

## RADIOTHERAPY OF NON-MALIGNANT DISEASES - SCLEREDEMA ADULTORUM BUSCHKE – A CASE REPORT

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### Summary

*Scleredema adultorum Buschke* is a very rare disease characterized by thickening of the dermis of the neck, head, and the upper trunk. The etiology of the disease is still unknown, and it predominantly occurs in young females. The disease is usually self-limiting with no optimal therapy modality for the severe forms of the disease having been yet defined.

A 62-year-old patient presented at our Hospital in 2006 with the already established diagnosis and clinical picture typical for *scleredema adultorum*. After several unsuccessful dermatology treatments, the patient underwent radiation therapy receiving a total irradiation dose of 20 Gy delivered in 10 daily fractions (2 Gy a day). During radiotherapy, and immediately after the therapy, no clinical improvement, or skin induration improvement was observed. A couple of months later, the skin infiltration was partially reduced. However, complete regression did not occur, and this finding remained unchanged to date.

KEY WORDS: *radiotherapy of non-malignant diseases, Scleredema adultorum Buschke*

### RADIOTERAPIJA NEMALIGNIH BOLESTI - SCLEREDEMA ADULTORUM BUSCHKE – PRIKAZ SLUČAJA

#### Sažetak

*Scleredema adultorum Buschke* izrazito je rijetka bolest obilježena zadebljanjem dermisa u području glave, vrata i gornjeg dijela trupa. Etiologija bolesti je nepoznata, a predominantno se pojavljuje u mlađih ženskih osoba. Obično je bolest samo-limitirajuća i do danas još nije definiran jedinstven terapijski modalitet za liječenje ovog poremećaja.

Bolesnik u dobi od 62 godine prvi se puta obratio se u našu Kliniku u 2006.g. s već postavljenom dijagnozom i tipičnom kliničkom slikom *scleredema adultorum*. Nakon višekratnog neuspjeha dermatološke terapije, bolesnik je podvrgnut radioterapiji, te je primio ukupnu iradijacijsku dozu od 20 Gy, podijeljenu u 10 dnevnih frakcija (2 Gy dnevno). Tijekom provođenja radioterapije, kao i neposredno po završenoj terapiji, klinički nije došlo do poboljšanja. Nakon nekoliko mjeseci kožna infiltracija se djelomično smanjila. Međutim, potpuna regresija nije nastupila, a takav nalaz je do danas ostao nepromijenjen.

KLJUČNE RIJEČI: *radioterapija nemalignih bolesti, Scleredema adultorum Buschke*

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### INTRODUCTION

*Scleredema adultorum Buschke* is a rather rare disease (with only some 400 cases reported in the literature to date) characterized by thickening of the dermis of the neck, head, and the upper trunk. The etiology of the disease is still unknown, but it is often preceded by acute febrile infection, and

there is also a known association with diabetes mellitus. It predominantly occurs in young adults, more frequently females. The disease is usually self-limiting (spontaneous partial or complete regression occurs within 18-24 months after the development of first symptoms), however in some patients a tendency to progression has been reported. In such cases, therapy options are rather

