Open Access Article

Ann. Acad. Med. Siles. (online) 2025; 79: 8–11
eISSN 1734-025X
DOI: 10.18794/aams/196560
www.annales.sum.edu.pl

OPIS PRZYPADKU
CASE REPORT

Morphea profunda – a case report of deep localized scleroderma with severe thorax deformity

Twardzina głęboka – opis przypadku ze znaczną deformacją klatki piersiowej

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ABSTRACT

Deep scleroderma is a rare form of limited scleroderma characterised by deep sclerosis that may involve the muscles, fascia, subcutaneous tissue and deep layers of the skin. The lesions usually occur in the paraspinal line and may be predisposed by factors such as infections, injuries, exposure to radiation or the use of stimulants. Due to an insufficient level of awareness among healthcare professionals, diagnosis can be significantly delayed, and irreversible as well as crippling deformities can result. The study presents the case of a 67-year-old female patient with deep scleroderma whose lesions occur in an unusual location on the anterior surface of the thorax. This case demonstrates the importance of early diagnosis and the introduction of appropriate therapy in the active phase of the disease to avoid such severe consequences.

KEYWORDS

localized scleroderma, deep localized scleroderma, connective tissue

Received: 17.09.2024 **Revised:** 08.10.2024 **Accepted:** 27.11.2024 **Published online:** 08.01.2025

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Publisher: Medical University of Silesia, Katowice, Poland

STRESZCZENIE

Twardzina głęboka jest rzadką formą twardziny ograniczonej, charakteryzującą się występowaniem głębokich stwardnień, mogących obejmować mięśnie, powięzi, tkankę podskórną oraz głębokie warstwy skóry. Zmiany zwykle występują w linii przykręgosłupowej, a do ich powstawania mogą predysponować czynniki wyzwalające, takie jak infekcje, urazy, ekspozycja na promieniowanie lub stosowanie używek. Ze względu na niedostateczny poziom świadomości pracowników opieki zdrowotnej diagnoza może być znacznie opóźniona, a w konsekwencji może dochodzić do nieodwracalnych i okaleczających deformacji. W pracy przedstawiono opis przypadku 67-letniej pacjentki z twardziną głęboką, u której zmiany chorobowe występują w nietypowej lokalizacji w obrębie przedniej powierzchni klatki piersiowej. Przypadek ukazuje, jak ważne jest wczesne postawienie właściwej diagnozy oraz wprowadzenie odpowiedniej terapii w aktywnej fazie choroby, aby uniknąć tak poważnych powikłań.

SŁOWA KLUCZOWE

twardzina ograniczona, twardzina głęboka, tkanka łączna

INTRODUCTION

Deep scleroderma is a rare form of localized scleroderma (LoSc, morphea; < 5%) characterized by the presence of deep sclerotic lesions affecting the muscles, fasciae, subcutaneous tissue and deeper layers of the dermis [1]. Infection, injury, radiation or drugs can potentially trigger the disease [2]. It may affect both children and adults. Skin manifestations are not accompanied by any subjective symptoms or the involvement of internal organs

CASE DESCRIPTION

A 67-year-old woman with a history of morphea profunda presented to the dermatology department with multiple diffuse sclerotic changes involving the characteristic of hardened tumors, poorly demarcated and overstretched scars, constant inflammation and discoloration in the chest region (Figure 1). On cutaneous examination in the right breast area, there was an erythematous infiltrative lesion present with a small crust on top, painful at palpation (Figure 2). Medial within the right breast there was a fistula, slightly painful at palpation. The nipples were free of lesions. Magnetic resonance imaging (MRI) showed the connective tissue changes of the characteristic of fibrous changes located in the skin and subcutaneous tissue of the anterior chest wall, which is typical for scleroderma. Computed tomography revealed sclerotic infiltration of the skin of both breasts, post-inflammatory fibrous changes with accompanying minor calcifications at the apices of the lungs, enlargement of the lymph nodes in the right axilla and Schmorl's nodes in the lower thoracic vertebrae. The symptoms were first observed in 1984; initially the differential diagnosis included breast cancer and panniculitis non febrilis, however, in 1995 a biopsy was performed and confirmed the diagnosis of morphea profunda. Over the years, the patient has been treated in various ways. Her medical history included medications such as thymostimulinum (TFX), Augmentin, dapsone, isotretinoin, ciprofloxacin, ceftriaxone, a eutectic mixture of local anesthetics (EMLA), cefuroxime, tobramycin ointment, cooling ointment with hydrocortisone and periodic topical mupirocin. The patient had concomitant diseases including chronic obstructive pulmonary disease (COPD), cardiac arrhythmia and a gallbladder polyp. The laboratory results only demonstrated a high cholesterol level (8.3 mm/l) and white blood cells in the upper limit of the norm (10 000/µl). *Staphylococcus epidermidis* was identified in the swab. The patient was started on Cefuroxime 2 × 1.5 g intravenously, probiotic and cold compresses with wound disinfection solution were applied locally. The treatment recommendations after hospitalization included methotrexate (MTX) 15–25 mg/week, prednisone 0.5–1 mg/kg body weight/day in 2 divided into doses for 2–4 weeks with a gradual dose reduction, and mycophenolate mofetil (MMF) 1–2 g/day.

