Case report

Sneddon- Wilkinson Disease in association with Non-Hodgkin Lymphoma

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Abstract: Sneddon-Wilkinson disease is a rare condition with chronic recurrent course. It may be a part of paraneoplastic manifestation of underlying malignancy. The disease is diagnosed on the basis of its characteristic clinical features and histopathology. A case of Sneddon –Wilkinson disease associated with non-hodgkin lymphoma in a 54 years old lady is presented.

Key words: Sneddon-Wilkinson disease, Non-Hodgkin lymphoma, Paraneoplastic manifestation, Dapsone.

Introduction

Sneddon–Wilkinson disease also known as subcorneal pustular dermatosis (SPD), is a rare idiopathic, chronic recurrent pustular eruption first described in 1956.[1] It is more common in females and in individuals above 40 years of age[2]. It manifests with[2]: 1) flaccid often hypopyon pustules, which can be in annular arrangements. 2) a chronic course. 3) an absence of psoriasis. SPD usually involves the axillae, groin, abdomen and flexural aspects of the proximal extremities of middle aged women. SPD is a part of spectrum of neutrophilic dermatosis, which is characterized histologically by an accumulation of neutrophils in subcorneal layer separated from a non-edematous epidermis without psoriasiform changes.[3] Culture of pustules fails to reveal bacterial growth.[2] Neutrophilic dermatosis including SPD are commonly associated with systemic inflammatory and neoplastic disorders, including inflammatory colitis, rheumatoid arthritis, monoclonal IgA gammopathy and hematological malignancies[4]. Subcorneal pustular dermatosis may be a part of paraneoplastic manifestation of underlying malignancy. These cutaneous manifestations can precede, occur concurrently with or follow the diagnosis of a neoplastic disease. They often represent the first clinical sign of an underlying neoplasm or the earliest symptom of relapse of a previous cancer.[5]

Case report

Fifty four years old lady, a teacher by profession, referred to dermatology out patient department of Tribhuvan University Teaching Hospital (TUTH) with two month history of progressive pustular eruptions appearing in crop, with slightly erythematous base distributed on flexural areas. The lesions were distributed bilaterally on axillae, infra-mammary folds, cubital areas, groins, lower abdomen, popliteal fossae and flexural surfaces of bilateral lower limbs including lower thigh (Figure 1). Characterstic hypopyon characterized by collection of fluid at lower dependent part of the lesion and clearing at upper half, was also noticed (Figure 1). Patient was treated with systemic antibiotic and steroid by a general practitioner, lesions improved in a week, then steroid was tapered, however lesions reappeared after a month of stopping the steroid. On examination, patient was febrile (high grade) and had generalized lymphadenopathy; the largest lymph node was on left axilla that measured 2x2 cm. There were multiple pustules over flexural areas of the body. The lesions were in different stage of evolution. Face, oral mucosa, palm and soles were spared. Liver and spleen were not palpable.

Routine laboratory investigations showed anemia (Hb, 9 gm %), leucocytosis (WBC, 14,000/cumm, with 84% polymorphs), ESR was elevated (55 mm at the end of one hour). Total protein was 55 gm/dl and serum albumin was

28 gm/dl. Peripheral blood smear showed normocytic normochromic anemia. Thyroid function test and liver function test were within normal limits. Serology for hepatitis B, hepatitis C and HIV I, II was negative. Antinuclear antibody (ANA) and Rheumatoid factor were also negative. *Brucella abortus* antibody titer was <1:80 and urine for Bence-Jones protein was negative.

A lesional biopsy showed features consistent with subcorneal pustular dermatosis; sections showed focal subcorneal collection of neutrophils and detached keratinocytes at the base of the pustule representing acantholytic cells, however, no typical suprabasal detachment noticed in the sections. The dermal papillae showed dilated capillaries and a few preivascular lymphocytes and occasional neutrophils. An axillary lymph node biopsy was performed and patient was discharged on supportive measure along with dapsone 100 mg per day. On follow up after two weeks, patient was afebrile and skin lesions were improved, however lymphadenopathy persisted. On next follow up after two weeks skin lesions recurred and was associated with high grade, intermittent fever. Patient was re-evaluated for skin lesions and fever. Lymph node biopsy was consistent with Non-Hodgkin lymphoma, diffuse large cell type *(Fig.2);* sections showed effacement of normal lymphoid architecture by proliferation of sheets of neoplastic large lymphoid cells. They showed polygonal shapes, moderate cytoplasm and large round nuclei with irregular membrane and coarse to vesicular chromatin pattern. Nuclei showed distinct prominent nucleoli 1-3 in numbers and increased numbers of mitosis, 2-5 per high power field. Both centroblastic and immunoblastic features were seen. Mature lymphocytes and a few plasma cells were seen in between neoplastic lymphoid cells.

