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Multiple Palpebral Carcinomas: Ophthalmologic And Maxillofacial Management: A Case Report

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Abstarct:

Introduction : Xeroderma pigmentosum (XP) is a rare genetic condition that makes individuals sensitive to ultraviolet (UV) rays and predisposed to skin and eye cancers. Mutations altering DNA repair favour the accumulation of lesions and the development of malignant tumours.

Observation:

A 21-year-old XP patient presented with multiple palpebral carcinomas 6 years after exenteration for squamous cell carcinoma of the left eye. The tumours were multiple, of varying sizes, located on the upper and lower eyelids and the medial canthus. Excisional surgery with skin grafting was successfully performed, followed by careful post-operative follow-up.

Discussion:

XP induces UV sensitivity, increasing the risk of skin and eye cancers, particularly basal cell and squamous cell carcinomas. Ophthalmological symptoms, such as photophobia, may precede skin manifestations. The management of palpebral BCC in XP patients is a challenge because of their number, their aggressiveness and the difficulties of reconstruction. Surgery remains the preferred treatment, requiring collaboration between ophthalmologists and maxillofacial surgeons.

Conclusion:

This case highlights the challenges inherent in the management of palpebral carcinomas in patients with xeroderma pigmentosum. A multidisciplinary approach is required for early diagnosis, appropriate surgical treatment and rigorous post-operative follow-up. Future research is crucial to developing innovative therapeutic strategies aimed at improving the quality of life and prognosis of these patients faced with this rare and complex condition.

Introduction:

Xeroderma pigmentosum (XP) is a rare genetic entity characterised by increased sensitivity to ultraviolet (UV) radiation and a predisposition to skin and eye cancers. Mutations affecting DNA repair mechanisms following UV exposure lead to an accumulation of lesions and genomic instability. Clinical manifestations of XP include photophobia, abnormal skin pigmentation and actinic keratoses, as well as the frequent development of basal cell carcinoma (BCC), squamous cell carcinoma (SCC) and other cutaneous and ocular malignancies. This study focuses on a singular case of multiple palpebral carcinomas in a 21-year-old patient with XP, outlining the particularities of the clinical presentation, the surgical management and the therapeutic challenges encountered.

Case report:

The patient was 21 years old and was being treated for xeroderma pigmentosum. He had undergone exenteration for squamous cell carcinoma of the left eye 6 years previously.

Presented for ophthalmological consultations after being lost to follow-up after 02 years following his first exenteration, following the appearance of multiple swellings of different sizes and locations in the right eye.

Macroscopic examination revealed:

- An upper palpebral mass measuring 35mm H and 30mm V, with the appearance of an ulcerating and budding tumour with a large haemorrhagic plaque.
- A budding mass extending from the internal canthus to the root of the nose.
- An inferior subpalpebral mass measuring 18mm H and 23mm V with a nodular pearly appearance and purulent foci..

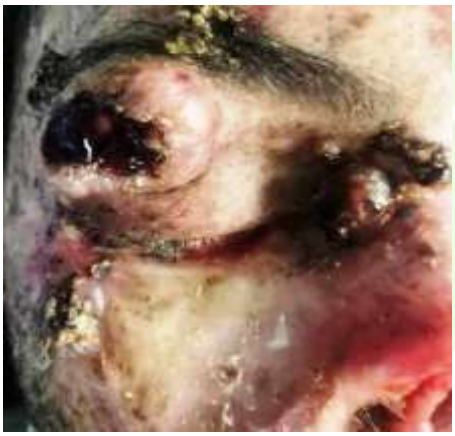

Figure 1: Upper palpebral, subpalpebral and medial canthal tumour lesions

Ophthalmological examination was difficult given the limited palpebral opening in the left eye, but the cornea appeared opacified with corneal neovessels.

An assessment of locoregional extension was carried out using cerebral MRI, which revealed subcutaneous nodular lesions more marked over the right orbit and the right internal canthus, with no invasion of adjacent structures.

Given the extent of the tumour lesions, a maxillofacial opinion was sought and the decision was made to perform an excisional biopsy with skin grafting.

During the operation, an excisional biopsy of the lesions was performed, with a healthy safety margin of 5mm, and the skin was grafted using an abdominal flap and sutured with 6/0 self-absorbable thread. A tarsorrhaphy was performed to protect the globe with the application of a compression dressing with vaseline on the grafted areas and the donor area..

