

Conference Abstracts

Sunlight, Skin Cancer, and Survival: Xeroderma Pigmentosum Story in Zanzibar

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Background

Xeroderma Pigmentosum (XP) is a rare genetic disorder caused by inherited mutations that affect the body's ability to repair DNA damage from ultraviolet (UV) light exposure. This condition leads to increased sensitivity to UV radiation, resulting in severe sunburn, changes in skin pigmentation, and a higher risk of skin cancer in those affected.

Zanzibar, a group of semi-autonomous islands in Tanzania with a population of approximately 1.8 million, has a notably high incidence of XP within its community, where many individuals present with skin cancer at an early age. The condition is particularly prevalent on Pemba Island, where intermarriage is a common practice. Currently, there is no known cure for XP. Management of the condition involves strict avoidance of direct sunlight and the use of sun protective measures. These include wearing long-sleeved clothing, wide-brimmed hats, and sunglasses, as well as taking vitamin D supplements and using sunscreen with a minimum SPF of 50. Regular skin examinations are also essential for early detection of potential skin cancer.

Methods

This was a retrospective cross-sectional observation, where data from the XP clinics were collected over 3 years with a gradual increase in

patient numbers as awareness and access to services were scaled up. The data were collected through the different outreaches in Pemba Island and the dermatology clinic at Mnazi Mmoja Hospital between 2019 and 2022. The diagnosis was made clinically.

Results

The mean age of XP was 8.7 years, ranging from 6 months to 30 years. The majority were male (53.6%) and came from Pemba Island. Micheweni had the highest concentration of XP, accounting for 32 cases.

All XP patients presented with classic symptoms, including fine scaling and freckle-like areas of hyperpigmentation (lentigines) and hypopigmentation on the face and back of the neck. Photophobia and tumours were in more than half of the population, 59.6%, primarily on the face, scalp, and tongue, likely due to chronic sun exposure and trauma to the tongue, respectively.

A significant number of these children (19, 33.9%) died at a very young age; almost all were <10 years of age. Studies suggested skin cancer and neurodegenerative disease as the main culprits.

Conclusion

Xeroderma Pigmentosum (XP) significantly affects the quality of life for those who have the condition and can lead to skin cancer at an early

age, which, if left untreated, can be fatal. Micheweni has a high number of patients with XP in Zanzibar, suggesting a possible considerable interaction between families, likely due to consanguinity. This points to an important

area for future research to establish causality and potentially design interventions aimed at reducing the incidence of skin cancer in these underserved communities. We believe that reducing skin cancer in children requires a collaborative effort.