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A Case of Elastosis Perforans Serpiginosa

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A 17-year-old white boy with no underlying connective tissue disorders presented with flat-topped annular plaques, with slight central atrophy on the bilateral neck. Results from histopathology revealed changes consistent with elastosis perforans serpiginosa (EPS). The idiopathic form of EPS occurs rarely in children. We report a patient with this rare pediatric diagnosis and review the literature.

Elastosis perforans serpiginosa (EPS) is a rare cutaneous connective tissue disorder, characterized by a thickened elastic fiber extrusion through narrow epidermal channels. It affects individuals between the ages of 6 and 30 years, with peak incidence in the second decade. Both sexes are affected, with males outnumbering females in a 4:1 ratio. Hyperkeratotic 1- to 3-mm reddish to skin-colored papules, with a central keratotic plug occurring singly or in arcuate and circinate patterns, are the common presentation of EPS. EPS can be asymptomatic or slightly itchy and usually affects the sides of the neck and face symmetrically. Histologically, it is characterized by an increase of eosinophilic elastic fibers in the papillary dermis, mixed inflammatory infiltrate, and acanthotic epidermis.

EPS may be associated with systemic disease, penicillamine treatment, or may be idiopathic. There have been several reported cases of idiopathic EPS in adults, but few cases have been reported in the pediatric population. In this study, we review the literature and report a case of idiopathic EPS in a pediatric patient.

Case Report

A 17-year-old white boy presented with a 6-month history of a scaly, nonpruritic area on his neck (Figure 1). It began on the left neck, spreading to the right neck and underneath the right chin. The area was treated initially with econazole nitrate for presumed tinea corporis, but without success. Subsequently, the lesions underwent slow expansion, with no spontaneous involution. Medical history was significant for a diagnosis of attention deficit hyperactivity disorder, for which he takes methylphenidate.

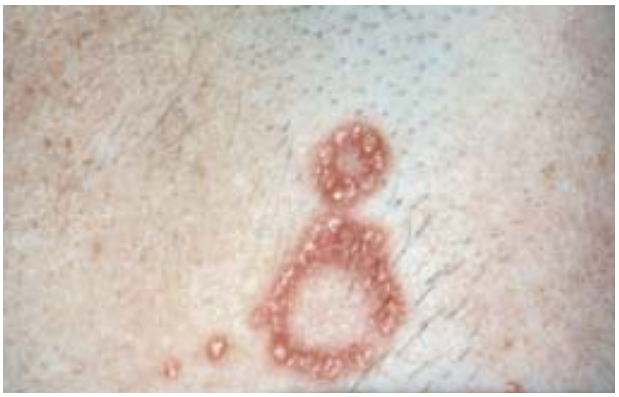

Figure 1. Minimally scaly, eythematous, annular 1-cm plaque, with central atrophy on the left neck.

Physical examination revealed an erythematous, flat-topped, annular, 1-cm plaque that was minimally scaly, with slight central atrophy on the left neck. A group of flat-topped, scaly lichenoid papules on the right side of the neck and under the right chin was noted (Figure 2). The patient did not have other similar lesions on the ankles, knees, lower extremities, wrists, or oral mucosa. One of the annular lesions was removed by punch biopsy for histologic examination (Figures 3 and 4).

Results from the biopsy specimen showed an epidermis with irregular acanthosis. In the papillary dermis, granular degeneration of the connective tissue was noted, with brightly eosinophilic strands that tested positive on the Weigert-van Gieson stain for elastin. The adjacent dermis showed a mild fibrotic reaction, with a patchy perivascular infiltrate of lymphocytes.

