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Subcutaneous edema: an "unrecognized" feature of acute polymyositis

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Summary. The case of a man with acute onset of muscle pain, weakness, anasarca, severe dysphagia and dysphonia, and biochemical, electromyographic and histologic evidence of polymyositis is presented. The literature on the occurrence of subcutaneous edema in polymyositis was reviewed. It is concluded that this particular symptom, with no other apparent cause, including heart failure from the underlying disease, is a rare but definite feature of polymyositis itself. A correlation of that with severe bulbar muscle involvement is also suggested.

Key words: Polymyositis – Subcutaneous edema – Bulbar muscle dysfunction

Introduction

Large series of cases of polydermatomyositis [1, 2] do not include subcutaneous edema among the clinical features of the disease, although this had been reported in the initial description of this entity by Wagner in 1877 [3]. A case of generalized subcutaneous edema associated with acute polymyositis and esophageal carcinoma in the French literature [4] and a report of three such cases in the English literature [5] with no underlying malignancy could be found. In two of the latter severe bulbar muscle dysfunction was present. We report here our experience with such an unusual case, in which generalized subcutaneous edema was the predominant initial finding. The etiology of the edema was an enigma for us and an extensive workup to rule out other causes was done. Finally, after the results of the literature search became available, the symptoms were attributed to the underlying rheumatic entity. The purpose of this report is to make clinicians aware of this symptom, which should be included in the disease's features in future reviews.

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Case report

Our patient was a 56-year-old white man, previously healthy, who presented with a 10-day history of arm and leg swelling, arthralgias and proximal muscle weakness and pain. On physical examination

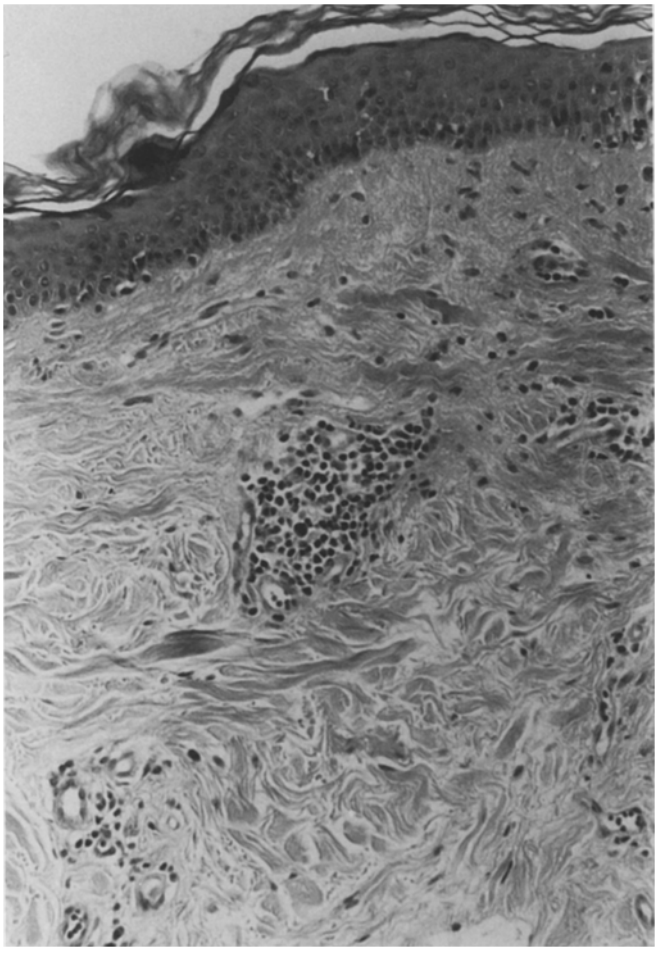

Fig. 1. Skin biopsy shows edema of the dermis, indicated by loose appearance of the tissue, and vasculitis of small blood vessels. HE × 160

