*Case Report*
*Olgu Sunumu*

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# Scleredema of Buschke in a little child and its successful treatment with ultraviolet A1 phototherapy

## *Küçük bir çocukta Buschke'nin sklerödemi ve ultraviyole A1 fototerapisi ile başarılı tedavisi*

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### Abstract

A 5-year-old girl with skin hardness and thickening was admitted to our clinic and diagnosed with scleredema of Buschke clinically and histopathologically. Ultraviolet A1 (UVA1) phototherapy at a dose of 30 J/cm$^2$ was planned for 3 days in 1 week. After 45 UVA1 phototherapy sessions (with a cumulative dose of 1350 J/cm$^2$), clinical complete remission was achieved. Ultrasonography revealed improvement in skin thickening, and no recurrence was noted in the 2-year follow-up period.
**Keywords:** Scleredema of Buschke, UVA1, phototherapy, scleredema, scleredema adultorum, ultrasonography

### Öz

Beş yaşında kadın hasta, deride sertlik ve kalınlık şikayeti ile kliniğimize başvurdu. Klinik ve histopatolojik bulgular ile Buschke'nin sklerödemi tanısı kondu. Haftada 3 gün 30 j/cm$^2$ ultraviyole A1 (UVA1) fototerapisi planlandı. Kırk beş seans UVA1 fototerapisinden sonra (kümülatif doz 1,350 j/cm$^2$ ile) klinik tam remisyon sağlandı. Ultrasonografi ile deri kalınlığında iyileşme gözlendi ve 2 yıllık takipte nüks izlenmedi.
**Anahtar Kelimeler:** Buschke'nin sklerödemi, UVA1, fototerapi, skleredema, sklerema adultorum, ultrasonografi

## Introduction

Scleredema of Buschke (SB) is a rare connective tissue disease and is characterized by skin induration. It typically starts from the neck and spreads to the upper half of the trunk, shoulders, back, and face[1]. It was first described by Curzio in 1752, but was named after Abraham Buschke, who reported a 46-year-old male patient who presented with skin thickening and hardening after an influenza epidemic in 1902[2].

SB has three types. Type 1 occurs following infectious diseases and tends to improve spontaneously. Type 2 is more persistent and associated with paraproteinemia, not with infectious diseases. Type 3 occurs in patients with diabetes mellitus and has a male predominance, unlike other types, and it is also called scleredema diabeticorum[3].

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Patients with SB have abnormal extracellular matrix gene expression of fibroblasts, and the accumulation of collagen and aminoglycans in the dermis is the main disease mechanism[4]. Immunosuppressive agents, antibiotics, corticosteroids, radiotherapy, and phototherapy are treatment options, but with unsuccessful results[1]. Possible treatment options are limited, especially in pediatric patients owing to their negative effects and side effects on growth and development. Herein, we present a child with SB treated with ultraviolet A1 (UVA1) phototherapy. In this report, we aimed to emphasize the effectiveness of UVA1 phototherapy, which is a safe and successful treatment method in the treatment of SB, especially in children.

## Case Report

A 5-year-old girl presented to our outpatient clinic because of skin hardness and thickening for 4 months. She had no history of any additional disease. A dermatological examination revealed woody hardness and skin thickening predominantly on her face and upper and lower extremities (Figure 1a-c). Laboratory tests were not remarkable. A 4 mm punch biopsy was performed, and histopathological examination revealed loss of hair follicles in the dermis, enlarged collagen bundles (Figure 2a, b), and mucin deposition between collagen fibers (Figure 2c). She was diagnosed with SB based on the clinical and histopathological findings.

Before treatment, skin thickness was evaluated by ultrasonography (Figure 3a, c). UVA1 phototherapy was planned for 3 days in 1 week. A dose of 30 J/cm$^2$ UVA1 was applied for each treatment session. After 45 sessions of UVA1 phototherapy (with a cumulative dose of 1350 J/cm$^2$), clinical complete remission was achieved, and treatment was concluded. Ultrasonography revealed improvement of the skin thickening (Figure 3b, d). No recurrence was noted in the 2-year follow-up period.

The UVA1 irradiation equipment consisted of a Waldmann 7001 K cabin with Waldmann TL10R low-pressure lamps (Waldmann Gmblt, Schvenningen, Germany). These lamps generate UVA1 wavelengths in the 340-400 nm range. Infrared irradiation is also emitted, but this is filtered by an acrylic glass screen. The UVA1 irradiation level is approximately 35 mW/cm$^2$. A dose of 30 J/cm$^2$ is achieved in approximately 30 min.