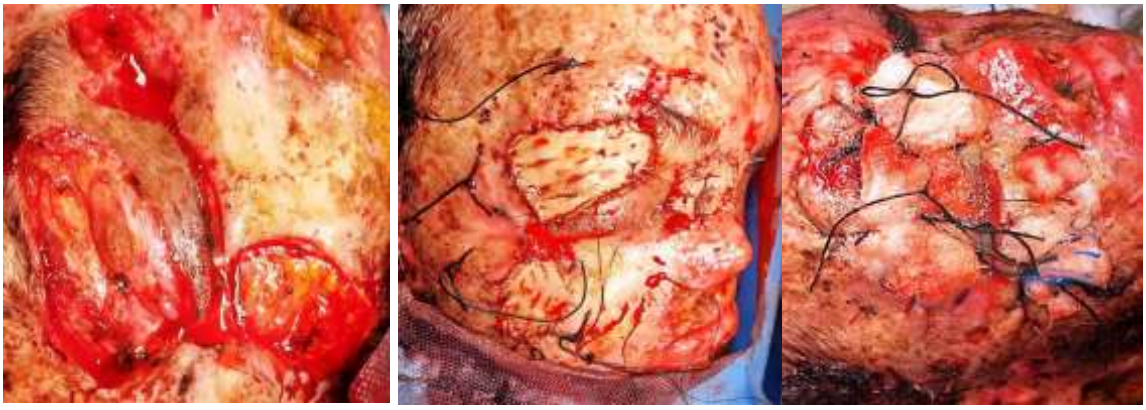

Figure 2: images showing the different stages of the operation

Post-operative follow-up was carried out, with a check-up every week for the first month and then every 2 weeks for the next 6 months, in order to detect recurrences and new lesions. In our case, the post-operative follow-up was straightforward with no significant recurrence, and the results were marked by complete healing..

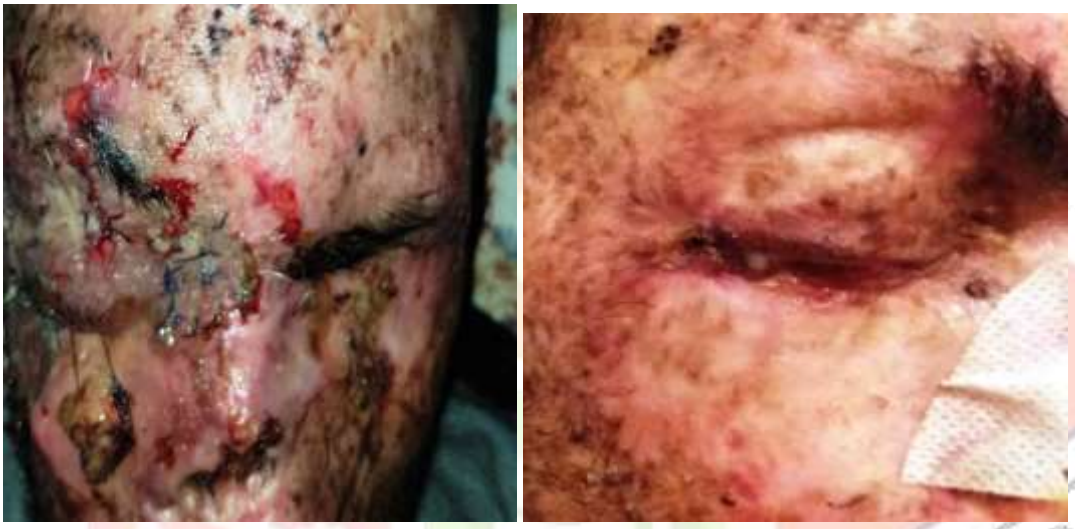

Figure 3: image taken post-operatively at D15 and at 6 months.

Discussion :

Hypersensitivity to ultraviolet (UV) rays and a predisposition to skin and eye cancers are the hallmarks of xeroderma pigmentosum (XP). XP patients have a defect in the repair of UV-damaged DNA, leading to an accumulation of mutations and genomic instability. Symptoms of XP include photophobia, abnormal skin pigmentation, actinic keratoses, skin ulcers and malignant tumours. Basal cell carcinoma (BCC), squamous cell carcinoma (SCC) and melanoma are the most common skin cancers in XP patients. Uveal melanoma, conjunctival carcinoma and palpebral carcinoma are the most common ocular cancers.