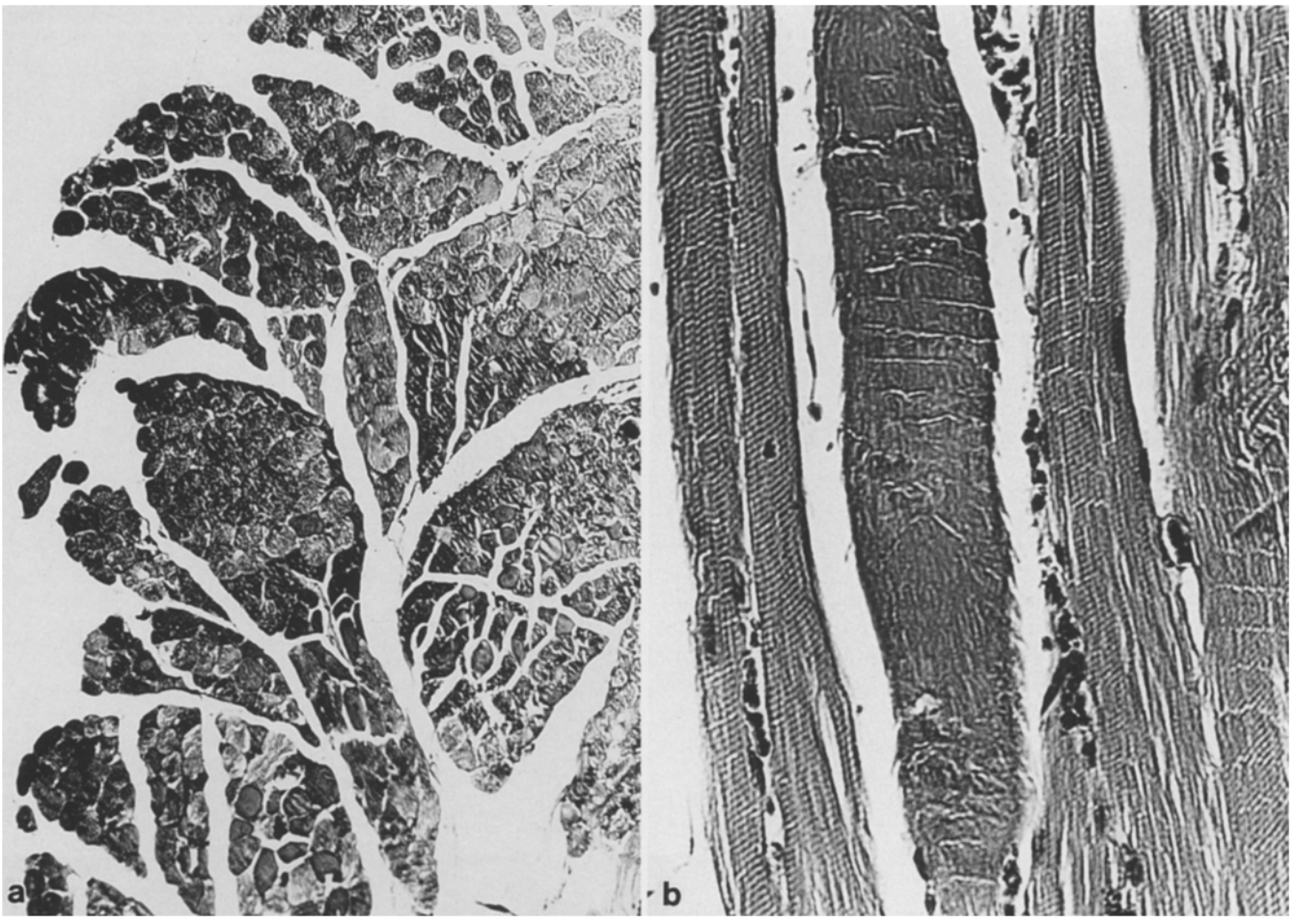

Fig. 2 a, b. Muscle biopsy. **a** A transverse section reveals degenerating and regenerating fibers, the latter characterized by basophilia of the sarcoplasm and a reduction in diameter. HE × 30. **b** A longitudinal section shows areas of degeneration, in which the muscle fiber exhibits loss of transverse striation and fragmentation of the sarcoplasm. HE × 280

he was afebrile with normal vital signs. Positive findings included diffuse pitting edema of the upper and lower extremities including hands and feet, the lumbar, abdominal and posterior thoracic region as well as the chest but not the face, decreased and painful range of motion of both shoulders, and proximal muscle weakness and tenderness.

