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74 HEREDITARY ANGIOEDEMA REVEALED BY COMPARTMENT SYNDROME

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Background
Hereditary angioedema (HAE) can present with a wide spectrum of clinical symptoms, including articular features like the compartment syndrome.
Objective
We present the peculiar case of a young female patient suffering from this rare complement system disorder and presenting as a « wrist oedema ».
Methods
A 14-year-old girl was admitted in the emergency room during a night shift. She presented with acute compartment (wrist) syndrome due to edematous compression of the median nerve, associated with a typical personal history of HAE. It resolved within 6 h with IV steroids and hyperhydration.
The quantitative and qualitative C1 INH esterase returning negative, the diagnosis of HAE type III was made.
The patient was treated with long-term treatment based on Danazol, (the most frequently prescribed attenuated androgen to treat HAE).
Results
HAE is due to the deficiency or dysfunction of the C1 inhibitor protein (quantitative or qualitative deficiency). A new nomenclature has replaced the initial use of denominations HAE type 1, 2 or 3; to speak rather of HAE with a deficient C1 inhibitor (type 1), with a dysfunctional C1 inhibitor (type 2) or with a normal C1 inhibitor (type 3) Type 3 HAE is rare and difficult to treat. It may respond to anti-estrogenic drugs such as progestins and especially to androgens; specific (very expensive) molecules do exist and act directly on the bradykinin pathways
Conclusion
A compartment syndrome may reveal extremely rare conditions such as hereditary angioedema.
Early recognition and management are the guarantee of a preserved functional prognosis.