

(lymphocytic leukemia and monoclonal gammopathy).^[8]

Clinical backgrounds of patients with adult-onset xanthogranuloma are somewhat different from those of patients with juvenile xanthogranuloma, but the histological findings of both forms of the disease are identical. Extracutaneous involvement of the eye orbit, lung, liver, testis, central nervous system, kidney etc. has been reported in childhood variants, but is not seen in the adult type. Though there has been established association of JXG with neurofibromatosis (NF-1) and juvenile chronic myelogenous leukemia (JCML) in the childhood type, this has never been reported in adults. Evaluation for extracutaneous JXG is not indicated, unless there are symptoms or findings suggesting their presence, as they also disappear spontaneously. Differential diagnosis includes molluscum contagiosum, cryptococcosis, benign cephalic histiocytosis (seen exclusively in children, infiltrate lacks foamy cells and multinucleated giant cells), generalized eruptive histiocytosis (absence of granulation and lipidation), xanthoma disseminatum (lesions tend to merge into plaques, mucous membrane involvement, associated diabetes insipidus and different biopsy findings), papular xanthoma (JXG histologically recognized by its pure primitive histiocytic phase and presence of inflammatory cells, not seen in papular xanthoma). The importance of presenting this case is to highlight the fact that while making a diagnosis of common disorders like molluscum contagiosum, one must keep in mind the adult form of xanthogranuloma in differential diagnosis.

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Bullous pyoderma gangrenosum associated with ulcerative colitis

Sir,

Pyoderma gangrenosum (PG) is a rare non-infectious neutrophilic dermatosis associated with underlying systemic disease, characterized by distinctive cutaneous ulcers with undermined borders; lesions usually require aggressive therapy and they heal with a characteristic cribriform scar. We report a case of bullous PG associated with ulcerative colitis.

A 55-year-old woman presented with multiple fluid-filled lesions and ulcers over the upper and lower limbs and trunk of two months duration. There was a history of fever and loose stools on and off and loss of weight since two years. History of development of lesions at the site of trauma was elicited. There was no history of extramarital sexual contact, joint pains, mucosal ulcers and any other systemic symptoms. Examination revealed multiple, tender ulcers of varying size with undermined violaceous borders, vesicles and bullae [Figure 1] distributed over bilateral lower and upper limbs, palms and soles and a few over the trunk. Mucosa, hair and nails were normal. Systemic examination revealed no abnormality.

On investigation, peripheral smear showed hypochromic anemia with neutrophilia; renal function tests and liver function tests were within normal limits, serologic testing revealed negativity for VDRL, HIV 1 and -2, ANA, HBSAg and RA factor. Both bacterial and fungal cultures from the surface of ulcer were sterile; chest X-ray and ultrasound abdomen did not reveal any significant abnormality. No reaction was observed with Mantoux test. Colonoscopy and sigmoidoscopy showed red colonic mucosa with multiple ulcers of varying sizes and shapes and few pseudopolyps distributed throughout the length of the colon [Figure 2].

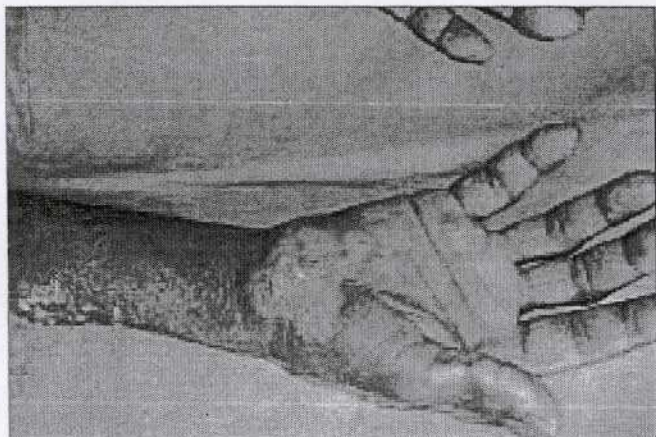


Figure 1: Multiple vesiculobullous and pustular lesions over the fore arm and extending to the palm



Figure 2: Dermis showing perivascular lymphocytic infiltration and focal neutrophilic abscess (H and E, X100)