Comment

EPS was first recognized by Lutz[1] in 1953. EPS is one of the 4 classical perforating skin diseases that include Kyrle disease, perforating folliculitis, and reactive perforating collagenosis. The cause of EPS

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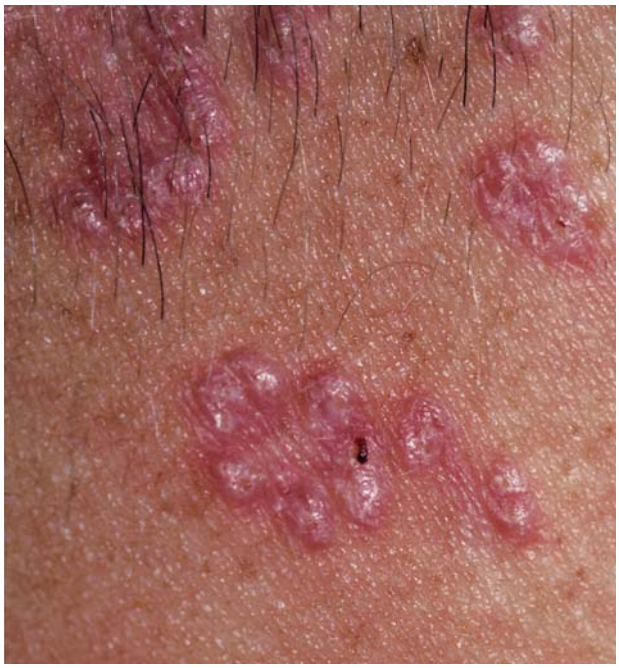

Figure 2. Flat-topped, lichenoid papules on the right side of the neck and under the right chin.

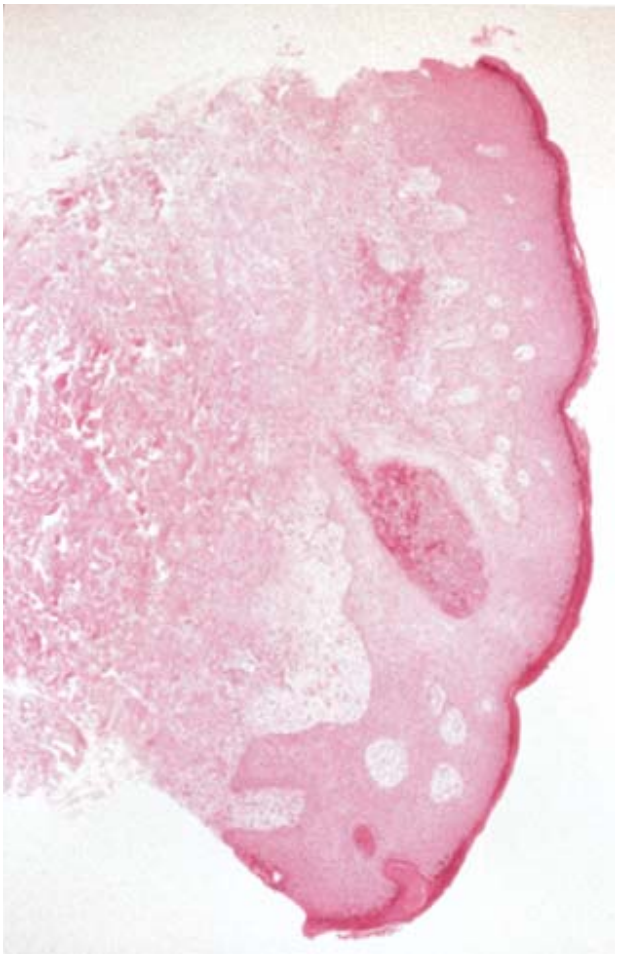

Figure 3. Epidermis with irregular acanthosis and granular degeneration of the connective tissue in the papillary dermis (H&E, original magnification ×10).

is unknown, but a genetically determined defect of elastic tissue with an autosomal-dominant mode of inheritance has been proposed for some cases.[2] Three forms of EPS are described: reactive, which is associated with genodermatoses and involves the connective tissue; secondary, which is caused by intake of penicillamine; and idiopathic, which is not correlated with systemic disease.

