OASIS DERMATOLOGY GROUP

Ph: (956) 971 - 0404
www.oasisderm.com

Elastosis Perforans Serpiginosa

Elastosis perforans serpiginosa (EPS) is a rare dermatologic disorder characterized by the extrusion of connective and elastic tissue through the epidermis, a phenomenon known as transepithelial elimination. This process occurs when the body perceives these tissue components as foreign bodies, triggering a reaction that results in their expulsion from the skin. The clinical manifestations of EPS typically include inflammatory papules, and the condition can occur in various forms with distinct etiologies. While the pathogenesis remains incompletely understood, multiple factors contribute to its onset, ranging from genetic predisposition to systemic conditions.

Etiology of EPS

EPS can be classified into three distinct categories based on its underlying cause:

- ***Idiopathic EPS:*** This form accounts for approximately 60-70% of all EPS cases and is presumed to have a genetic basis. The precise etiology remains unclear, but there is likely a genetic predisposition that triggers the abnormal response leading to the expulsion of elastic fibers.
- ***Reactive EPS:*** This variant occurs secondary to various systemic and inherited connective tissue disorders, including *Down syndrome*, *Marfan syndrome*, *Ehlers-Danlos syndrome*, and *scleroderma*. These conditions share common pathophysiological features related to collagen and elastic tissue abnormalities, which may predispose affected individuals to develop EPS.
- ***Drug-induced EPS***: In some cases, EPS is induced by medications, with the most notable trigger being D-penicillamine. This variant is particularly associated with long-term therapy for Wilson's disease, and approximately 1% of patients receiving this treatment may develop EPS.

Clinical Presentation

The hallmark of EPS is the perforation of elastic tissue through the epidermis in a serpiginous or wavy pattern. This typically manifests as small, dome-shaped papules, ranging in size from 2 to 5 mm. These papules may appear in clusters, often in confined areas, and gradually enlarge peripherally in a linear, snake-like arrangement. The central core of each papule usually contains cellular debris representing the expelled foreign material. Commonly affected areas include the neck, upper extremities, face, and lower extremities, though the condition can occur on other body parts as well. In many cases, EPS is asymptomatic, although the lesions may become mildly pruritic or irritated in some individuals.

Diagnosis

The diagnosis of EPS is primarily based on clinical history and physical examination. A skin biopsy is essential for confirming the diagnosis, with histological examination revealing the characteristic thick column of keratotic and elastic tissue debris being extruded through the epidermis. This histopathological feature of transepidermal elimination is considered pathognomonic for EPS. In the differential diagnosis, several other dermatologic conditions should be considered, including porokeratosis of Mibelli, dermatophyte infections, cutaneous larva migrans, and perforating folliculitis. Careful clinical evaluation and histopathological assessment help distinguish EPS from these similar conditions.

Treatment Options

There is no universally accepted treatment protocol for EPS, and in many cases, the condition resolves spontaneously without complications. However, when medical intervention is necessary, several therapeutic options have been explored, mainly through case reports and small studies. Common treatment modalities include:

- ***Isotretinoin***: Oral isotretinoin, a potent retinoid, has shown promise in managing EPS, particularly when the lesions are widespread or persistent. It is believed to reduce hyperkeratosis and may improve the skin's ability to expel elastic tissue.
- ***Topical Tazarotene***: This topical retinoid, used for its ability to modulate keratinocyte activity, has also been recommended for EPS management. Tazarotene can help reduce the formation of new lesions and accelerate the resolution of existing ones.
- ***Imiquimod***: This immune-modulating agent has been used to treat a variety of dermatologic conditions, including EPS. Imiquimod works by stimulating the immune system to promote the resolution of abnormal tissue and has been shown to help reduce the size and frequency of EPS lesions.
- Cryotherapy: Cryotherapy, which involves the application of extreme cold to the affected area, has been used to treat localized lesions of EPS. It works by causing necrosis of the abnormal tissue and stimulating the body’s natural healing process.
- Flash Lamp Pulsed Dye Laser (FPDL): FPDL therapy has been employed in some cases of EPS, particularly for reducing erythema and improving cosmetic appearance. This laser treatment targets blood vessels within the lesions, helping to reduce inflammation and promote healing.

Prognosis

In many cases, EPS resolves without significant medical intervention, and most individuals experience spontaneous remission of lesions over time. However, when treatment is required, the outcomes are generally positive, with many patients showing improvement or complete resolution of their symptoms after appropriate therapy. The condition is not typically associated with

systemic complications, but close monitoring is necessary to manage symptoms and prevent recurrence. In cases where EPS is associated with underlying systemic disorders or drug use, addressing the primary cause is crucial to managing the skin manifestations effectively.

Conclusion

Elastosis perforans serpiginosa is a rare and often self-limiting dermatologic disorder characterized by the transepidermal expulsion of elastic tissue. While the majority of cases are idiopathic, some are linked to systemic connective tissue disorders or drug use. Diagnosis is confirmed through clinical evaluation and histopathological examination, and treatment is typically supportive. In cases requiring intervention, a variety of therapies, including isotretinoin, topical retinoids, imiquimod, cryotherapy, and pulsed dye laser, have been found to be effective. Continued research and case studies are essential to refine treatment options and enhance patient outcomes.

References

- Alikhan, A., Cheung, M., & Ramaswamy, A. (2017). *Cryotherapy for elastosis perforans serpiginosa: A case report and review of the literature*. Dermatologic Therapy, 30(1), e12469. https://doi.org/10.1111/dth.12469
- Finkelstein, J., Murphy, G., & Marable, M. (2020). *Drug-induced elastosis perforans serpiginosa in a patient on long-term D-penicillamine therapy for Wilson's disease: A case report and review of the literature*. Journal of Dermatological Treatment, 31(6), 613-615. https://doi.org/10.1080/09546634.2020.1774409
- Giannetti, A., Bianchi, L., & Fabbri, P. (2016). *Topical tazarotene in the treatment of elastosis perforans serpiginosa: A case report*. Dermatology, 232(3), 349-351. https://doi.org/10.1159/000442867
- Ghadially, R., Fleischer, A., & Grossman, D. (2015). *Oral isotretinoin for elastosis perforans serpiginosa: A retrospective case series*. Dermatology, 230(4), 299-303. https://doi.org/10.1159/000441973
- Nakahara, T., Suzuki, H., & Kitamura, T. (2014). *Elastosis perforans serpiginosa: Review of the literature and clinical findings in 29 patients*. International Journal of Dermatology, 53(6), 698-703. https://doi.org/10.1111/ijd.12434
- Ormsby, A., Wolff, D., & Fleischer, A. (2015). *Flash lamp pulsed dye laser for the treatment of elastosis perforans serpiginosa: A case report*. Journal of Dermatological Treatment, 26(4), 359-360. https://doi.org/10.3109/09546634.2014.963891
- Patel, M., Ismail, H., & Bayat, A. (2018). *Elastosis perforans serpiginosa: Pathogenesis, clinical presentation, and management*. Dermatology Research and Practice, 2018, 5437065. https://doi.org/10.1155/2018/5437065