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En coup de sabre linear scleroderma – diagnostic difficulties. Case report

Twardzina linijna typu *en coup de sabre* – trudności diagnostyczne. Opis przypadku

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Abstract

Frontal linear scleroderma (morphea en coup de sabre) is a rare disease from the group of limited connective tissue diseases, most often affecting children. Due to the fact that it is not a common dermatosis, diagnosis and treatment may be difficult. Therefore, we believe it is advisable to present the diagnostic difficulties in a patient of the Provincial Integrated Hospital in Elbląg. It is also noteworthy that the patient's neurological symptoms preceded the appearance of skin lesions. Clinical symptoms and imaging studies led us to observe progressive hemifacial atrophy. The patient requires interdisciplinary specialist care and diagnosis in the field of neurology, dermatology, rheumatology and ophthalmology.

Keywords: linear scleroderma, saber cut, morphea en coup de sabre

Streszczenie

Twardzina typu cięcia szablą (*morphea en coup de sabre*) jest rzadko występującą chorobą z kręgu ograniczonych schorzeń tkanki łącznej, pojawiającą się najczęściej w dzieciństwie. Jako że nie jest to częsta dermatoza, jej rozpoznanie i leczenie mogą sprawiać trudności, dlatego autorki uznały za celowe przedstawienie trudności diagnostycznych w przypadku pacjenta Wojewódzkiego Szpitala Zespolonego w Elblągu. Na uwagę zasługuje też fakt, że u opisywanego chorego wystąpienie zmian skórnych poprzedziły problemy neurologiczne. Objawy kliniczne i badania obrazowe skłaniają do obserwacji pacjenta w kierunku postępującego połowiczego zaniku twarzy. Chory wymaga interdyscyplinarnej specjalistycznej opieki i diagnostyki w zakresie neurologii, dermatologii, reumatologii, a także okulistyki.

Słowa kluczowe: twardzina linijna, typu cięcia szablą, *morphea en coup de sabre*

INTRODUCTION

Frontal linear scleroderma (morphea en coup de sabre) is a rare disease from the group of limited connective tissue diseases, occurring most often in children and females. The incidence is estimated at less than 3 per 100,000. The clinical picture initially presents as a unilateral, erythematous plaque, which then transforms into a discoloured, deep furrow involving not only the skin, but also the subcutaneous tissue, muscles and bones. Due to its rarity and the coexistence of cutaneous, neurological and ophthalmological symptoms, scleroderma en coup de sabre poses a diagnostic challenge. It is an interdisciplinary problem that requires the cooperation of doctors of many specialties.

CASE REPORT

A 23-year-old patient was admitted to the Department of Dermatology of the Provincial Integrated Hospital in Elbląg with suspicion of en coup de sabre linear scleroderma (saber cuts, ECDS). During medical interview, he reported the appearance of skin discoloration changes in the left frontal area about 6 years ago. So far diagnosed mainly neurologically, without making a final diagnosis. Dermatologically, the diagnosis of a verrucous nevus or a post-traumatic lesion was initially suggested (the patient denied the injury). Physical examination on admission revealed two linear lesions in the form of induration of the skin in the fronto-parietal region, brown in colour with discrete translucency of blood vessels. The patient initially noticed a red streak on the left side of his forehead, which over time spread to the scalp (Fig. 1). With the passage of time, the lesion darkened, and atrophy of the frontal belly of the fronto-occipital muscle developed within it. Atrophy of soft tissues and bones with visible asymmetry of eyeballs and superciliary arches was also observed. The patient had no cicatricial alopecia on the scalp. Histopathological examination of the skin sample collected in 2019 showed signs of increased fibrosis with the presence of preserved single skin appendages, without signs of inflammation. The patient has not used topical treatment so far. He is permanently on levetiracetam for seizures. Allergies, sudden weight loss, fever, mucosal dryness, deterioration of exercise tolerance, and swallowing disorders were denied by the patient. Family history of autoimmune diseases and skin diseases was negative. Despite the lack of visual disturbances, an initial posterior subcapsular cataract was found in the ophthalmological examination. Laboratory tests revealed normocytic anaemia (Hgb 13 g/dL), hyperuricaemia (uric acid 7.52 mg/dL), and vitamin D deficiency (vitamin 25-(OH) D 11.10 ng/mL). Abdominal and retroperitoneal ultrasound, high resolution computer tomography of the chest, X-ray of the hands and ECHO showed no abnormalities. Capillaroscopy showed single dilated capillaries of the fifth finger of the left and right hand, as well as abnormal morphology and microextravasations.

