Pustular Psoriasis, Annular Type

Pustular psoriasis is an uncommon form of psoriasis whose main features include sterile pustules on an erythematous base. It is a rare disease. One study of 112 children with psoriasis found that 0.9% of cases involved generalized pustular psoriasis. Pustular psoriasis afflicts all races, is slightly more common in males, and may occur as early as the first week of life. Several cases of pustular psoriasis have been described in the context of Kawasaki disease.

VARIANTS OF PUSTULAR PSORIASIS

There are several forms of pustular psoriasis. The most common form is generalized pustular psoriasis (von Zumbusch type), an acute, severe form that is associated with fever, severe pruritus, toxicity, malaise, and painful pustulation. Generalized pustular psoriasis lesions in children have an annular morphology in 60% of patients. By contrast, our patient had the annular, or circinate, form that classically follows a subacute or chronic course and has less systemic involvement. Pustulosis palmaris et plantaris involves pustules limited to the palms and soles bilaterally and is associated with chronic recurrent multifocal osteomyelitis, which involves osteolytic bone lesions and arthritis. Other forms of pustular psoriasis include exanthematic, localized, juvenile, infantile, and pregnancy-associated types.

PATHOLOGY

Pustular psoriasis lesions develop on normal skin or on psoriatic plaques as red plaques covered with pinpoint subcorneal pustules. The sterile pustules then coalesce to form collections of pus, eventually leaving dry, brownish crusts followed by exfoliative dermatitis. Flexures, genital areas, webs of fingers, and periangual areas are predisposed. Mucous membrane involvement in the mouth and tongue is common. Laboratory tests may reveal lymphopenia; neutrophil leukocytosis; a high erythrocyte sedimentation rate; low levels of albumin, calcium, and zinc; and negative blood and pustule cultures. Antistreptolysin O titers should be checked and hypocalcemia. Causative medications include lithium, upper respiratory infection, pregnancy, stress, vaccinations, tonsillitis, steroid withdrawal, group A streptococcal infection, and hypocalcemia. Topical compresses, wet wraps, and baths with saline or oatmeal are soothing and gently debride the skin lesions. Topical steroids can also be used albeit very cautiously, as there is a high rate of absorption. Hydration, bed rest, avoidance of excess heat loss, and monitoring for secondary infections are important. Systemic therapy for cases recalcitrant to topical treatment includes oral retinoids, methotrexate, cyclosporine, hydroxyurea, and 6-thioguanine. Oral steroids are generally discouraged because they may cause the disease to flare upon withdrawal of the medication.

Although tumor necrosis factor inhibitors have been reported to cause the development of new pustular psoriasis in adults, they have been used to treat the condition in children. Phototherapy is rarely used in children, but 1 case report described use of acitretin and narrowband UV-B phototherapy as maintenance therapy after induction with cyclosporine.

NATURAL HISTORY AND PROGNOSIS

The course of pustular psoriasis is cyclical, with unexplained intermittent flares of disease that may occur across decades. Relapses are common and may become progressively severe. Flares may resolve spontaneously, leaving behind normal skin.

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REFERENCES