

## Last Onset Unilateral Pansclerotic Morphea

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### Letter to Editor

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### Letter to the Editor

Pansclerotic morphea is characterized by rapidly progressing sclerosis involving the entire skin, trophic cutaneous ulcers, painful contraction and limited joint mobility [1]. The classification of PM is controversial. Some authors consider it as the most severe variant of linear scleroderma, while other authors classify it as deep morphea due to the involvement of all structures overlying the bone [2]. It is distinguished from systemic scleroderma by the absence of visceral and vascular involvement [3]. No reliably effective treatment has yet been established. The disease is generally resistant to usual treatments. The prognosis is poor since the disease has a major functional, aesthetic damage and potentially fatal outcome [1]. We report a new observation of this unusual disease.

A 76-year-old patient, with three years' history of benign prostatic hyperplasia. He was referred by her urologist for management of an in duration and hyperpigmentation, evolving since 6 months, of the right hand. Within a few months of onset, painful contracture deformities of the hand, joints developed, impairing the patient's normal daily activities. Otherwise patient did not report any history of trauma in childhood, raynaud phenomenon or other systemic signs. Clinical examination revealed a sclerotic hyperpigmented cup of the hand and the right forearm with amyotrophy and a pudgy aspect of the fingers [Figure 1,2]. The diagnosis of deep morphea was confirmed by skin biopsy. The biological assessment was normal. Magnetic resonance imaging did not show joint damage. The patient was put on corticotherapy bolus for 3 days and then oral relay (0.5 mg / kg / day) combined with methotrexate (15 mg /

week) with functional rehabilitation sessions. The decline is 2 months with a clear improvement in sclerosis and stiffness.



Figure 1: A sclerotic hyperpigmented cup of the hand and the right forearm with amyotrophy and a pudgy aspect of the fingers.



Figure 2: Asymmetry of both hands.

Morphea or localized scleroderma, is characterized by sclerosis of the skin and subcutaneous tissues. The recent classification individualizes 5 subtypes of localized scleroderma: plate, linear, generalized, mixed and pansclerotic [1]. Pansclerotic morphea is a rare, severe, and mutilating subtype of localized scleroderma characterized by severe course with generalized full-thickness skin involvement and possible growth and functional impairment [4]. The prevalence of this Pansclerotic morphea in adults has never been estimated [5]. Genetic background with predisposition to autoimmune disorders, immunologic alterations and environmental factors such as trauma, toxins exposure or infections (especially infection with *Borrelia burgdorferi*) are considered to play a role in the development of morphea [6]. Although the etiology is unclear, T lymphocytes, Langerhans cells and natural killer cells are thought to be pathogenic in localized scleroderma [5]. PM is usually seen before the age of 14 years with the patient complaining of arthralgia and stiffness at the time of onset [3]. It is a chronic disease which manifests clinically by initially inflammatory, violaceous plaques, that later become indurate and atrophic. The fibrotic process may progress to different depths, involving the dermis, the subcutaneous fat, and sometimes even the underlying soft tissues and bone [5,6]. In fact, the disease has a wide clinical spectrum, ranging from mild hyperpigmented plaques to severe, invalidating generalized and pansclerotic forms [6]. It is important to differentiate between the aggressive form of localized scleroderma and progressive systemic sclerosis. scleroderma spares the central back, whereas Pansclerotic morphea can involve the whole back, tends to be less intense in the upper pectoral area and spares the nipples and periareolar skin [5]. However, the presence of sclerodactyly, with characteristic sparing of the tip of the fingers and absence of Raynaud phenomenon, are findings that help us to differentiate between the two diseases [3]. The most common complications observed in DPM patients are generalization of cutaneous sclerosis; irreversible ankylosis; development of cutaneous trophic ulcers with superinfection; neurocompressive syndromes; development of squamous cell carcinoma and transformation into systemic sclerosis [3]. To date, there is no standard strategy of treatment, as no therapy has been consistently proven to be efficient in stopping the

progression of the disease [6]. PM treatment comprises a combination of immunosuppressive agents (such as corticosteroids, methotrexate, mycophenolate mofetil), PUVA, anti-thymocyte globulin and biological agents used in off-label [4].

This form of scleroderma is poorly known and sometimes confused with systemic scleroderma. It is individualized by its rapidly aggressive evolution and damage to the skin and deep structures causing contractures and trophic disorders complicated by superinfection and squamous cell carcinoma. It is important to report all cases of this rare and very serious form, including cases of therapeutic resistance and fatal outcome, to improve knowledge about the disease.

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