Laboratory tests included normal complete blood count and differential, urinalysis, blood glucose, blood urea nitrogen, creatinine, alkaline and acid phosphatase, sodium, potassium, calcium, phosphorus and magnesium, uric acid, bilirubin and prothrombin time. Erythrocyte sedimentation rate was 35 mm/h. Serum creatine kinase (CK) was 1300 U (upper normal 300 U) and SGOT and SGPT were 133 U and 50 U respectively (upper normal for both 40 U). Serum albumin was 3 g% and protein electrophoresis showed a slight elevation of alpha$_2$-globulin. A 24 h urine protein was 44 mg. C-reactive protein was normal, while rheumatoid factor, antinuclear antibodies and hepatitis B surface antigen were absent. Chest X-ray, electrocardiogram and echocardiogram were normal. A promptly performed electromyogram revealed a pattern typical for polymyositis. Skin biopsy revealed vasculitis and edema of the dermis (Fig. 1), and muscle biopsy showed polymyositis (Fig. 2).

The patient was placed on 48 mg of methylprednizolone per day, but at the same time an extensive workup to rule out underlying malignancy, quite likely in such a setting, was undertaken. This included a detailed otolaryncologic examination, esophagogastroscopy, thoracic and abdominal CT scans, barium enema, and serum carcinoembryonic antigen and alpha-fetoprotein measurements. The results of all these were normal.

On the above-mentioned regimen serum CK responded favorably and became normal after 20 days, at which time the patient was discharged from hospital. However, the weakness of the proximal muscles persisted, as did the edema. The latter waxed and waned until it finally disappeared 1.5 months after the initiation of steroids. By that time muscle weakness had also decreased somewhat. Interestingly, though, it was then that severe dysfunction of the bulbar muscles, manifested by serious dysphonia and dysphagia for both solid and liquid food, appeared. The scheduled tapering of steroids was cancelled and the patient was hospitalized following an episode of aspiration pneumonia. He was treated with antibiotics and parenteral fluids and the same steroid dose. On this regimen he showed a considerable improvement in both the dysphonia and dysphagia, and gradual steroid tapering was started. Bulbar muscle dysfunction and muscle weakness disappeared after 2 months, as did the arthritis of the shoulders. Today, almost a year after the onset of his symptoms, the patient is in a very satisfactory condition, fully functional, on 6 mg of methylprednizolone every other day.

Discussion

The initial presentation of the reported case posed a serious diagnostic problem. The combination of proximal muscle weakness and elevated muscle enzymes was very

suggestive of polymyositis. This was promptly confirmed by electromyography and muscle biopsy, and appropriate treatment was instituted. However, the etiology of the generalized subcutaneous edema was not clear. Although there was no clinically obvious cause, extensive attempts were made to rule out all common entities that could be responsible and none was identified. The edema, though, waxed and waned for quite some time, on adequate steroid doses, and this posed further questions as to etiology. Although the feeling was that it should have been related to the underlying myositic process, uncertainty remained until the relevant literature became available.

It is interesting that, although subcutaneous edema was included in the initial description of the disease in its acute form by Wagner [3] more than a century ago, this feature has not been mentioned in subsequent large series and textbook chapters [1, 2, 6]. A French report of a case of acute polymyositis associated with esophageal carcinoma described anasarca in 1985 [4]. Before that, in 1982, a British paper described three cases [5]. It is worth noting that two of these three patients died of respiratory failure between 2 weeks and 2 months after the onset of the disease, although neither of them had an underlying malignancy. Of further interest may be the fact that they both had severe bulbar muscle involvement. The cause of death is not mentioned, but aspiration pneumonia and respiratory muscle involvement are possibilities. Interestingly, our patient developed severe dysphonia and dysphagia, to the point of being unable to talk and swallow, followed by aspiration pneumonia, at a time when muscle enzymes had normalized. Fortunately these symptoms did not last long and responded to supportive measures, and the patient is now in remission on small steroid doses.

The purpose of the present report is to emphasize the existence of subcutaneous edema within the clinical spectrum of polymyositis and to sensitize the clinician to this symptom. The mechanism is not clear. We suspect that either it may appear as a reaction to the adjacent muscle inflammation, or it may be secondary to an accompanying vasculitis in the dermis, as in our case. It is known that edema of the subcutaneous tissues may be a symptom of certain vasculitides, such as allergic purpura [7]. In any case, in polymyositis it seems to indicate serious acute disease, likely to be associated with bulbar muscle dysfunction, a feature rather uncommon in polydermatomyositis [1]. It therefore appears reasonable to include subcutaneous edema, no matter how rare it may be, in the clinical picture of polymyositis.

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