A 9-year-old boy presented with a 1-month history of asymptomatic facial lesions. The lesions started around his mouth and spread to a perinasal and periorcular location. His mother had been applying over-the-counter hydrocortisone twice daily for 2 weeks without noted improvement.

His medical history was unremarkable. Family history was negative for skin disease. Physical examination revealed multiple erythematous 1 to 2 mm papules with scale in a periorificial location (Figure). The remainder of the physical examination was within normal limits.

Editor's note: Each month, this department features a discussion of an unusual diagnosis in genetics, radiology, or dermatology. A description and images of the patient and his or her condition will be presented, with the diagnosis and an explanation of how the diagnosis was determined following. As always, your comments are welcome.
DIAGNOSIS

Granulomatous perioral dermatitis

DISCUSSION

Granulomatous perioral dermatitis was initially described in the French literature by Gianotti et al. in 1970 and was later reported in the United States by Frieden et al. as a distinct clinical and histopathological entity. This disorder most commonly occurs in prepubertal children and is seen equally in males and females without a racial predilection.

The cutaneous lesions are erythematous or flesh-colored papules distributed in a perioral, periorcular, and perinasal location. The term perioral is somewhat of a misnomer; periorificial has been used to more accurately describe this eruption. The presence of scale and pustules in the affected areas may be noted, and some children complain of pruritus. Lesions may also occur on the neck and trunk. Histopathology of the papules reveals a dermal granulomatous infiltrate with surrounding lymphocytes. This granulomatous infiltrate typically predominates in perifollicular regions and follicular rupture may be noted. A biopsy is rarely indicated, as the diagnosis can be made by the presence of the clinical features alone.

The differential diagnosis includes acne rosacea, benign cephalic histiocytosis, allergic and irritant contact dermatitis, and sarcoidosis. Although the histologic features of acne rosacea are similar to those of perioral granulomatous dermatitis, other features of acne rosacea, including flushing, pustules, nodules, cysts, and telangiectasias are not typically seen. In addition, the distribution of the lesions in acne rosacea is not primarily periorificial. Benign cephalic histiocytosis is an eruption of yellow-brown and pink papules on the forehead, cheeks and eyelids. Histologically, a dermal histiocytic infiltrate is present. Perioral granulomatous dermatitis can be distinguished from cutaneous sarcoidosis by the presence of lymphocytes and the lack of systemic findings in the former.

Treatment options include topical metronidazole cream or gel or systemic erythromycin (or tetracycline in children older than 8). Many children respond to topical therapy alone, but systemic therapy may be needed for persistent or severe cases. Therapeutic response often occurs slowly, and a flare of skin lesions may occur in the first weeks of therapy. Children often require treatment for several months. Topical corticosteroids have been implicated in the pathogenesis of this disorder and should be discontinued. Abrupt discontinuation of topical corticosteroids may lead to temporary worsening of the eruption. Pitted scars can be seen after resolution, but recurrences are rare.

REFERENCES