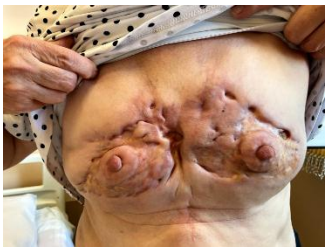

Fig. 1. Skin lesions in form of hardened tumors. Poorly demarcated, overstretched scars. Skin is discoloured, covered by chronic inflammatory process. Nipples are free of lesions.

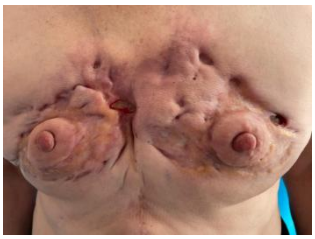

Fig. 2. In region of right breast, erythematous and infiltrative lesion with crust on top and fistula at sternum.

DISCUSSION

Limited scleroderma is a chronic connective tissue disease. The peak incidence is between the ages of 20 and 40 [1]. The disease is more common in Caucasians and females (female:male – 4:2) [3]. The etiopathogenesis of LoSc is not fully understood, but genetic and environmental factors, as well as immunological and vascular disorders, are thought to be crucial in the development of the condition [1].

There are known cases that LoSc-type lesions occurred due to mechanical injury, long-term pressure, and the usage of drugs. The role of *Borrelia burgdorferi* spirochetes in the development of LoSc is debatable, but currently it is not recommended to routinely determine the level of anti-*B. burgdorferi* antibodies [4].

Clinically, the disease involves three phases: early inflammatory (active, lasting an average of 3–4 years), progressive sclerosis and atrophic (atrophic). Limited scleroderma usually does not progress to systemic scleroderma [1].

According to the latest classification, there are five main clinical types of LoSc – deep scleroderma is one of them. The case we present is an example of a severe deformity in the thorax resulting from a long-standing course of deep scleroderma.

In every patient with deep morphea, a thorough medical history should be taken to determine the onset of lesions, conduct a thorough physical examination, and assess disease activity or severity and tissue damage, as well as the possible progression of lesions. A special questionnaire for assessing activity/severity and tissue damage (Localized Scleroderma Cutaneous Assessment Toll – LoSCAT) is used for this purpose [5]. Investigations in cases of morphea profunda may show peripheral eosinophilia, high gamma-globulinemia and raised erythrocyte sedimentation rate (ESR) [5]. Although serologic abnormalities like antinuclear antibodies, anti-double stranded-DNA, anti-single stranded-DNA, anti-histone antibodies and a rheumatoid factor have been discovered in morphea patients, regular testing for them is not advised [6]. It is also worthwhile to perform imaging studies (especially MRI) [7], which will undoubtedly facilitate the evaluation of the effectiveness of the implemented treatment or the progression of the disease in further follow-up [4]. MRI has its limitations, such as the high cost of the examination, the long examination time, accessibility or the low signal-to-noise ratio for the superficial layers [8]. Deep scleroderma does not involve internal organs, hence diagnosing the patient for organ changes is not required [4].

Excised skin should be taken for histopathological examination only in doubtful situations with an unclear clinical picture, in order to establish the diagnosis. Deep scleroderma should be differentiated from subcutaneous tissue inflammation and the subcutaneous (deep) variety of lupus erythematosus [4].

