OASIS DERMATOLOGY GROUP

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

Atrophoderma of Pasini and Pierini

Atrophoderma of Pasini and Pierini (APP) is a rare, benign cutaneous condition characterized by the development of hyperpigmented depressions on the skin, primarily affecting the dermis. These lesions are typically oval or circular, with a distinct "cliff-drop" border and well-defined edges. APP is considered a slowly progressive condition, with a bilateral, symmetrical distribution that predominantly affects the back and lumbosacral region. While it more commonly occurs in females and typically emerges during adolescence or early adulthood, rare cases of congenital presentation have also been reported. Though the disorder tends to self-limit over time, its benign nature and lack of associated symptoms often lead to underdiagnosis.

Pathogenesis and Etiology

The etiology of APP remains unknown, though several potential factors have been suggested. One hypothesis is that the condition may be triggered by infectious agents, particularly due to an association with *Borrelia burgdorferi*, the bacterium responsible for Lyme disease. This has been supported by reports of patients with APP testing positive for *B. burgdorferi* antibodies. However, this association remains controversial, as not all patients with APP have a history of Lyme disease.

Another point of contention is whether APP represents a variant of morphea or a distinct clinical entity altogether. Morphea, or localized scleroderma, shares some clinical similarities with APP, such as the presence of skin atrophy and induration, but the underlying pathophysiology may differ. Some studies have suggested that both conditions may share similar autoimmune or inflammatory pathways, but definitive evidence is lacking.

The absence of a clear genetic link or environmental trigger further complicates our understanding of the disease, making it a subject of ongoing research.

Clinical Presentation

Patients with APP typically present with well-defined, hyperpigmented macules or plaques, which are violet to brownish in color. These depressed lesions are usually asymptomatic, with no associated itching, pain, or other discomfort. Over time, the size and number of the lesions can increase, although the condition is self-limiting and often halts progression by late adulthood. The "cliff-drop" border, where the lesion appears to descend sharply into the surrounding skin, is a distinctive feature used to differentiate APP from other dermatologic conditions.

Although the condition can affect any area of the skin, it predominantly involves the back and lumbosacral region. In rare cases, APP has been seen in other areas, including the upper limbs and abdomen.

Diagnosis

The diagnosis of APP is primarily clinical. The characteristic appearance of hyperpigmented depressions with well-demarcated borders is often sufficient for identification. However, due to the overlap with other conditions, particularly morphea and lupus, a skin biopsy is frequently performed to exclude differential diagnoses. While serologic tests for *Borrelia burgdorferi* antibodies may be useful in certain cases, they are not definitive, and the presence of these antibodies does not establish a causative relationship.

Management and Treatment

Currently, there is no universally accepted or definitive treatment for Atrophoderma of Pasini and Pierini, and therapeutic options remain largely empirical. Treatment goals are primarily aimed at controlling symptoms, preventing progression, and improving the cosmetic appearance of the lesions. Several approaches have been explored with varying success:

- ***Hydroxychloroquine***: This antimalarial drug has shown potential in some case reports for reducing skin atrophy and inflammation in APP patients. Hydroxychloroquine is thought to exert immunomodulatory effects, which may be beneficial in cases where autoimmune mechanisms are implicated.
- ***Retinoids***: Topical and oral retinoids, such as tretinoin and isotretinoin, have been used to treat a variety of skin conditions, including APP. Retinoids are thought to stimulate epidermal turnover and may help improve the cosmetic appearance of lesions by promoting cell regeneration and reduction of hyperpigmentation.
- ***Topical Steroids:*** Though the use of topical steroids has not been extensively studied for APP, some practitioners have reported mild improvements in inflammation and skin texture when used in combination with other treatments. These steroids help reduce the inflammatory component of the condition, though their efficacy for APP is limited.
- ***Antibiotics***: In patients who test positive for *Borrelia burgdorferi* antibodies, the use of antibiotics (such as doxycycline) may be considered, particularly in cases where an infectious etiology is suspected. However, the role of antibiotics in improving the dermal atrophy associated with APP remains unclear, and evidence for their effectiveness is limited.
- ***Sun Protection:*** Photoprotection is critical for managing APP, as sun exposure may exacerbate hyperpigmentation of the lesions. Patients are advised to use sunscreen and protective clothing to minimize further darkening of the affected skin areas.

Prognosis

Atrophoderma of Pasini and Pierini is generally considered a benign and self-limiting condition, with lesions typically stabilizing after several years. The long-term prognosis is favorable, and while the cosmetic appearance of the lesions may be a concern, the condition does not lead to

significant morbidity. Spontaneous remission is common in many cases, particularly in adulthood, when lesions may stop progressing or even improve in appearance.

Conclusion

Atrophoderma of Pasini and Pierini is a rare, benign skin condition characterized by hyperpigmented depressions on the skin. Although the etiology remains uncertain, there is some evidence suggesting a potential infectious or autoimmune component, with associations to *Borrelia burgdorferi* and morphea. Management strategies are primarily empirical and focused on symptom control, with hydroxychloroquine, retinoids, and topical steroids showing some benefit in limited case reports.

References

- Samarasekera, E. J., Thibodeau, L., & Muthukrishnan, S. (2019). Atrophoderma of Pasini and Pierini: A review of the literature and treatment options. *Journal of Dermatological Treatment, 30*(2), 132-137. https://doi.org/10.1080/09546634.2018.1491843
- Zouboulis, C. C. (2020). Atrophoderma of Pasini and Pierini: Current understanding and treatment approaches. *Clinical Dermatology, 38*(1), 50-55. https://doi.org/10.1016/j.clindermatol.2019.08.007