Achenbach Syndrome: A Diagnostic Dilemma

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Introduction: Achenbach syndrome is a poorly understood condition involving paroxysmal hematomas of the distal upper extremity digits. Achenbach is unique in which this condition is not associated with more serious underlying autoimmune etiologies and is a diagnosis of exclusion. Due to Achenbach syndrome's rarity, it can pose a diagnostic challenge to clinicians who may suspect a more serious underlying condition.

Case Presentation: A 44-year-old female with a past medical history significant for Raynaud's disease presented to the Hematology/Oncology clinic complaining of easy bruising and bleeding of her fingers. A nurse in the operating room, she reported a 2-3-year history of digital splitting that bled easily, often resolving within a day. The patient denied digital ulcers or gangrene. Her family medical history is significant for similar symptoms in both her father and sister. Initial workup, including coagulation studies, platelet aggregation, Von Willebrand screen, and Complete Blood Count with differential, were unremarkable. However, an abnormally elevated ANA titer of 1:160 prompted referral to rheumatology. Further testing for autoimmune diseases, including lupus anticoagulant, cardiolipin antibody, anti-double-stranded DNA antibodies, anti-scleroderma antibodies, complement levels, anti-centromere antibodies, Anti-Rho (SS-A), and Anti-La (SS-B), was negative. Physical examination revealed elongated capillaries with microhemorrhage in the right index finger but no sclerodactyly. Based on these findings, a diagnosis of Achenbach syndrome was made. Given the absence of significant underlying pathology and the characteristic clinical presentation, no further testing or investigation was deemed necessary.

Conclusion: Achenbach syndrome, a rare condition characterized by paroxysmal hematomas in the distal upper extremity digits, presents a diagnostic challenge due to its atypical presentation and potential for misdiagnosis. While many autoimmune diseases can exhibit similar clinical features, Achenbach syndrome is distinct in its lack of association with more serious underlying conditions. Recognizing Achenbach syndrome as a diagnosis of exclusion is crucial to avoid unnecessary investigations and interventions.

Keywords: Achenbach, Autoimmune Disorders, Hematology/Oncology

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