If morphea profunda is diagnosed, aggressive treatment is required in the active phase of the disease. The treatment of choice is glucocorticosteroids in the form of intravenous infusions of methylprednisolone at a dose of 500–1000 mg/day for 3 days. Treatment should be continued for at least 3–6 months. An alternative to parenteral treatment is oral prednisolone therapy. It is recommended to administer 0.5–2 mg/kg/day of prednisolone for 2–4 weeks, followed by a gradual reduction of the dose. During long-term glucocorticosteroid therapy, patients should be monitored for adverse effects such as osteoporosis [9]. Glucocorticosteroids can be used in monotherapy or in combination with methotrexate. The recommended dose of methotrexate in adults is 12.5–25 mg/week. Treatment should be continued for at least 12 months. Methotrexate intolerance or drug resistance is an indication to consider mycophenolate mofetil (1–2 g/day) [10]. Patients should be screened for hepatitis B and C before starting methotrexate treatment. While taking methotrexate, patients require a complete blood count every three months to monitor for methotrexate toxicity, which can cause cytopenia and elevated liver parameters. Folic acid preparations should also be supplemented during therapy with this drug to prevent side effects [9].

During the inactive period, the morphology of the plaque changes – erythema disappears, hyper-pigmentation occurs, sclerosis subsides and features of atrophy of the skin and/or subcutaneous tissue are present. Such lesions that do not show activity for at least 6 months do not require drug treatment [11].

CONCLUSIONS

The case is reported in view of its extensive morphea profunda with irreversible chest deformity. This case demonstrates the importance of introducing appropriate therapy in the active phase of the disease to avoid such severe complications.

Author's contribution

Study design – N. Tekiela, A. Guberniak, K. Polak, B. Bergler-Czop
Manuscript preparation – N. Tekiela, A. Guberniak K. Polak
Literature research – N. Tekiela, K. Polak, B. Miziołek
Final approval of the version to be published – B. Bergler-Czop, B. Miziołek

REFERENCES

1. Wolska-Gawron K., Krasowska D. Localized scleroderma – classification and tools used for the evaluation of tissue damage and disease activity/severity. Dermatol. Rev./Przegl. Dermatol. 2017; 104(3): 269–289, doi: 10.5114/dr.2017.68775.

2. Cassisa A., Vannucchi M. Morphea profunda with tertiary lymphoid follicles: Description of two cases and review of the literature. Dermatopathology (Basel) 2022; 9(1): 17–22, doi: 10.3390/dermatopathology9010003.

3. Shajil C., Sathishkumar D., Babu J.S., Babu R., Kumar S. Spontaneous crateriform indentation in a child. Pediatr. Dermatol. 2023; 40(4): 715–717, doi: 10.1111/pde.15241.

4. Krasowska D., Rudnicka L., Dańczak-Pazdrowska A., Chodorowska G., Woźniacka A., Lis-Święty A. et al. Localized scleroderma (morphea): Diagnostic and therapeutic recommendations of the Polish Dermatological Society. Dermatol. Rev./Przegl. Dermatol. 2019; 106(4): 333–353, doi: 10.5114/dr.2019.88252.

5. Budamakuntla L., Malvankar D. Extensive morphea profunda with autoantibodies and benign tumors: A rare case report. Indian Dermatol. Online J. 2012; 3(3): 208–210, doi: 10.4103/2229-5178.101823.

6. Anggawirya B.Y., Indramaya D.M., Wardhani P.H., Widia Y., Citrashanty I., Sawitri S. et al. Deep circumscribed morphea: A case report. J. Pak. Assoc. Dermatol. 2024; 34(2): 582–586.

7. Horger M., Fierlbeck G., Kuemmerle-Deschner J., Tzaribachev N., Wehrmann M., Claussen C.D. et al. MRI findings in deep and generalized morphea (localized scleroderma). AJR Am. J. Roentgenol. 2008; 190(1): 32––39, doi: 10.2214/AJR.07.2163.

8. Khorasanizadeh F., Kalantari Y., Etesami I. Role of imaging in morphea assessment: A review of the literature. Skin Res. Technol. 2023; 29(7): e13410, doi: 10.1111/srt.13410.

9. Penmetsa G.K., Sapra A. Morphea. StatPearls, 2023 [online]. Treasure Island (FL): StatPearls Publishing; 2024, https://www.ncbi.nlm.nih.gov/books/NBK559010/ [accessed on 27 November 2024].

10. Wolska-Gawron K., Michalska-Jakubus M., Krasowska D. Localized scleroderma – current treatment options. Dermatol. Rev./Przegl. Dermatol. 2017; 104(6): 606–618, doi: 10.5114/dr.2017.71833.

11. Florez-Pollack S., Kunzler E., Jacobe H.T. Morphea: Current concepts. Clin. Dermatol. 2018; 36(4): 475–486, doi: 10.1016/j.clindermatol.2018.04.005.