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# Giant Spontaneous Hematoma of the Thoracic Wall in a Patient with REST Syndrome

M. BÖHM[1], N. HAAS and B. M. CZARNETZKI

*Department of Dermatology, University Hospital Rudolf Virchow, Free University, Berlin, Germany*

**We report on a 56-year-old patient with systemic sclerosis of one and a half year's duration who was admitted with an acute soft tissue swelling over the left scapula and a marked reduction of joint mobility of the left shoulder. Over the following days, the swelling extended distally and a hemorrhagic discoloration developed, reaching down to the scrotum. Nuclear magnetic resonance imaging revealed a giant intermuscular hematoma between the serratus anterior and the costal muscles. This is the first report of an extensive spontaneous hematoma of the thoracic wall in a patient with progressive systemic sclerosis. Its development and unusual localization may be explained by both the underlying systemic sclerosis and the patient's previous profession as a trapeze artist. Hemorrhagic complications of systemic sclerosis are rare but should be watched for in these patients.** ***Key words: hemorrhage; progressive systemic sclerosis.***

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M. Böhm, Yale University School of Medicine, Dept. of Dermatology, P.O. Box 3333, New Haven, Ct 06510–8059, USA.

## CASE REPORT

A 56-year-old white male, having suffered from REST syndrome (an incomplete variant of the CREST syndrome) for one and a half year, was referred to the dermatology department with an acute painful swelling of the left scapular region which had developed spontaneously. He did not recall any trauma or major physical exertion during the previous days. His systemic sclerosis had previously been treated by his family doctor with prednisolone in a dose-reducing regimen from 100 mg to 15 mg/day. At the first visit to the dermatology clinic, he presented with an orange-sized, non-erythematous, fluctuating swelling over the left scapular region. The joint mobility of the left shoulder was impaired, showing a marked reduction of adduction/abduction (10–0-40°) and anteversion/retroversion (55–0-20°). Leathery, palpable and audible friction rubs were detectable over the shoulder muscles as well as the proximal limbs. The face and neck showed multiple telangiectasis, and the skin on the fingers felt hard and taut. Subsequently, the swelling extended distally, and a hemorrhagic discoloration developed between the distal border of the subscapular swelling and the scrotum (Fig. 1).

Initial laboratory findings showed an elevated white blood cell count of 15800/μl, a decreased red blood cell count ($2.9 \times 10^6$/μl) and a hemoglobin of 9.4 g/dl. Over the following days, there was a progressive drop of hemoglobin to 7.9 g/dl, followed by a slow rise and a normalization of the white blood cell count. Coagulation parameters (including thromboelastogram, bleeding time and Rumpel-Leede test) were normal except for a slight thrombocytosis (418000/μl) and a hyperfibrinogenemia (647 mg/dl). Serum activity of muscle enzymes (creatinine kinase and aldolase) was normal. The following antibody titers were obtained: antinuclear antibody, 1:5120, with a homogeneous fluorescence pattern; anti-histone antibody: positive; antibodies against ds-DNA, ss-DNA, U1-RNP, SSA-ENA, SSB-ENA, SmA-ENA, SCL-70: negative.

Ultrasonography of the scapular region showed an echo-poor structure of 5 cm length and 2 cm breadth under the soft tissue swelling. Nuclear resonance imaging revealed a clearly identifiable intermuscular hematoma between the left serratus anterior muscle and the sceletal thorax. It was located mainly in the subscapular region and extended distally to the 7th rib (Fig. 2). The source of the hemorrhage was not detectable by digital subtraction angiography. Electromyography of the affected shoulder muscles showed regular findings.

A biopsy taken from the region of the hemorrhage showed a massive infiltration of erythrocytes in the dermal and subcutaneous compartment which was more pronounced in the deeper layers. Collagen fibers were thickened and sclerosed. Direct immunofluorescence was negative.

The patient was initially treated with non-steroidal antiphlogistics (diclofenac, 3 × 100 mg) and tilidin for relief of pain. Sterile puncture of the hematoma was not successful, yielding only some clotted blood. The dosage of prednisolone was subsequently increased to 50 mg/day and azathioprine started at a dosage of 100 mg/day. During a period of 4 weeks, the dosage of prednisolone was reduced to 20 mg/day. The hemorrhagic discoloration disappeared by the third week, but the swell-

