

# Denouement and Comment

## Pustular Psoriasis, Annular Type

**P**ustular 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.<sup>1</sup> Pustular psoriasis affects all races, is slightly more common in males, and may occur as early as the first week of life.<sup>2</sup> Several cases of psoriasis have been described in the context of Kawasaki disease.<sup>3</sup>

### 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.<sup>2</sup> 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.<sup>4</sup> 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.<sup>2</sup> Flexures, genital areas, webs of fingers, and periungual 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.<sup>5</sup> Antistreptolysin O titers should be checked to rule out a triggering group A streptococcal infection.

Reported triggers of pustular psoriasis include medications, steroid withdrawal, group A streptococcal infection, upper respiratory infection, pregnancy, stress, vaccinations, and hypocalcemia. Causative medications include lithium, iodine, penicillin, and interferon.<sup>1,5</sup>

### COMPLICATIONS

Fortunately, complications in pustular psoriasis are rare. Bacteremia may occur secondary to loss of skin-barrier function. Liver impairment can occur secondary to the disease itself or as a result of hepatotoxicity from methotrexate or retinoids used to treat the disease. Renal impairment has also been reported. Death may result from cardiorespiratory failure during the acute eruptive phase.

### TREATMENT

The approach to treatment of pustular psoriasis should be conservative, starting first with topical therapy and then considering systemic medication if the disease progresses. Topical

compresses, wet wraps, and baths with saline or oatmeal are soothing and gently debride the skin lesions.<sup>5</sup> Topical steroids can also be used albeit very cautiously, as there is a high rate of absorption.<sup>6</sup> 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.<sup>5,7</sup> 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.<sup>8,9</sup> 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.<sup>10</sup>

### 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.<sup>2</sup> Flares may resolve spontaneously, leaving behind normal skin.

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### REFERENCES

1. Cassandra M, Conte E, Cortez B. Childhood pustular psoriasis elicited by the streptococcal antigen. *Pediatr Dermatol.* 2003;20(6):506-510.
2. Paller A, Mancini A. *Hurwitz Clinical Pediatric Dermatology: A Textbook of Skin Disorders of Childhood and Adolescence.* 3rd ed. Philadelphia, PA: WB Saunders Co; 2006.
3. Zvulunov A, Greenberg D, Cagnano E, Einhorn M. Development of psoriatic lesions during acute and convalescent phases of Kawasaki disease. *J Paediatr Child Health.* 2003;39(3):229-231.
4. Girschick H. Chronic recurrent multifocal osteomyelitis in children. *Orphanet Encyclopedia.* March 2002. <http://www.orpha.net/data/patho/GB/uk-CRMO.pdf>. Accessed January 2007.
5. Baron E, Taylor C. Psoriasis, pustular. eMedicine Web site. <http://www.emedicine.com/DERM/topic366.htm>. Updated Jan 16, 2007; Accessed January 2007.
6. Rogers M. Childhood psoriasis. *Curr Opin Pediatr.* 2002;14(4):404-409.
7. Generalised pustular psoriasis. DermNet NZ Web site. <http://dermnetnz.org/scaly/pustular-psoriasis.html>. Updated March 18, 2008; Accessed May 17, 2007.
8. Callen JP, Jackson JH. Adalimumab effectively controlled recalcitrant generalized pustular psoriasis in an adolescent. *J Dermatolog Treat.* 2005;16(5-6):350-352.
9. Pereira TM, Vieira AP, Fernandes JC, Antunes H, Basto AS. Anti-TNF- $\alpha$  therapy in childhood pustular psoriasis. *Dermatology.* 2006;213(4):350-352.
10. Kim HS, Kim GM, Kim SI. Two-stage therapy for childhood generalized pustular psoriasis. *Pediatr Dermatol.* 2006;23(3):306-308.