cells prompted us to make a diagnosis of paraneoplastic Stevens Johnson syndrome. The occurrence of target lesions on the face and acral parts are points in favour of our diagnosis.

Atypical cases of ulcerative and blistering mucocutaneous disease associated with pempigus-like antibodies are well-known.[2-6] In retrospect, all patients with high titres of auto-antibodies had underlying neoplasms, frequently lymphoma. These patients had painful erosions of oropharynx and vermilion borders of the lips, which were resistant to conventional therapy, and most patients also had severe pseudomembranous conjunctivitis. The cutaneous lesions were pruritic and characteristically polymorphous, and the affected sites eroded, causing extensive denudation. Erythematous papules of the trunk and extremities evolved to form target lesions with central blisters and resembled erythema multiforme or toxic

epidermal necrolysis in the advanced stages.[7]

The exact pathogenesis of paraneoplastic Stevens Johnson syndrome is unknown, though a hypersensitivity mechanism has been postulated.[8] Recognition of an apparently irrelevant alteration of the skin, as being due to the presence of a malignant disease, may help in early diagnosis and successful treatment of the disease. Stevens Johnson syndrome, when persistent and atypical in presentation, should prompt investigation for underlying malignancy.

References

1. Fuller CJ. Hodgkins disease with erythema nodosum. Br Med J 1934 ; 2 : 1172.
2. Matsuoka LY, Wortsman J, Stanley JR. Epidermal autoantibodies in erythema multiforme. J Am Acad Dermatol 1989 ; 21 : 677-80.
3. Cruz PD Jr, Coldiron BM, Sontheimer RD. Concurrent features of cutaneous lupus erythematosus and pemphigus erythematosus following myasthenia gravis and thymoma. J Am Acad Dermatol 1987 ; 16 : 472-80.
4. Ansel J, Petrozzi JW, Kumar V. Possible drug-induced pemphigus-like antibodies with the clinical manifestations of erythema multiforme. Arch Dermatol 1983 ; 119 : 1006-9.
5. Ramseur WL, Richards FH, Duggan DB. A case of fatal pemphigus vulgaris in association with beta interferon and interleukin-2 therapy. Cancer 1989 ; 63 : 2005-7.
6. Panteleyeva GA. Paraneoplastic bullous dermatoses. Vestn Dermatol Venereol 1990 ; 2 : 50-2.
7. Anhalt GJ, Kim S, Stanley JR, et al. Paraneoplastic pemphigus. N Engl J Med 1990; 323 : 1729-35.
8. Elias PM, Fritsch PO. Erythema multiforme. In : Dermatology in General Medicine (Fitzpatrick TB, Eisen AZ, Wolff K, et al, eds), 2nd edn. New York : McGraw-Hill, 1979 ; 295-303.