

# Elastosis Perforans Serpiginosa: A Case Report

Farhad Malekzad, MD  
Mojdeh Sarlak, MD

Department of Dermatology, Shaheed  
Beheshti University of Medical Sciences,  
Tehran, Iran

Corresponding author:  
Farhad Malekzad, MD  
Loghman Hospital, Tehran, Iran

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## Abstract

We report a 61-year-old man with multiple colored skin papules with central crusts in an archiform and linear pattern on his forehead as well as verrucus and hyperkeratotic plaques with central perforation on hands. Histologic examination of the lesions demonstrated a narrow channel of epidermal perforation containing notrophils, basophilic debris and elastic fibers with eosinophilic degeneration compatible with the diagnosis of elastosis perforans serpiginosa. (*Iran J Dermatol* 2008;11:89-91)

**Keywords:** perforating, elastosis, transepidermal elimination

## Case Report

A 61-year-old man presented with multiple colored skin papules with central crusts in an archiform and linear pattern on his forehead (figure 1) from 8 months ago. Also, verrucus and hyperkeratotic plaques with a steady growth and central perforation were seen on the back of hands (figure 2).

There were no specific signs or symptoms. Mental and physical examination, hair, nail and oral examination were normal. But there was a medical history of chronic renal failure and haemodialysis since 12 years ago. No drug history was detected. 2 biopsy specimens were obtained from the lesions on his forehead and dorsum of the hand.

Histologic examination of specimens revealed a narrow channel of epidermal perforation containing notrophils, basophilic debris and elastic fibers with eosinophilic degeneration (figure 3). Based on these findings, a diagnosis of elastosis perforans serpiginosa (EPS) was made and the patient was treated with cryotherapy, with some improvement and some regression in size.

## Discussion

Perforating dermatoses are a group of dermatoses with transepidermal elimination in which dermis is extruded through the epidermis to the exterior with little or no disruption of the surrounding structures.<sup>1</sup> The extruded material may include inflammatory cells, red cells, microorganisms and extra cellular substances such as musin or altered connective tissue components.<sup>2,3</sup>

There are four conditions that are regarded as a primary perforating disorder, i.e. Kyrle's disease, perforating folliculitis, reactive perforating collagenosis and perforating srpiginous elastosis. It is possible that these primary disorders might be due to a defect in the epidermal keratinocytes, hair follicle, collagen and elastic fibers respectively, with TEE being the final common pathway.<sup>2,3</sup>

Four conditions mentioned above occur in diabetes mellitus or in patients with chronic renal failure, most of whom may have undergone hameodialysis. Keratolic lesions in this condition develop on the trunk and limbs and are usually pruritic, dome shaped papules with central crusts. The term "acquired perforating dermatosis" is suggested for this condition.<sup>4</sup>

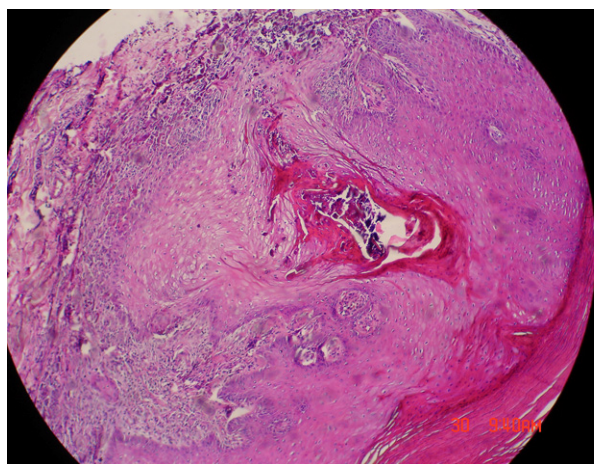
In 1953, Lutz described a chronic popular keratotic eruption in an archi form located on the sides of the nape of the neck.<sup>5</sup>



**Figure 1:** Multiple papules with central crusts on forehead



**Figure 2:** Hyperkeratotic plaques with central perforation on back of hands.



**Figure 3:** Transepidermal elimination of elastic fibres (H & E staining; X40)

In this perforating disorder, abnormal elastic fibers project above the surface. The cause is unknown but a genetically determined defect of elastic tissue may be involved, which provokes a cellular response that ultimately lead to extrusion of the abnormal elastic tissue.<sup>6</sup> Lesions are commonly seen in areas subjected to wear and tear. Some 40% of reported cases have been associated with connective tissue disorders such as pseudo xanthoma elasticum, Ehlers-Danlos syndrome, osteogenesis imperfecta, Marfan's syndrome, and acrogeria.<sup>7</sup> Rothman Thomson syndrome, systemic sclerosis and morphea, XYY syndrome and renal failure have also been associated with EPS. Most frequent concomitant disorder is Down syndrome. Approximately 1% of patients with Down syndrome have EPS, and the lesions are likely to be more extensive and persistent than in other patients. Several patents have developed EPS after prolonged treatments with penicillamin.<sup>5</sup> The earliest detectable pathologic change is focal development of elastotic tissues and basophilic

debris in the dermis. This is followed by a reaction of the overlying epidermis, which grows down to engulf the elastotic material. The distinctive histopathologic change consists of elongated, tortuous channels in the epidermis communicating directly with the dermis, forming horny material in its upper third and amorphous debris from elastin in its lower two-thirds. The epiderm surrounding the fully developed lesion is acanthotic and hyperkeratotic. In the dermis beneath and around the lesion, there is degeneration and alteration in the elastic tissue with foreign body giant cell reaction. The elastotic material is finally extruded which leaves irregular scarring and warty thickening.<sup>8</sup>

Clinically small, horny or umbilicated papules are characteristically arranged in lines or circles in a serpiginous pattern. The individual papules may remain small or may enlarge slightly to assume an elevated edge and a central plug or further to leave an area of atrophic skin surrounded by smaller papules.

The rings may reach a diameter of 15-20 cm. The disease is most common in young adults (6-20 y) and men. Back and sides of the neck are most commonly affected but the lesions may also occur elsewhere and are sometimes bilaterally symmetrical. They may persist for several years, but eventually involute spontaneously to leave reticulate atrophic scars.<sup>9,10</sup>

Annular and linear arrangement of the papules and their distribution suggest the diagnosis. Conditions which may cause confusion include prokeratosis of mibelli, reactive perforating collagenosis and perforating granuloma anulare.<sup>11</sup>

Careful removal of the nodules with a curette may give a reasonable result. Freezing has also been recommended.<sup>9</sup>

Excision should be avoided and dermabrasion may make the condition worse.<sup>2</sup> There are reports of improvement following pulsed dye laser, CO<sub>2</sub> laser and tazarotene.<sup>14</sup> Overall, clinical and histological data in our patient was compatible with the diagnosis of EPS.

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