Basal cell carcinoma is a major public health issue, with an incidence rate of 4-8% that increases over time [1]. Several risk criteria, such as cumulative UV exposure and phototype [2, 3], have been associated with the epidemiology of basal cell carcinoma. Various hereditary conditions that increase the risk of skin cancer have been described [4].

BCC cells originate from the basal epithelial layer. The tumour resembles the normal basal epithelial cell layer on histology. The threads, cords and islands of tumour are created by the BCC. Previously, the majority of BCCs were considered to be clinically nodular. It has now been shown that the perilesional region shows two types of tumour growth: nodular and morpheiform (fibrosing or sclerosing), which has a higher recurrence rate.

Periocular BCC occurs most frequently on the lower eyelid and medial canthus, and less frequently near the lateral canthus [5][6]. In our case the locations were upper palpebral, lower palpebral and medial canthal.

In the literature, there is a delay in diagnosis due to the late realisation of the seriousness of the lesion in almost 10% of patients, who may consult a specialist after 5 years [7]. Ophthalmological symptoms are generally bilateral, correlated with skin involvement, and photophobia is the most consistent and earliest sign, making it possible to orientate the diagnosis even before skin symptoms [8][9]. Photophobia gradually diminishes with the appearance of corneal opacification. The severity of the condition depends on the appearance of benign and, above all, malignant tumours [10, 11], which may be located in the palpebral, limbal or conjunctival areas, compromising functional and vital prognosis.

Because these tumours are often multiple, recurrent and large, the management of palpebral BCCs in XP patients is a therapeutic challenge. Although the loss of substance and the difficulties of reconstruction may limit surgery, it remains the reference treatment. XP patients should not receive radiotherapy as it increases the risk of new cancers.

Palpebral carcinomas are primarily managed surgically, with the aim of achieving complete removal of the tumour with sufficient safety margins, with anatomopathological analysis of the cuts at the edges. In a recent study of 470 extemporaneous examinations, only 4 were falsely negative, and the same study also found that extemporaneous histological examination reduces the rate of recurrence compared with conventional surgery by using probabilistic margins. The reliability of extemporaneous examination is as high as 98% [12]. Topical treatments, such as imiquimod or 5-fluorouracil, may be useful for superficial lesions, but are not very effective for nodular or infiltrating lesions. Advanced or metastatic cases may require systemic treatments, such as Hedgehog pathway inhibitors, but these are associated with serious side effects and a risk of resistance.

To ensure optimal management of palpebral BCC in XP patients, ophthalmologists and maxillofacial surgeons need to work together. Ophthalmologists are responsible for diagnosis, work-up, monitoring and treatment of ocular complications. Surgeries involving extensive lesions require extensive resection and complex reconstruction. In addition, depending on the case, they may perform local or distant skin grafts or flaps. The aim is to ensure total removal of the tumour while preserving the function and appearance of the eyeball and eyelids.

Our case presents several particularities that distinguish it from cases reported in the literature. Firstly, this was a very young patient, aged 21, with an early basal cell carcinoma, given the association with XP. Secondly, there were multiple lesions of different sizes and in different locations, demonstrating the aggressiveness of BCC in XP patients. Finally, this case required abdominal flap skin grafting, a technique rarely used for palpebral reconstruction. Skin grafts are often taken from the auricle, forehead, neck or inguinal region, as they offer a better colour and texture match with the palpebral skin. However, the abdominal flap made it possible to cover a large surface area of loss of substance while guaranteeing effective vascularisation and healing in our case.

Conclusion :

The management of palpebral basal cell carcinomas in patients with xeroderma pigmentosum represents a major clinical challenge. This case, because of its young age, the multiplicity of lesions and the surgical complexity required for their treatment, highlights the importance of a multidisciplinary approach. Close collaboration between ophthalmologists and maxillofacial surgeons is essential for early diagnosis, effective treatment and rigorous follow-up of these patients, who are at high risk of recurrence and serious complications. By exploring and documenting cases such as this, it becomes imperative to seek innovative therapeutic strategies aimed at improving the quality of life and prognosis of these individuals faced with this rare and complex condition.

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