

Criminal Exploitation of a Child with Xeroderma Pigmentosum

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Abstract: Xeroderma pigmentosum (XP) is a rare autosomal recessive disease with cutaneous, ocular and neurological symptoms. People suffering from this disease are very sensitive to light and are very likely to develop cancer; so they need to be protected from sunlight.

Keywords: Criminal, exploitation, child, Xeroderma pigmentosum.

INTRODUCTION

Xeroderma pigmentosum (XP) presents as extreme photosensitivity of the skin and eyes with a 1000-fold increased frequency of cutaneous basal and squamous cell carcinomas and melanomas and a small increase in nervous system neoplasms [1]. It is a rare autosomal recessive genodermatosis, reported to have a worldwide incidence of 1: 250,000 live births. Patients have a genetic inability to repair DNA damage, induced by ultraviolet light. It is estimated to vary from 1 in 20,000 in Japan to 1 in 250,000 in the USA, and approximately 2.3 per million live births in Western Europe [2].

The critical mediator of the cellular response to DNA damage is the p53 tumor suppressor gene. The function of p53 tumor suppressor gene has been attributed to its role as a transcription factor regulating expression of genes involved in DNA damage response pathways that affects apoptosis, DNA repair, and cell cycle regulation [3, 4].

Ocular complications are nearly as common as skin lesions with keratitis progressing to corneal opacification, loss of eyelashes, ectropion, entropion and benign and malignant lesions of the cornea and eyelids. Neurological complications have found to occur in about 30% of cases that can be severe [5]. XP

is usually inherited in an autosomal recessive manner with phenotypically normal heterozygotes. At least seven different subtypes (complementation groups A-G) as well as XP variants are reported. Various rare forms occur have been found to occur in combination with other disorders such as Cockayne's Syndrome [6]. The diagnosis of XP is considered in a young patient with marked photosensitivity, xerosis and multiple pigmented lesions [7, 8].

CASE REPORT

We report the case of a 7-year-old male child, presented with photophobia. He is an orphan, since he lost his father when he was 1-year-old. The child was enrolled in a kind of informal koranic school when he was 5-year-old. Instead of receiving instruction and teaching from his master (a marabout); he was compelled to beg for this one. The clinical findings were: bilateral xerosis and multiple pigmented lesions on the skin, multiple wounds on the scalp, bilateral punctuate keratitis, and chemosis.



Fig. 1: Xeroderma pigmentosum in a 7- year- old male

DISCUSSION

Usually Xeroderma pigmentosum has found to occur at the age of 1-2 years with photosensitivity and burning after minimal sun exposure. Later cutaneous manifestations include increasing dryness of skin, freckling, and telangiectasia, with increased incidence of skin cancer on sun exposed sites. The median age of onset of skin cancer is 8 years. Ocular abnormalities include photophobia, ectropion, conjunctival injection, keratitis, and tumours [6, 9].

People suffering from XP should not face sun exposure. Early age XP patients are sensitive to even minimal sun exposure resulting in development of erythema, vesicles and oedema. By the age of two years solar lentigos, xerosis and pigmentation occur. Later in childhood dysplastic and neoplastic lesions occur with the development of actinic keratosis, keratocanthoma, basal cell carcinoma, squamous cell carcinoma and malignant melanoma [10].

The current case points out the violation of all the rights of the child to protection and even to health care. Children are fragile human beings and need therefore to be protected particularly when they are sick or disable.

Motherhood and childhood are entitled to special care and assistance. All children, whether born in or out of wedlock, shall enjoy the same social protection [11]. It is unacceptable that such a fragile child be used as a beggar, since he is obliged to move under the sun shine and jump between wheeler engines during harsh times. This child with this terrible genetic disease and above all, orphan does not deserve to be deprived.

Many international conventions protect the rights of child; but their enforcement requires the commitment of States [12].

All children have the right to be protected from violence, exploitation and abuse. Yet, millions of

children worldwide from all socio-economic backgrounds, across all ages, religions and cultures suffer violence, exploitation and abuse every day [13].

CONCLUSION

People suffering from this disease are very sensitive to light and are very likely to develop cancer; so they need to be protected from sunlight.

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