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GENETICS AND GENODERMATOSES

REPORT OF TWO CASES OF XERODERMA PIGMENTOSUM IN MAURITIUS: PHOTOPROTECTION A MUST

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Background: Xeroderma pigmentosum (XP) is a rare genetic condition caused by defective nucleotide excision repair and characterised by skin cancer, ocular and neurological involvement. In order to prevent the occurrence of skin cancer, which can be sometimes fatal mainly in cases of melanoma, stringent sun protection is one of the important measures. Despite of these risks some patients neglect its importance essentially in the sunny tropical countries like Mauritius.

We here report two cases diagnosed as XP in the same year but with a different evolution in response to degree of sun protection.

Observations: Case A, a 13-year-old male patient diagnosed at the age of 5 years, used rigorous sun protection including sunscreen, photoprotective clothes, special head and eyewear. He has not developed until now any premalignant or skin cancer.

Case B, a 9year old girl diagnosed with the disease at the age of 2 years, used inadequate sun protection and followed poorly the given medical advice. She developed multiple skin tumours amongst a recently diagnosed facial melanoma.

Key message: Through comparative pictures of these two cases, we would like to emphasise on the importance of strict photoprotection and the role as well as the education of parents in the implementation of these measures, and hereby inhibiting the development of fatal complications including melanomas. This education is pivotal in the management of these patients in countries like Mauritius.