Figure 3: Colonoscopy showing multiple ulcers in the colon

The skin biopsy taken from the edge of an ulcer revealed normal epidermis with dermis showing perivascular lymphocytic infiltration with endothelial swelling and focal

neutrophilic abscess [Figure 3]. Pyoderma gangrenosum commonly occurs in immunosuppressed patients secondary to accompanying disease, infections or therapy.^[1] The pathophysiology is poorly understood, an immune-mediated pathogenesis is suspected, both humoral and cell-mediated abnormalities have been associated with PG.^[2]

The skin lesions of PG appear as tender vesiculobullous, papulopustular lesions that develop into painful ulcers with dusky purple borders that are raised and undermined. The base of the ulcer typically reveals both granulation tissue and necrotic material. Lesion may be solitary or multiple and shows pathergy phenomenon. Several variants of PG have been described; ulcerative, pustular, bullous and vegetative, vulvar and peristomal PG. The peristomal PG is a recently recognized variant that occurs primarily in patients with inflammatory bowel disease. The skin lesions usually appear during the course of active bowel disease and frequently concur with exacerbation of colitis.^[3]

Other diseases commonly associated with PG include arthritis, hematological diseases and rarely chronic active hepatitis, myeloma, Takayasu's arteritis, systemic lupus erythematosus, Wegener's granulomatosis, diabetes mellitus, HIV infection and other neutrophilic pustular dermatoses, particularly Behçet's syndrome.^[4] Several studies have documented patients with PG and Behçet's syndrome, the two diseases share certain features such as arthritis, pustulation, aphthous lesions of mucous membranes and the phenomenon of pathergy.^[4,5]

The association of PG with ulcerative colitis varies from 1-50% in various studies, however, PG is an extremely rare disease occurring in less than 1% of patients with inflammatory bowel disease with an equal ratio of patients with Crohn's disease and ulcerative colitis, and similarly various other studies also demonstrated an incidence of less than 1%.^[6,7] Conversely 0.5-5% patients with ulcerative colitis have PG.^[8]

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Sporotrichoid pattern of malignant melanoma

Sir,

Malignant melanoma is a highly invasive neoplasm of skin, strongly influenced by environmental factors that develop in a genetically susceptible host. Incidence of melanoma is rising in Caucasians although it is a rare presentation in India.^[1]

A 28-year-old female presented with multiple painful lesions over her right leg of eight months duration. She was a farmer and claimed that the lesions started as a single painful nodule on the heel following a thorn prick. She ignored it then and subsequently the size and number of the lesions increased to involve the whole right lower limb as linearly arranged nodules. Cutaneous examination revealed multiple tender nodulo-ulcerative lesions with discharge, arranged in linear fashion from heel to thigh, present over the right leg [Figure 1]. Other findings included edema of the feet, thickened lymphatic channels between nodules along with bilateral stony hard, tender inguinal lymph nodes. The patient was previously diagnosed as sporotrichosis and treated with antifungal (itraconazole) drugs for six months



Figure 1: Multiple deep seated nodules of malignant melanoma in sporotrichoid pattern

in another healthcare facility without any clinical response. A differential diagnosis of sporotrichosis and malignant melanoma was considered. Limb X-ray, KOH smear, gram stained smear and fungal culture were negative. Skin biopsy revealed atypical melanocytes in the epidermis and dermis in nests. General examination revealed multiple generalized lymphadenopathy (axillary, cervical and submandibular lymph nodes). Abdominal sonography showed liver metastasis. Chest X-ray was normal. A diagnosis of malignant melanoma was established and the patient was referred to the oncology department for further management.

The incidence of melanoma continues to rise at an epidemic rate as evidenced by a 101.5% increase from the 1970s to the 1990s.^[2,3] Melanoma represents the fifth most common type of cancer, the most common type in women 25-29 years of age and the most common type in Caucasian men 25-44 years of age. But it is rare in Indian patients. Nodular melanoma and melanoma d'emblee are rare types of primary cutaneous malignant melanoma that are invasive and lack intraepidermal component.^[4] These lesions when first noted clinically are always palpable, convex in shape, of varying shades, rapidly increasing in size; neglected tumors may be several centimeters in diameter. Ulceration occurs fairly early. They can occur in any portion of the skin/mucosa.^[4]

Our patient presented with history of trauma with multiple nodular ulcerative lesions arranged in a linear fashion along the lymphatics over the lower limb which clinically simulated lymphocutaneous sporotrichosis. However, histopathology helped us to reach the correct diagnosis.

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