In recent years, the patient has reported attacks of headaches, numbness and hyperaesthesia of the right upper limb, as well as episodes of poor concentration and memory. In January 2019, he experienced the first incident of convulsive seizure with turning of the eyeballs and biting the tongue, with complete amnesia of the event. At that time, the first magnetic resonance imaging (MRI) of the brain was performed along with a stereotactic biopsy, after which nodular arteritis was suspected, which was then treated with systemic steroid therapy. Head MRI performed in March 2023 showed a T2 and FLAIR hyperintense area of approximately 54/14 mm in the white matter of the posterior part of the frontal lobe and the parietal lobe on the left side, which was already visible in previous imaging studies. Numerous foci of signal attenuation in the hemo sequence were visible within the left hemisphere of the brain. After administration of the contrast agent, the cortical vessels of the left hemisphere, especially the posterior frontal, parietal and occipital lobes, appeared more numerous, slightly wider than in the corresponding regions of the right hemisphere, with a reduced fluid space of the parabrain. On this basis, angiopathy in the course of localized scleroderma was suspected. During the diagnosis, tests were performed for Lyme disease, viruses: cytomegalovirus (CMV), Epstein–Barr virus (EBV), herpes simplex virus (HSV), varicella zoster virus (VZV), human herpesvirus 6 and 7 (HHV-6, -7),

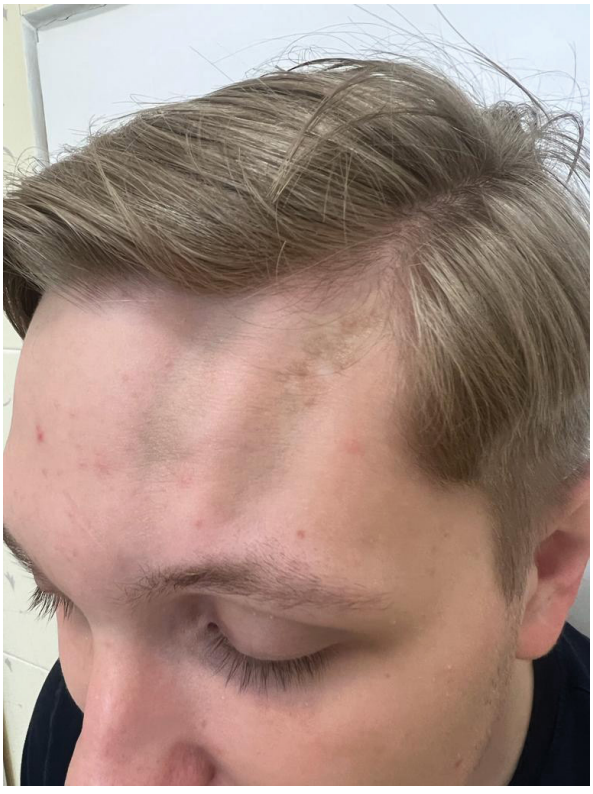

Fig. 1. A patient presenting with frontal linear scleroderma (morphea en coup de sabre)

