GENETICS AND GENODERMATOSES

SKIN CANCER PREVENTION IN XERODERMA PIGMENTOSUM

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Introduction: Xeroderma pigmentosum (XP) is characterised by extreme sensitivity to light and early onset of oculocutaneous cancers. This study aims to evaluate the impact of photoprotection in preventing and slowing the progression of tumors during XP.

Material and Method: Thirty patients with XPC ({XPC V548A fs X572} mutation) participated in this study. Photoprotective methods (UV filtering film, special clothing, sunglasses, sunscreen (SPF 50+/UVA 40), stick for the lips (SPF 50+), etc.) were given to the patients. Dermoscopic follow-up for control purposes took place every 3 months over 2 years for all patients.

Results: Seventy percent of patients developed melanoma and non-melanoma cancers during the two years prior to the start of the study (83 basal cell carcinomas (BCC), 13 squamous cell carcinomas (SCC), 5 keratoacantomas (KA), and 3 Melanoma (MM)). Examination on day 0 (D0) showed 194 suspicious lesions. Of these lesions, 64 were considered malignant and were scheduled for treatment in the months following the start of the study. The remaining 130 suspicious lesions were smaller than 6 mm. Nineteen of them regressed and 111 were classified according to their dermoscopic features as BCC in 25 cases and undetermined in 86. An evaluaton of the overall photoprotective use was performed in all patients. The size of the BCCs in the patients with the least photoprotection progressed slowly. In two years, they indeed doubled instead of quadrupling. In patients with a good photoprotection, the size of the BCC stayed stable (increase of only 13.9%) The undetermined lesions had a decrease in their average size from D0 to D720 by 4.5%.

Conclusion: XP is a heterogeneous disease clinically and genetically. XP-C group is the most severe form.
Photoprotection must be started as early as possible and must be continued for life, even in the advanced stages of the disease.