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IMAGE ARTICLE

Vitamin-K Dependent Protein Deficiency and Pseudoxanthoma Elasticum

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27-year-old male referred from Dental clinic, due to asymptomatic incidental abnormal coagulation parameters.

Clinical examination showed loose skin folds of axilla, anterior abdominal wall and chest which were suggestive of Pseudoxanthoma Elasticum. Rest systemic examinations WNL. Patient has similar family history.

Coagulation profile showed PT-46 seconds, (control 12.50), INR 4.64 seconds, APTT 61.30 seconds (control 23.80), Mixing Studies-PT 13.8 seconds, PTT-29.7 Seconds.

Factor-VII-12.00%, Factor-XIII-113.10U/DL, Factor-V-73.00 seconds, Factor-IX-21 seconds, Rest laboratory tests were normal. Patient was diagnosed with

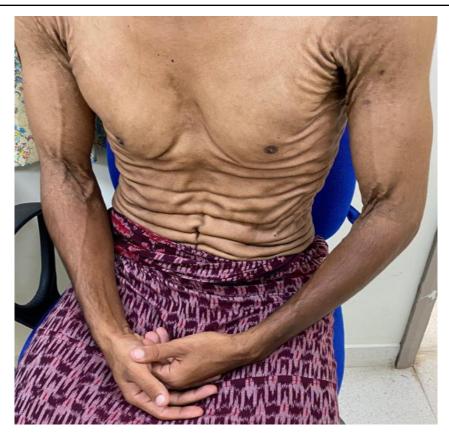


Figure 1: Patient was diagnosed with multiple coagulation factor deficiency.



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multiple coagulation factor deficiency Figure 1 and Figure 2.

There is a known association of VKDPD and Pseudoxanthoma Elasticum due to Gamma Glutamyl Carboxylase (GGCX) mutation. This patient has Gamma Glutamyl Carboxylase (GGCX) mutation which led to VKDPD & PXE!

If he bleeds, needs factor replacement with a PPC (4-Factor), not Vitamin-K. No need for prophylactic treatment as he is not bleeding phenotype.



Figure 2: Patient was diagnosed with multiple coagulation factor deficiency.