## Discussion

UVA1 is a part of the UV spectrum that emits photons with wavelengths of 340-400 nm[5]. The two main sources of UVA1 are fluorescent lamps and metal halides. Fluorescent lamp cells only allow low dose (LD) (10-30 J/cm$^2$) or medium dose (MD) (40-70 J/cm$^2$). High-output, high-dose metal halide sources allow up to (130 J/cm$^2$)[6]. UVA1 phototherapy was reported to be effective in the treatment of inflammatory skin diseases such as atopic dermatitis[7]. Mempel et al.[8] observed that mature collagens 1 and 3 decreased in skin biopsies after UVA1 treatment in 14 patients with atopic dermatitis. In addition, in 1981, Mutzhas et al.[5] showed that UVA wavelengths reached the deep dermis and subcutis. In later studies, UVA1 treatment was thought to be an effective treatment option for connective tissue diseases with sclerosis.

*In vitro* studies in cultured fibroblasts applying UVA1 phototherapy have shown an increase in collagenase production[9,10]. However, UVA1

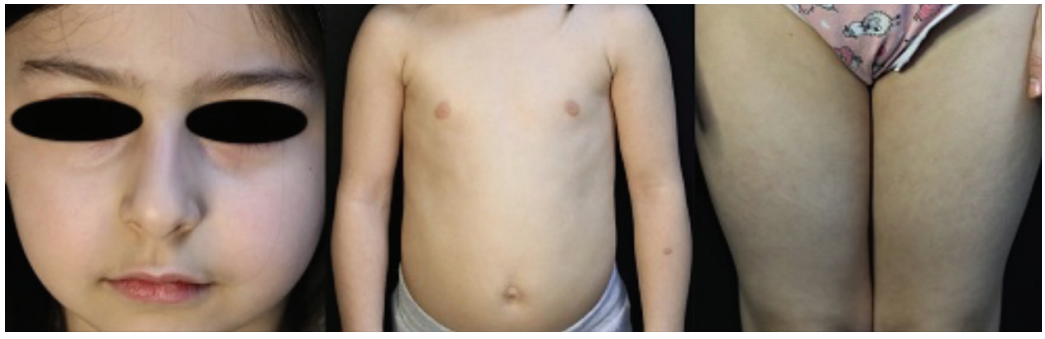

**Figure 1. (a-c)** Woody hardness and thickness of the skin predominantly on extremities

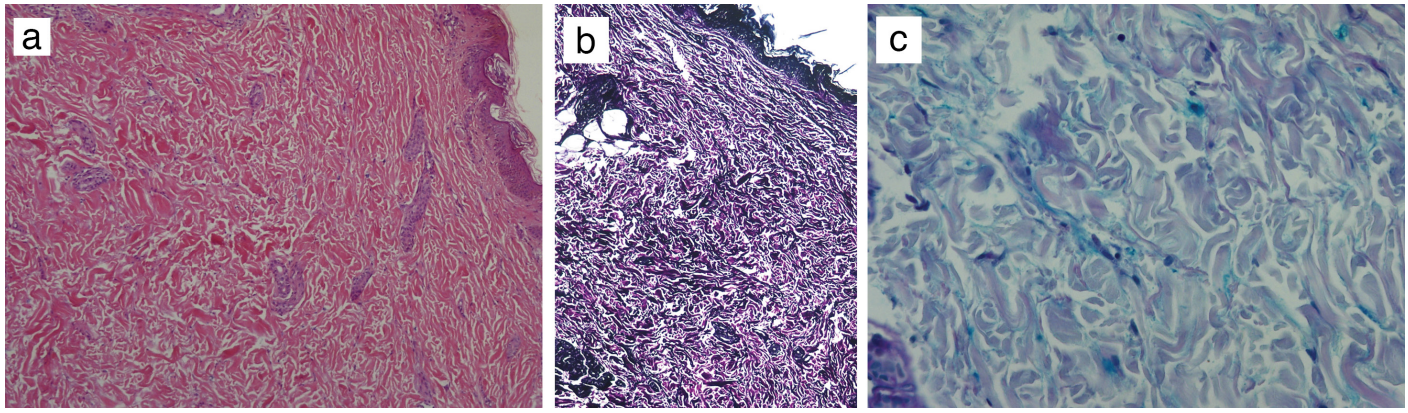


**Figure 2. (a, b)** Loss of hair follicles in the dermis, enlarged collagen bundles (hematoxylin and eosin staining, x10; Verhoeff Von Gieson staining, x10). **(c)** Mucin deposition between collagen fibers (periodic acid Schiff staining, x40)