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B19, adenoviruses, enteroviruses, parechoviruses and anti-NMDA, anti-neutrophil cytoplasmic antibodies (ANCA), α-amino-3-hydroxy-5-methyl-4-isoxazolepropionic acid 1 and 2 (AMPA 1, 2), contactin-associated protein-like 2 (CASPR2), leucine-rich glioma-inactivated 1 (LGI1) antibodies, gamma-aminobutyric acid B (GABA B), and were negative. During hospitalisation in the Department of Rheumatology, a dermatological consultation was held, during which systemic treatment with methotrexate and local treatment with tacrolimus 0.1% ointment twice a day were recommended. In the Department of Rheumatology, pulses of methylprednisolone in a dose of 500 mg were administered intravenously for 3 days, and methotrexate in a dose of 15 mg together with folic acid in a dose of 5 mg was started. Continuation of systemic steroid therapy in the form of oral prednisone 10 mg and vitamin D supplementation were recommended. The patient remains under the supervision of the Rheumatology and Dermatology Clinic with good treatment tolerance.

DISCUSSION

Frontal linear scleroderma (morphea en coup de sabre) is a rare disease from the group of limited connective tissue diseases, which is most prevalent in children and females[1–3]. The incidence is estimated at less than 3 per 100,000[4]. Although the aetiology of limited scleroderma is not fully understood, it has been observed that the disease can be provoked by certain factors, including injections, injuries, mechanical compression, and infection with *Borrelia burgdorferi*[5,6]. In the clinical picture, a unilateral, erythematous plaque is initially observed, which then transforms into a discoloured, deep furrow. Over time, it covers not only the skin, but also involves the subcutaneous tissue, muscles and bones[1,7,8]. The lesion initially begins at the level of the superciliary arch and reaches the fronto-parietal area, where the focus of scarring alopecia usually appears. Bilateral linear scleroderma has been described less frequently in the literature[3–5]. There are known case reports of patients with spreading lesions below the eyebrow line, involving the eyelids, eyelashes or the skin of the nose[9]. Morphea en coup de sabre presents an increased risk of central nervous system and ocular involvement. This is a rare complication that may manifest as episodes of migraine, epilepsy, trigeminal neuralgia, vascular malformations, uveitis, or eyelid distortion. MRI abnormalities have also been reported, even in patients without concomitant neurological disorders[5,8,10]. Cases of maxillary and mandibular involvement with tooth root deformation and atrophy have also been described[1,2].

En coup de sabre may coexist in 20–40% of patients with progressive hemifacial atrophy (Parry–Romberg syndrome), which involves the skin, subcutaneous tissue, muscles and bones of half the face. The disease can occur at any age, but most often affects children under 10 years of age. Hemifacial atrophy may be accompanied by neurological symptoms in 20% and by ophthalmic symptoms (enophthalmia, strabismus, eyelid atrophy, uveitis, glaucoma, diplopia, Horner's syndrome, mydriasis) in 15% of cases[11]. Subcutaneous or oral methotrexate (MTX) at a dose of 15–25 mg/week in adults and 15 mg/m^2/week in children, and glucocorticoids (GCS), i.e. oral prednisone at 0.5–1.0 mg/kg body weight per day for 2–4 weeks or methylprednisolone in the form of intravenous pulses at 30 mg/kg per day, up to a maximum of 1,000 mg/day for 3 consecutive days per month for 3–6 months, are the treatment of choice. In the case of first-line treatment failure, the treatment may be supplemented with or switched to mycophenolate mofetil (MMF) at a dose of 1–2 g/day[1]. The probable mechanism of action of methotrexate in the treatment of scleroderma is to influence the expression of cytokines. Decreased levels of soluble circulating interleukin 2 receptors, decreased serum levels of interleukin 6 and 8 have been reported as a result of methotrexate treatment in both juvenile and adult rheumatoid arthritis patients, while elevated levels of the above-mentioned cytokines are associated with the active phase of scleroderma[12].

