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Facial resurfacing in xeroderma pigmentosum: are we spoiling the ship for a ha'p'orth of tar?

G. ASHALL, A. A. QUABA and M. E. J. HACKETT

North East Thames Regional Plastic Surgery Unit, St Andrew's Hospital, Billericay, Essex

Summary—A case of xeroderma pigmentosum (XP) is reported whose treatment included sub-total excision of facial skin and resurfacing in aesthetic units with grafts of relatively undamaged buttock skin. The principle of resurfacing is logical and supported by this and other reports which are reviewed. Though control was achieved in the grafted areas, tumours continued to develop in adjacent ungrafted skin. The importance of radical excision of exposed skin in severe cases of XP is emphasised.

In spite of the discovery of the genetically determined DNA defect in XP (Cleaver, 1968), there is no cure and the outlook for patients with this disease remains bleak. Survival far into adulthood is rare (Kucan and Orgel, 1985; Montandon, 1985) and much of the patient's time may be spent as the surgeon reaps the harvest of skin tumours resulting from earlier unguarded exposure to sunlight. A case is reported of an attempt to control tumour formation by facial resurfacing.

Case report

The patient was born in 1963, the second child of non-consanguineous West Indian parents with no family history of skin disease. When aged 9 years he moved with his family to St Lucia where patches of altered pigmentation appeared on his face and arms. Gradually all exposed parts of his body became involved and skin tumours developed. After 3 years of heavy exposure to sunlight he returned to the United Kingdom in 1976. The clinical diagnosis of XP was confirmed by studies of DNA repair in fibroblast cultures. Numerous lesions on the face (Fig. 1) were treated by excision or curettage and diathermy while generalised changes in the facial skin were treated with 5 fluorouracil cream.

Over the next 2 years, in spite of the avoidance of sunlight and the use of ultra-violet barrier cream, basal and squamous cell lesions continued to appear. In June 1977 11 basal cell carcinomata were excised from the face, incompletely in several cases owing to the difficulty in identifying tumour margins in a background of unstable skin.

In January 1978 the worst affected skin, *i.e.* that over both malar regions, bridge of nose and upper lip, was excised and replaced with split thickness grafts taken with Padgett dermatome from the buttocks where solar damage was minimal. Between 1978 and 1981 at least 10 lesions were excised from outside the grafted areas.

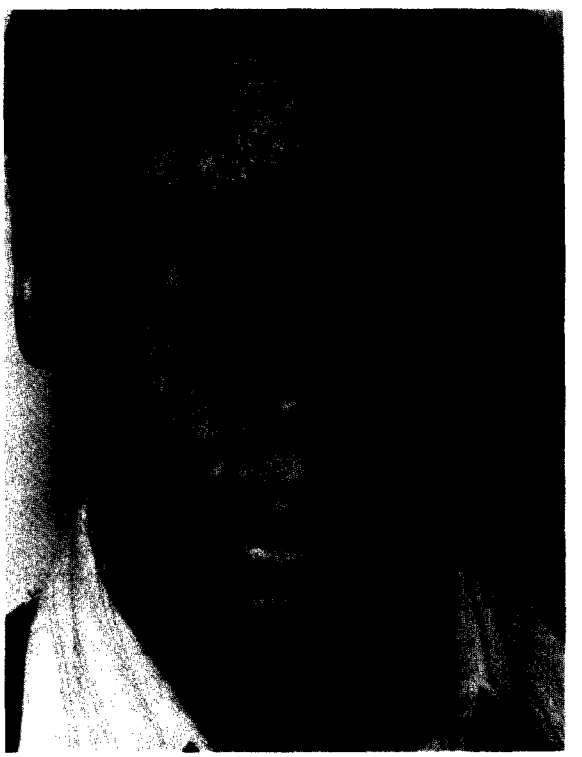

Fig. 1

Figure 1—Patient in 1976 soon after presentation.

By March 1982, 4 years after grafting, there were new lesions affecting much of the ungrafted areas of the face. At this stage it was realised that the strategy of excising

lesions as they arose was failing to control the disease. The entire skin of the forehead, lower eyelids and lower cheeks (the malar regions having been grafted previously) was excised and the defects resurfaced with thick partial thickness grafts, again taken from the buttocks. Several deeply infiltrating lesions of the nose precluded excision of skin alone and therefore amputation was performed.

Between 1982 and 1985 a number of lesions developed outside or at the edge of the grafts, the most significant of which was a basal cell carcinoma of the left inner canthus which invaded the orbit necessitating exenteration and temporalis flap cover (Fig. 2). In January 1986 the patient was admitted with a neglected ulcerating basal cell carcinoma centred over the lateral margin of the right pyriform fossa and extending on to the right lower eyelid, cheek and lateral wall of nose. This was excised and the defect grafted with relatively undamaged skin from the left iliac region. The patient's most recent admission was in September 1986 for excision of basal cell carcinomata of the lower lip vermilion and left ear lobe.

Discussion

Genetic engineering may eventually provide a cure for XP. Present management must include prevention of further damage to the skin, but for areas which have been heavily exposed prior to diagnosis (usually including the face) measures such as avoidance of sunlight and application of ultra-violet barrier cream are very often too late. Although the exact mechanism of carcinogenesis is not clear, once DNA within skin cells has been altered by ultra-violet radiation the natural history of the disease is for malignancy to develop. It is logical, therefore, to give these areas a second chance by resurfacing them with relatively undamaged skin.