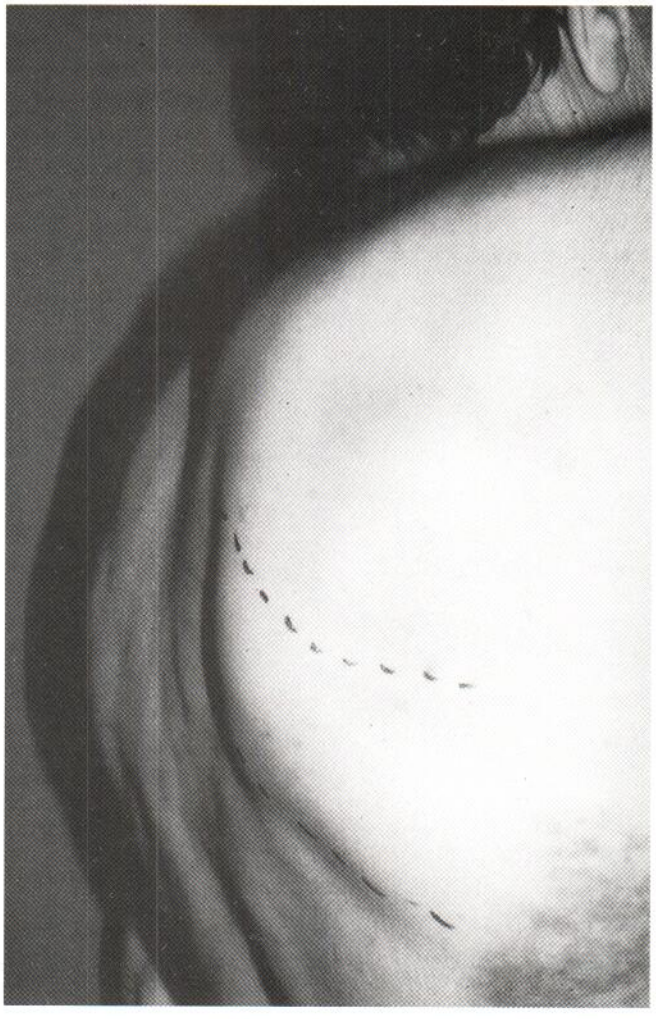

*Fig. 1.* Dorsal view of the giant hematoma; the subscapular swelling was prominent particularly at abduction.



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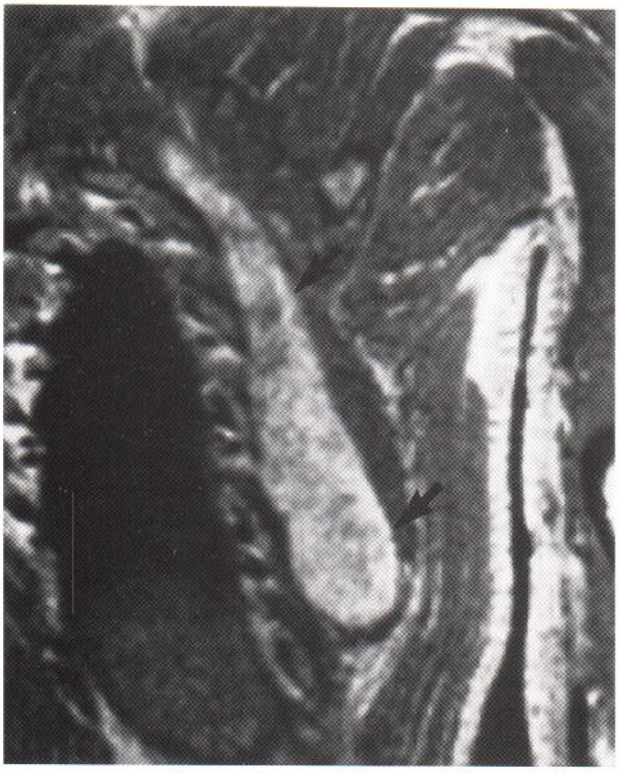

*Fig. 2.* NMR scan of the left lateral thorax; *arrows* indicate the hematoma extending between the subscapular region and the 7th rib.

ing of the scapular and subscapular regions persisted for another 5 weeks.

## DISCUSSION

The CREST syndrome and its incomplete variants REST and CRST are considered to be benign manifestations of progressive systemic sclerosis (PSS). Although cutaneous telangiectasis is a hallmark of the CREST syndrome, bleeding has seldom been reported, in contrast to Osler-Weber-Rendu disease, even when mucous membranes are involved. Gastrointestinal telangiectasis, however, has been reported in a few cases of CREST variants, leading to chronic blood loss and secondary anemia (1–3). In addition, diffuse pulmonary hemorrhage has been described in one case with PSS (4).

To our knowledge, this is the first report of a patient with a CREST variant in whom a giant spontaneous hematoma developed in the thoracic wall. The initial swelling was strictly confined to the scapular region on the day of admission, with subsequent distal migration and final subscapular hemorrhagic discoloration of the left thoracic side. The initial deep scapular swelling might have been due to an erosive injury to vessels in the scapular muscles (infra- and supraspinous muscles and teres minor) which are known to be encapsulated by a robust fascia (5). Hematomas and abscesses in this region can therefore remain localized for a longer period before they penetrate. An exact detection of the bleeding source, however, was not possible in the case presented.

Vascular alterations in muscles affected during the course of PSS include obliterative arteritis of interseptal vessels, perivascular infiltrates or necrotizing vasculitis (6). In our case, electromyography and muscle enzyme tests failed to demonstrate muscular involvement, and the palpable and audible friction rubs are more likely clinical manifestations of a tenositis rather than a myopathy. On the other hand, electromyography is abnormal in only 50% of patients with early disease, and histologic changes are present in only about 40% (7). Therefore, muscular involvement as the causative factor of the spontaneous hematoma cannot be ruled out.

Although the patient denied any trauma or unusual exertion during the days preceding the acute event, the localization and the massive extent of this spontaneous intermuscular hematoma might be related to both PSS and the patient's previous profession as a trapeze artist.

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