Pustulocrustaceous Secondary Syphilis

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Sir,
A generalized pustular eruption in an adult may be the presenting feature of viral infections, drug eruptions, psoriasis, subcorneal pustular dermatosis or impetigo herpetiformis. Rarely, generalized pustulation is a manifestation of secondary syphilis. We present here a case of secondary syphilis with a dramatic generalized eruption of pustules followed by impetiginous lesions. The classification of pustular syphilis is also reviewed herein.

CASE REPORT
A 20-year-old man presented with multiple ulcerated lesions over the trunk, extremities and genitalia of approximately 4 weeks’ duration. About 1 month earlier, the patient had developed pustular lesions all over the body. The lesions later burst open discharging pus and giving rise to multiple ulcers. The onset of ulcerated lesions was accompanied by fever of a moderate degree, malaise and sore throat. The patient gave a history of multiple, premarital, unprotected sexual contacts, over a span of 3 years. Further, the patient gave history of genital ulcer, about 2 years earlier, which was cured with some injections. The patient was a bachelor and a shopkeeper by occupation.

Examination of genitalia revealed multiple, variably sized, sharply defined, clean-looking mucous erosions on the undersurface of prepuce. There were multiple ulcerated, crusted lesions, ranging in size from 0.2 to 0.6 cm in diameter, over the front and back of the trunk (Fig. 1a), upper and lower extremities and the shaft of the penis (Fig. 1b).

The lesions were round to oval in configuration and sharply demarcated with erythematous borders. The covering crusts were brown-black in colour. Mechanical debridement readily removed the crusts and revealed a thin, suppurative layer of granulating tissue beneath. The posterior cervical, posterior auricular, occipital, axillary and inguinal lymph nodes were enlarged and firm, bilaterally, but not tender. There were no lesions in the oral mucosa or eyes. There was no evidence of an active or healed primary lesion.

Laboratory data disclosed the following values: haemoglobin 13.4 g/100 ml; haematocrit 40%; white blood cell count of 9600/mm³ with a normal differential count; normal urinalysis and a normal chest roentgenogram. Results of an intradermal skin test using intermediate strength purified protein derivative of tuberculin were negative at 48 and 72 h. Tzanck smear from one of the pustules showed many polymorphs without multinucleate giant cells. A Gram stain of the purulent material harvested from the pustules showed polymorphonuclear leukocytes. Neither multinucleate giant cells nor bacteria were identified. Results of dark-field examination of this material were negative for spirochaetes. HIV status of the patient was negative.

Results of cultures of sputum and from the pharynx and rectum were negative for pathogens, including Neisseria gonorrhoeae. Results of multiple cultures of the blood, urine and the cutaneous pustules were also negative. VDRL was positive to a dilution of 1:128 and...
the fluorescent treponemal antibody absorption test was positive.

A biopsy specimen taken from one of the ulcerating crusted lesions showed acanthosis and ulceration in an area of epidermis where there was an inflammatory exudate and necrosis. Dermis showed a chronic inflammatory infiltrate, comprising lymphocytes and plasma cells, which was perivascular in certain areas.

A presumptive diagnosis of pustular secondary syphilis was made. The patient was treated with a single dose of intramuscular benzathine penicillin G, 2.4 million units, after sensitivity testing. Within 10 days, the lesions healed completely.

DISCUSSION
Pustular secondary syphilis is a rare form of secondary syphilis, accounting for <2% of all secondary syphilis (1). It has been thought to occur commonly in debilitated patients; however recent reports, including our case, argue against that observation (2–5). Lesions of pustular syphilis consist of crops of pustules which may be follicular (2, 3) perifollicular (4) or associated with other forms of secondary syphilis such as framboesiform, papulosquamous and ulcerated.

The pustular syphilitic exanthemata have been divided on the basis of their clinical morphology into four prototypical categories (3). Small accumulate pustular syphilid (miliary pustular syphilid) consists of diffuse, small 1–3 mm, discrete, perifollicular pustules, symmetrically distributed on the trunk and extremities, often occurring with papular lesions and leaving depressed hyperpigmented areas when healed.

Large accumulate pustular syphilidem, also called acneiform, varioliform or obtuse syphilidem, is characterized by large, 3–6 mm, discrete accumulate peri-follicular pustules with infiltrated bases. They tend to show polymorphism and may be umbilicated, resembling variola lesions.

Flat pustular syphilidem (papulocrustaceous or impetiginous or eczymiform syphilidem) occurs on the face, scalp, trunk, genitalia and perianal skin. The lesions are superficial flat pustules ranging from 4 to 20 mm in diameter, surrounded by dull red areola. The pustules dry, leaving dirty yellow to brownish crusts covering shallow ulcers. The lesions may become confluent, forming a large convex crust called a carapace (4).

Pustuloulcerative syphilidem is similar to the impetiginous type, except that the inflammatory reaction extends deeper into the tissues. The ulcers are covered with black-brown crusts, which often have the appearance of oyster shells and are called rupial syphilidem. The term ‘lues maligna’ is used when the lesions of pustuloulcerative syphilidem are especially acute, widespread or severe and are associated with constitutional symptoms (6, 7).

The case presented in this report principally had impetiginoid and pustulocrustaceous lesions and should be best described as the pustulocrustaceous type of secondary syphilis.

Histopathological findings of pustular syphilidem include endothelial swelling and proliferation and fibrinoid necrosis resulting in obliterator endarteritis. There are dense perivascular lymphoplasmacytic infiltrates throughout the dermis. Subcorneal pustules, intrafollicular abscesses with necrosis of the follicular epithelium (2, 3), infarct necrosis of the upper dermis and epidermis (6) and non-caseating granulomatous infiltrates have also been reported (5).

The differential diagnosis of pustular syphilidem includes viral infection, pustular psoriasis, pustular drug eruptions, subcorneal pustular dermatosis and impetigo herpetiformis. Dark-ground examinations from the pustular lesions are frequently positive for spirochaetes (2, 4). The titre of VDRL is usually high in pustular secondary syphilidem (2, 4).

Pustular syphilidem responds to penicillin therapy at the same dosages recommended for the usual forms of secondary syphilis. Pustular secondary syphilidem takes a longer time to convert to seronegative than the maculopapular form (8). Jarisch-Herxheimer reaction after penicillin therapy is common in pustular syphilidem (2–4).

Dubbed ‘the great imitator’, syphilidem is notorious for its capacity to mimic a variety of other diseases and entities. Subtleness of symptoms and signs and mimicry of other diseases are hallmarks of secondary syphilidem, which have always made diagnosis by clinical examination difficult. Although the spectrum of clinical manifestations has not changed substantially, clinicians still must maintain a high index of suspicion and a reliance on confirmatory serological tests to establish a definite diagnosis.

REFERENCES