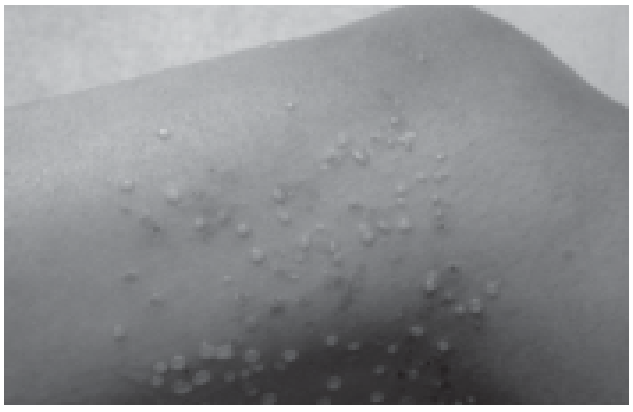

Fig. 1: Sneddon-Wilkinson disease: Crops of pustular eruptions at different stages of evolution, on flexural aspect of lower limb. A hypopyon is shown by an arrow

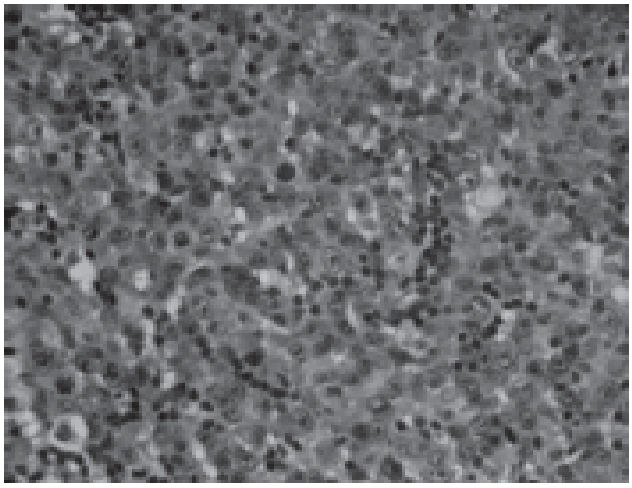

Fig. 2: Axillary lymph node biopsy in Sneddon-Wilkinson disease showing histology of diffuse large cell type of Non-Hodgkin lymphoma

Discussion

Our patient presented with pustular eruption characterized histologically by spongiform pustule formation. Clinical features and settings were characteristics of Sneddon-Wilkinson disease within the large spectrum of neutrophilic dermatomes. [1,2,3] Sneddon- Wilkinson disease can be a part of paraneopalstic manifestation of underlying malignancy.[5] Sneddon-Wilkinson disease has been reported in association with anaplastic large cell type of Non-Hodgkin lymphoma, and multiple myeloma. [6, 7, 8] Present case was a part of paraneoplastic manifestation of concurrent non-hodgkin lymphoma, diffuse large cell type. Clinically Sneddon-Wilkinson disease is characterized by small discrete flaccid pustules or vesicles that rapidly turn pustular and usually arise in crops within a few hours on normal or slightly erythematous skin. In dependent areas the pus characteristically accumulate in lower half of the pustules. As the lesion evolves they coalesce to form annular, circinate or bizarre patterns. Pustules rupture and dry up to form thin superficial scar and crust resembling impetigo.[2] There are no associated atrophy and scaring. Variable interval of quiescence lasting from few days to weeks may be followed by sudden development of new lesions[2] as in present case. The eruptions tends to occur symmetrically affecting mainly axilla, groin, sub-mammary areas, abdomen and flexural aspects of limbs with relative sparing of face, palm, soles, scalp and mucus membrane as was in present case.[2] Diagnosis is mainly based on typical distribution and characteristics of lesions. Absence of bacterial growth in pustules, ineffectiveness of systemic and topical antibiotics and the course of the disease differentiate it from impetigo.[2]

Absence of characteristic immunofluorescence findings differentiates it from dermatitis herpetiformis and pemphigus foliaceaus.[2] Its characteristic histopathology and response to Dapsone differentiate from pustular psoriasis[2]. The drug of choice is sulphones such as dapsone. A dose of 50-150 mg per day shows slower but good response as seen in present case. Systemic steroids are less effective and higher doses are required.[9]

Conclusion

Sneddon-Wilkinson disease in analogy to other neutrophilic dermatoses, however, it might be a manifestation of underlying severe disease. A complete evaluation of the patient is mandatory for proper management. An underlying malignancy should always be ruled out in the patient presenting with features of neutrophilic dermatosis. To the best of our knowledge, the present case is first reported case of Sneddon-Wilkinson disease in association with Non-Hodgkin Lymphoma in Nepal.

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