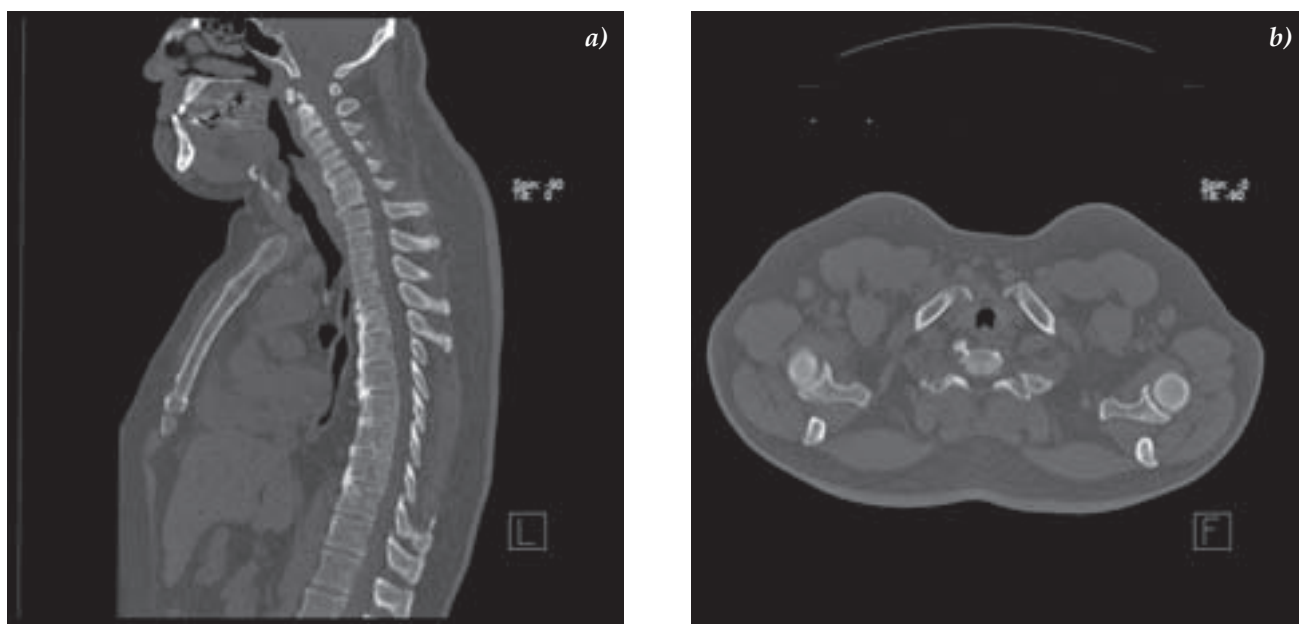


Figure 1.



Figure 2.

poor, with no optimal therapy modality for the severe forms of the disease having been yet defined (1-3).

## CASE REPORT

A male patient, V.F., born in 1944, presented at our Hospital in July 2006 with an already established diagnosis and clinical picture typical for *scleredema adultorum*: horny thickening of the skin in the back of the head, back of the neck, shoulders, interscapular region and along the middle

back. Due to the above changes, the patient had limited mobility of the head, neck and shoulders (especially in performing rotation movements) with a subjective 'tightening' sensation. The patient is a long-term diabetic on insulin therapy, with already developed complications (diabetic foot). After unsuccessful dermatology treatment (with local corticosteroids, keratinolytics, antibiotics, PUVA cream therapy and alike), and an attempt at physical therapy (US therapy to the affected region) he was referred to our Department for favorable response to radiotherapy reported in the scarce literature. Before radiation therapy, the thickness of the back skin (measured by computer tomography) was 12.8 mm (Figure 1a and b), or 4-5 times greater than in a healthy individual.

In the period July 13 - July 27, 2006, the patient received a total irradiation dose of 20 Gy delivered in 10 daily fractions (2 Gy a day) (4). The direct electron beam radiation technique was applied, with electron beam energy of 8 MeV delivered to the affected skin region; the field size in the back and in the neck was 25 x 25 cm and 10 x 10 cm, respectively. During radiotherapy, and immediately after the therapy, no clinical improvement, or skin induration improvement was observed, although the patient reported less discomfort related to the skin tightening.

A year after completing radiotherapy, the patient underwent PUVA cream therapy (15 frac-

tions) again, resulting in a partial reduction of the skin infiltration. However, complete regression did not occur, and this finding remained unchanged to date (Figure 2).

Control examination performed in December 2007 still showed induration and thickening of the skin in both the shoulders and the back, but the patient felt significantly less subjective symptoms.

## CONCLUSION

Although various authors were applying various radiotherapy modalities (beam type and energy, total and daily dose, number of fractions) in the treatment of *scleredema adultorum Buschke*, in our case we adopted the approach of Könemann and colleagues (5) from the University Hospital in Münsteru, Germany. They applied the same radiotherapy modality to treat a patient with the disease of very similar characteristics as we did in our case. In their patient, too, the treatment resulted only in a partial remission of the disease, while the complete remission occurred more than a year after the therapy completion, most probably due to the natural, self-limiting course of the disease.

Regardless of the outcome, the application of ionizing radiation in *scleredema adultorum Buschke* should be taken into consideration as one of important, effective and well tolerable therapy options.

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