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# And what have you got at the end of the day? Private investigations in rheumatology

In this “Rheumatology Forum” issue, we reference the lyrics of Dire Straits’ “Private Investigations” to illustrate the complexities of investigations in rheumatology. Our goal is to discuss several challenging cases presented in this edition, with the hope that they will provide insight and guidance for your investigations in daily rheumatological practice.

It is now well established that patients with rheumatoid arthritis, and more broadly those with autoimmune disorders, often experience multiple comorbidities, which complicate their clinical presentation. Notably, premature atherosclerosis and its associated fatal cardiovascular complications, recognized many years ago, pose a significant risk to these patients. Current understanding indicates that individuals with autoimmune diseases face a 1.5-fold increased risk of cardiovascular complications compared to the general population, as calculated using the Framingham study model. Therefore, there is a continuous need to investigate all clinical factors contributing to the development and progression of atherosclerosis in this specific patient population. In accordance with this perspective, metabolic syndrome, a key contributor to atherosclerosis in the general population, has garnered significant scientific attention in patients with inflammatory arthropathies as well. An exemplary study addressing the role of metabolic syndrome in this context is the paper by Grzechnik et al. [1]. The authors provide a comprehensive and insightful review of the pathophysiological mechanisms underlying the role of metabolic syndrome in disease development and progression, with particular emphasis on lipid profile disturbances. Notably, in contrast

to the general population, normal values of triglycerides, cholesterol, and its subfractions do not necessarily indicate a protective state. High rheumatoid arthritis (RA) activity significantly affects lipid metabolism, and paradoxically, low cholesterol levels in RA patients are often associated with high disease activity. In the conclusions of this paper, the authors assert that the presence of metabolic syndrome (MetS) in patients with RA is associated with increased disease activity, elevated inflammatory markers, functional impairment, and reduced quality of life. The diagnosis of MetS in RA patients appears to represent an additional risk factor for heightened disease activity, underscoring the need for therapeutic interventions aimed at addressing metabolic disturbances [1].

The “Rheumatology Forum” also serves an educational purpose for early-career rheumatologists, bridging the gap between academic textbooks and the continuous influx of new data. As you may recall, the series titled “Everything You Always Wanted to Know About Systemic Sclerosis” garnered significant scientific attention. It is, therefore, unsurprising that a new paper on this enigmatic disease appears in the current issue. The research group from Katowice presents a challenging case of sine syndrome in systemic sclerosis (systemic sclerosis sine scleroderma), which is particularly noteworthy. The patient experienced scleroderma renal crisis without a formal diagnosis of systemic sclerosis, and cutaneous involvement only emerged after kidney damage had occurred. This complex case highlights the importance of recognizing atypical presentations and underscores the need for accurate differential diagnosis, offering valuable in-

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sights for rheumatologists [2]. This aligns well with the preceding paper on scleroderma-like syndromes. The paper emphasizes that while skin hardening is a characteristic feature of systemic sclerosis, it does not necessarily indicate the development of the disease. A broad range of conditions can present with skin induration. The authors highlight the importance of correctly interpreting additional tests, such as antinuclear antibody testing, capillaroscopy, and identifying scleroderma-related manifestations. A current limitation in diagnosing these conditions lies in the absence of specific diagnostic criteria, as the existing criteria are primarily for classification. Strict adherence to these criteria may, in some cases, delay diagnosis. To address this, a critical interpretation of patients' clinical syndromes remains essential for achieving an accurate diagnosis [3].

We continue to gather interesting case reports on challenging conditions to diagnose and treat. An excellent example is the case report on pyoderma gangrenosum (PG), a rare inflammatory skin disease and a representative of the familial group of neutrophilic dermatoses. Moreover, the authors discussed the current diagnostic criteria, which may aid in establishing an accurate diagnosis. Given the rarity of this condition, clinicians should be attentive to nonspecific skin changes that could be associated with autoinflammatory skin disorders [4].

The case report section of this "Rheumatology Forum" issue also includes a report on eosinophilic fasciitis, which aligns well with the comprehensive review on scleroderma-like syndromes published in this issue. Unlike in systemic sclerosis, histopathological examination remains the gold standard for diagnosis. The typical histopathological findings include inflammatory infiltrates composed of lymphocytes, histiocytes, plasma cells, and eosino-

phils, localized in the deep fascia and subcutaneous tissue [5].

A more representative case report originates from the Children's Hospital in Cracow. The authors described the clinical course of RF-negative polyarticular juvenile idiopathic arthritis in a 5-year-old girl. They highlighted the complex clinical presentation and outlined the therapeutic modalities employed for this patient. Finally, they emphasized the role of biologic therapy as the appropriate treatment approach for managing the patient's condition.[6]

The final case report in this issue of "Rheumatology Forum" highlights the challenges in diagnosing and treating a specific subtype of inflammatory myopathy: inclusion body myositis (IBM). This rare and difficult-to-treat form of myositis is frequently associated with the presence of cN-1A autoantibodies. Unfortunately, unlike more common forms of inflammatory myopathies, IBM is resistant to conventional treatments, including steroid therapy. However, novel therapeutic strategies offer some promise, such as biologics like follistatin, which inhibits the myostatin pathway, and arimoclomol, a drug targeting the heat shock response (HSR). Another potential treatment is sirolimus, a drug widely used in transplantation that inhibits the mTOR pathway. An additional approach involves the use of monoclonal antibodies that directly target T-cells. However, despite high expectations from phase IIb clinical trials involving bimagrumab, a fully human monoclonal antibody that binds to the activin receptor, the results have not been confirmed in subsequent studies [7].

Finally, the "historical corner" of this issue of "Rheumatology Forum" features the profile of Jan van Breemen, a pioneer in rheumatology collaboration, elegantly presented by Eugeniusz J. Kucharz [8].

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