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CASE DISCUSSION

Hot and hobbling with hives: Schnitzler syndrome

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Case

A 52-year-old man has recently been referred for evaluation of a periodic fever syndrome. He has had recurrent fevers for the past 2 years and often misses work with the fevers because his legs hurt and he is unable to function. The fevers occur at random intervals and last approximately 1 day. When the fevers began 2 years ago, he was thoroughly evaluated for infectious and malignant causes of his fevers and a bone marrow at that time was negative. Extensive cultures and serologic evaluations were also negative. He was born in the United States but is of Italian ancestry. He has no history of travel nor of ingestion of imported foods. An empiric trial of colchicine for familial Mediterranean fever was not helpful. Beginning approximately a year ago, he has had a chronic skin condition consisting of red raised macules of approximately 1 cm. The macules are not fixed and evolve and change every day but he has had them consistently for the past year. A biopsy of an active skin lesion was non-specific. A trial of a steroid cream and oral anti-histamine was not helpful. He has postponed previous medical evaluations because he generally feels well between fevers. Recently, however, he has lost weight and has felt fatigued even on his "good" days. On examination, he was found to have mild cervical and inguinal adenopathy and extensive non-pruritic urticarial lesions over his extremities. Protein was found in his urine on a rapid dipstick evaluation.

Commentary from Dr. Dan Lipsker

I have chosen to narrow the differential diagnosis by discussing first the skin rash. It is described as "red raised macules of approximately 1 cm". The lesions are not fixed, evolve and change every day, but are consistently present. This short-lived chronology of the primary lesion is highly suggestive of urticaria. The elementary lesions in most other skin eruptions are more long-lasting. The causes of patients with recurrent fever and skin rash are illustrated in Table 1.

The possibility of *chronic meningococcemia* should always be raised in patients with recurrent fever and an eruption. However, the rash in this condition is usually maculo-papular and sometimes petechial, and patients complain about arthralgia and/or tenosynovitis. Blood cultures should always be performed and the microbiologist should be aware of the suspected diagnosis. A dramatic improvement with antibiotic treatment within 24—48 h is a further indication of this diagnosis. Yet, this diagnosis can probably be ruled out in this case because this patient was said to have had extensive cultures and serologic evaluations 2 years ago.

A biopsy of an active cutaneous lesion was said to be non-specific. This eliminates the possibility of *urticarial vasculitis*. Indeed, hypocomplementemic urticarial vasculitis, often occurring in the setting of lupus erythematosus had to be ruled out in this patient. This non-specific biopsy

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Table 1 Causes of a recurrent febrile rash

Disease			Distinguishing features
Chronic meningococcemia			Blood cultures; rapid improvement within 24—48 h after antibiotic treatment
Adult onset Still disease			Elevated, low-glycosylated, ferritine levels
Hypocomplementemic urticarial vasculitis			Cutaneous biopsy shows vasculitis; low complement levels; lupus erythematosous is often associated
Acquired C1 esterase inhibitor deficiency, especially in the setting of lymphoma and/or paraproteinemia			Usually angio-oedema; low C4 levels, low functional C1-inh levels
Cryoglobulinemia			Temperature-dependency of clinical signs, presence of cryoglobulins Usually vasculitis on cutaneous biopsy
Auto-inflammatory syndromes	Hyper IgD syndrome		Increased IgD levels ; mevalonate kinase mutation
	Tumor necrosis factor receptor-associated periodic syndrome (TRAPS)		Mutations in the soluble TNFRSF1A gene in the 12p13 region
	CIAS1 mutation	CINCA*syndrome	Begins in childhood; neurological involvement; joint deformities; dysmorphic facial appearance
		Muckle—Wells syndrome	Deafness and amyloidosis; family history
		Familial Cold Autoinflammatory Syndrome	Temperature-dependency of clinical signs; conjunctivitis; family history
Erythema marginatum			Preceding streptococcal infection; elevated anti-streptococcal antibodies
Schnitzler syndrome			Monoclonal IgM gammopathy

CINCA (chronic infantile neurological cutaneous and articular syndrome) is also called NOMID (neonatal onset multisystem inflammatory disease).

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