In a 2021 meta-analysis summarising 34 articles assessing treatment outcomes in patients with morphea en coup de sabre, methotrexate was concluded as the most extensively studied drug with the highest 100% rates of positive responses to treatment in children and adults. In addition, cases of positive responses to treatment with systemic GCS, ultraviolet A1 (UVA1) phototherapy, narrow-band UVB (NB-UVB), psoralen ultra-violet A (PUVA) therapy, mycophenolate mofetil, hydroxychloroquine, abatacept, tocilizumab, cyclosporine A, gamma interferon, and pulsed dye laser (PDL) were reported[9].

CONCLUSIONS

Frontal linear scleroderma typically occurs in children and females, which may have made the diagnosis difficult in this male patient, who developed first symptoms in early adulthood. All patients with head and neck morphea should be closely monitored for central nervous system complications and ophthalmic abnormalities.

Due to the asymmetry of the size of the eyeballs and central nervous system (CNS) involvement, the described patient requires further follow-up for Parry–Romberg syndrome. Our patient is a rare case, where neurological symptoms and diagnosed changes in the nervous system preceded the correct diagnosis of en coup de sabre. Scalp lesions are most commonly seen on the same side as CNS lesions, and thinning of the cortical skull bone should allow for a correct diagnosis.

Due to the coexistence of cutaneous, neurological and ophthalmological symptoms, scleroderma en coup to sabre is an interdisciplinary problem that requires the cooperation of dermatologists, rheumatologists, neurologists, ophthalmologists and paediatricians.

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Conflict of interest

The authors do not declare any financial or personal links to other persons or organisations that could adversely affect the content of this publication or claim rights thereto.

References

1. Szymoniak-Lipska M, Cieplewicz P, Polańska A et al.: En coup de sabre – a case report of therapeutic difficulties. J Face Aesthet 2020; 3: 139–144.
2. Stępień K, Reich A: [Linear morphea *en coup de sabre*]. Forum Derm 2018; 4: 153–154.
3. Vargas-Mora P, Orlandi D, Aranibar L et al.: Bilateral morphea *en coup de sabre*. A rare presentation. Indian J Dermatol 2021; 66: 446.
4. Kashyape P, D'Souza AP, Fathalla B: En coup de sabre presenting as status epilepticus. Clin Rheumatol 2020; 39: 3885–3886.
5. Khan HQ, Adil M, Amin SS et al.: Bilateral morphea *en coup de sabre*: a rare presentation of linear morphea. Przegl Dermatol 2021; 108: 59–66.
6. Rudnicka L, Olszewska M, Rakowska A et al. (eds.): Współczesna dermatologia. Vol. 1, PZWL, Warszawa 2022: 307.
7. Bolognia JL, Schaffer JV, Cerroni L et al.: Dermatology. 4th ed., vol. 1, Elsevier, 2018: 711.
8. Ulc E, Rudnicka L, Waśkiel-Burnat A et al.: Therapeutic and reconstructive management options in scleroderma (morphea) en coup de sabre in children and adults. A systematic literature review. J Clin Med 2021; 10: 4517.
9. Mears KA, Servat JJ, Black EH: Linear scleroderma en coup de sabre affecting the upper eyelid and lashes. Graefes Arch Clin Exp Ophthalmol 2012; 250: 1097–1099.
10. Amaral TN, Peres FA, Lapa AT et al.: Neurologic involvement in scleroderma: a systematic review. Semin Arthritis Rheum 2013; 43: 335–347.
11. Krasowska D, Rudnicka L, Dańczak-Pazdrowska A et al.: Twardzina ograniczona (*morphea*). Rekomendacje diagnostyczno-terapeutyczne Polskiego Towarzystwa Dermatologicznego. Przegl Dermatol 2019; 106: 333–353.
12. Platsidaki E, Tzanetakou V, Kouris A et al.: Methotrexate: an effective monotherapy for refractory generalized morphea. Clin Cosmet Investig Dermatol 2017; 10: 165–169.