Reactive EPS accounts for 25% of all cases of EPS. The most common concomitant disease is Down syndrome, in which the lesions are more extensive.[3] Other associations with disorders of the dermal connective tissue include acrogeria,[4] Ehlers-Danlos syndrome,[4] Marfan syndrome,[5] morphea,[6] osteogenesis imperfecta,[7] pseudoxanthoma elasticum,[8] Rothmund-Thomson syndrome,[9] and scleroderma.[10]

The association between long-term penicillamine therapy and EPS also is well recognized.[11-15] In particular, patients with cystinuria who have been treated with long-term penicillamine are at risk for EPS.[15] Although the mechanism is unclear, EPS results from the direct effect of penicillamine on elastic fibers or the depletion of tissue copper—a cofactor in elastin synthesis[11]; this results in weakened or poorly cross-linked elastin and an increase in tropoelastin. EPS has not been reported in untreated Wilson disease. In rare cases, EPS appeared during a short course of low cumulative dose of penicillamine, as described in a patient with juvenile rheumatoid arthritis.[16]

Idiopathic EPS has been well reported in healthy adults. However, there are very few reports in the literature of this type of EPS in pediatric patients. Tuyp and McLeod[17] reported the case of a 14-year-old girl with an 8-month history of asymptomatic, gradually growing papules on both cheeks. Rubio et al[18] reported the case of a 12-year-old boy with a 9-month history of an asymptomatic, gradually enlarging papular eruption on the skin of his upper lip. In both cases, the diagnosis of EPS was confirmed by histologic examination.

Most treatments for EPS are unsatisfactory. Various therapies were used in our case with limited success. Application of topical or intralesional corticosteroids was largely unsuccessful. There are several case reports in which treatment with liquid nitrogen cryotherapy,[19,20] 13-*cis*-retinoic acid,[21] and oral isotretinoin[22] was successful, without scar formation. It has been reported that resurfacing with a CO_2 laser[23] and stripping of the surface

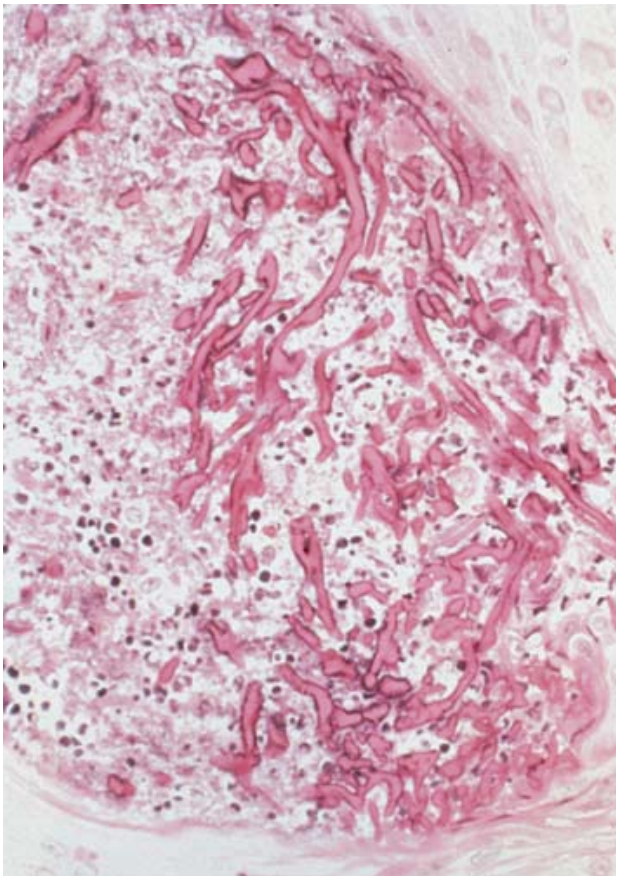

Figure 4. Adjacent dermis showed a fibrotic reaction, with a patchy perivascular infiltrate of lymphocytes (H&E, original magnification ×40).

keratinous material with cellophane[2] have some beneficial effects.

Although these lesions may persist for 6 months to 3 years without treatment, they eventually involute, leaving linear or reticulate scars that are hypopigmented and atrophic. Most children diagnosed with EPS are not disturbed by this disorder. Physicians should be aware of the association of EPS with underlying disorders of the connective tissue.

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