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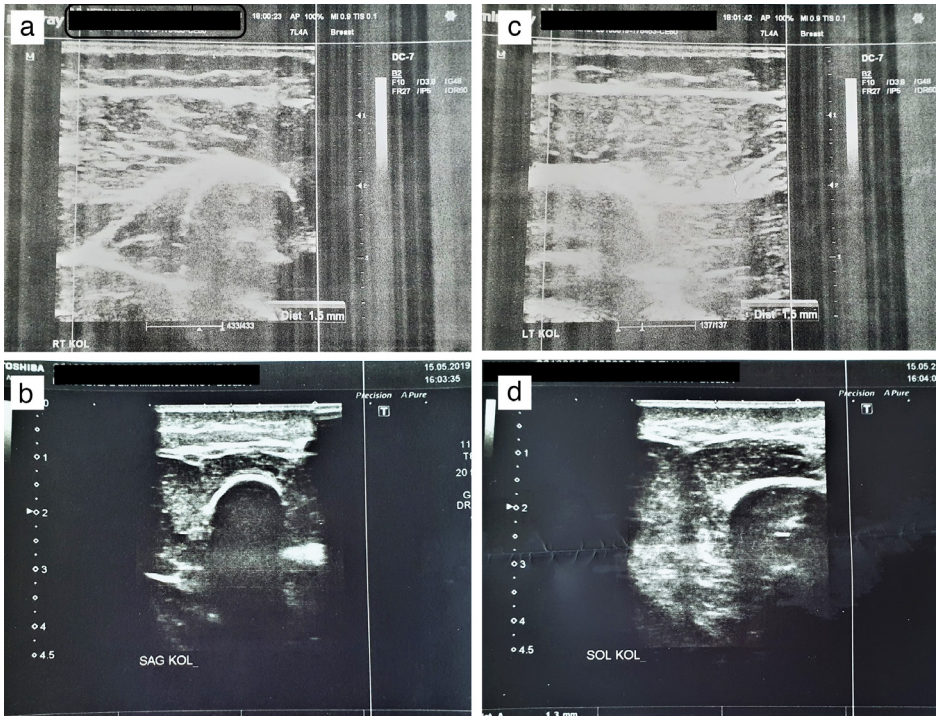


**Figure 3. (a)** Pretreatment ultrasonographic skin thickness of the right arm: 1.5 mm, **(b)** post-treatment ultrasonographic skin thickness of the right arm: 1.2 mm, **(c)** pretreatment ultrasonographic skin thickness of the left arm: 1.5 mm, and **(d)** post-treatment ultrasonographic skin thickness of the left arm: 1.3 mm

reduces the enzyme activity of propyl hydroxylase, which stabilizes the triple helix structure of collagen, and causes disruption of the cross-linking between collagen fibrils[11,12]. In addition, many *in vivo* and *in vitro* studies have shown that UVA increases the number of matrix metalloproteinase (MMP)-specific mRNA receptors in cultured fibroblasts. UVA1 stimulated three members of the MMP family, i.e., MMP1, MMP3, and MMP9, which are responsible for the degradation of type 1 collagen[13]. On the contrary, UVA1 reduces transforming growth factor (TGF)-beta cell surface receptors, which play an important role in procollagen synthesis, growth, and differentiation[14]. Recently, some studies have focused on SMAD, which is a family of transcription factor proteins involved in TGF-beta signaling from the cell surface to the nucleus. *In vitro* and *in vivo* studies have shown that SMAD7 is an important inhibitor of the TGF-beta SMAD pathway and UVA1 stimulates SMAD7[15,16]. Based on these mechanisms of action, UVA1 phototherapy was reported as an effective treatment in different connective tissue diseases with sclerosis.

SB is a chronic skin disease with sclerosis. The effectiveness of UVA1 for the treatment of SB was first reported in 2004[17]. Two patients diagnosed with type 2 and type 3 SB were treated with LD UVA1 phototherapy, and regression was reported in all lesions. In 2005, Eberlein-König et al.[18] reported thinning of the skin following MD UVA1 treatment in a patient diagnosed with type 3 SB. In a study retrospectively evaluating 92 patients with different skin diseases treated with UVA1 in 2006, five had SB, clinical regression was detected in four patients, and treatment was terminated in one patient because of polymorphous light eruption[19]. In our case, pre- and post-treatment ultrasonography findings were also evaluated. After 45 sessions of UVA1 treatment, ultrasonography detected regression of the lesions and significant skin thinning. No treatment-related side effects occurred.

UVA1 phototherapy is an effective and safe treatment method for patients with SB. It may be a treatment option, especially in pediatric patients.

### Ethics

**Informed Consent:** Retrospective study.

**Peer-review:** Externally and internally peer-reviewed.

### Authorship Contributions

Surgical and Medical Practices: D.D., M.G.K., F.C.K., İ.Z., E.Z., Concept: D.D., M.G.K., F.C.K., İ.Z., E.Z., Design: D.D., M.G.K., F.C.K., İ.Z., E.Z., Data Collection or Processing: D.D., M.G.K., F.C.K., İ.Z., E.Z., Analysis or Interpretation: D.D., M.G.K., F.C.K., İ.Z., E.Z., Literature Search: D.D., M.G.K., F.C.K., İ.Z., E.Z., Writing: D.D., M.G.K., F.C.K., İ.Z., E.Z.

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