Several authors (Vaillant *et al.*, 1979; Mouly *et al.*, 1980; Lemaitre *et al.*, 1983) have discussed the surgical treatment of XP which includes (i) local excision of tumours ('chirurgie a la demande'), (ii)

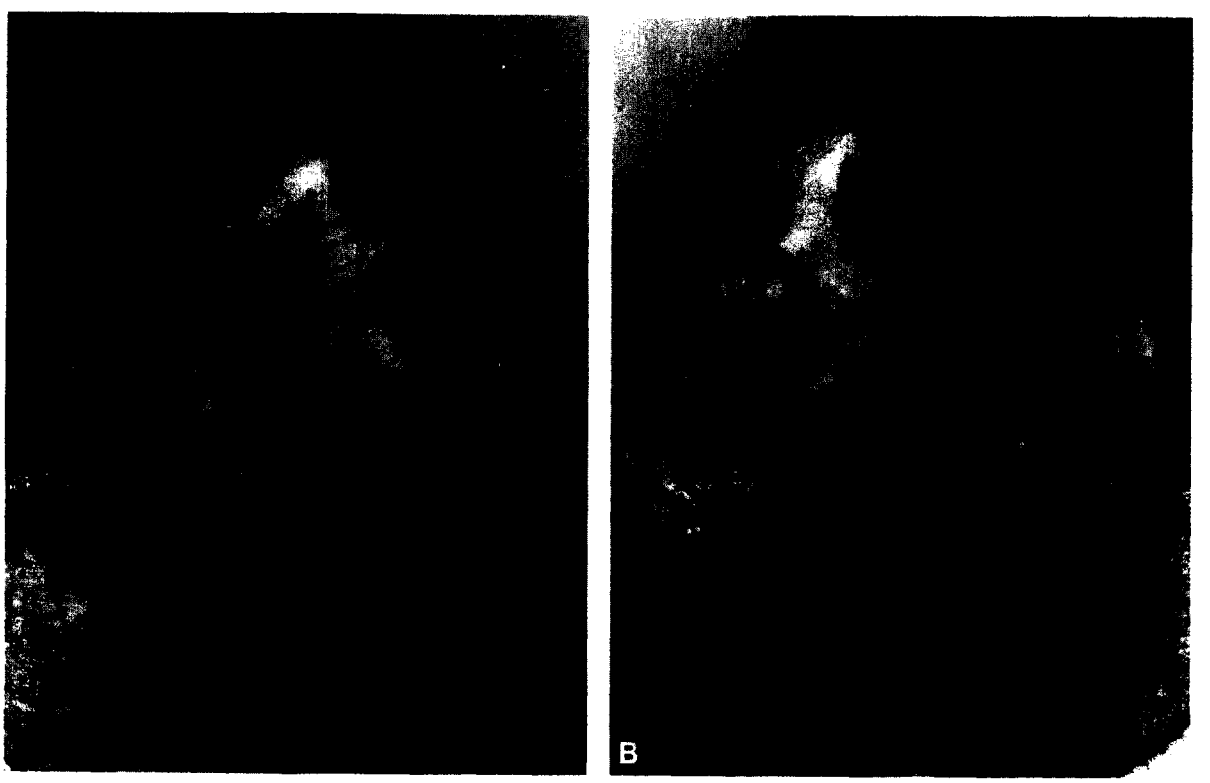

Fig. 2

Figure 2--Patient in 1986 having previously undergone facial resurfacing, nasal amputation, and left orbital exenteration. Note healthy appearance of grafts on cheeks compared with ungrafted neck and auricular skin. A lesion has recently been excised from the right post-auricular region and two small new lesions are visible in the left pre-auricular region.

excision of unstable or tumour-bearing skin and resurfacing in aesthetic units, and (iii) dermabrasion. The rationale behind (ii) and (iii) is that resurfacing is achieved with skin less exposed to solar radiation. Kubacek (1975), however, in a human experiment unlikely to be repeated, found that even sun-damaged skin lost the characteristic signs of XP when transplanted from one site to another and he postulated that transplantation *per se* may have an ameliorating effect.

Moore and Iverson (1954) were the first to suggest radical excision of large areas of skin involved by malignant or pre-malignant lesions followed by graft replacement. This and subsequent reports are summarised in Table 1. Martins' (1965) patient underwent mono-block excision of the entire face, lasting 8 hours and requiring tracheostomy; other authors performed staged or sub-total resurfacing. Grafts were usually harvested from the thigh or trunk. Mouly *et al.* recommend scalp as a donor site likely to provide disease-free grafts.

Dermabrasion as a prophylactic manoeuvre for pre-cancerous skin was first described by Epstein (1956) and later reports of its use in XP are summarised in Table 2. It is thought that benefit accrues from re-epithelialisation by cells derived from those lying deep in the skin adnexae and which therefore have received little exposure to ultra-violet radiation (Epstein *et al.*, 1972). In addition to the improvement achieved by dermabrasion in Epstein *et al's* (1972) 26-year-old patient, fewer changes of XP occurred in thigh split skin graft donor sites and this was put to therapeutic use when the remainder of both lower limbs was shaved with a dermatome. The authors considered dermatome shaving more practical than dermabrasion for large areas. They also claimed a better cosmetic result from dermabrasion than was achieved in

Table 1 Resurfacing in xeroderma pigmentosum

Year	*Author*	*Patient sex & age*	*Areas resurfaced*	*Donor site*	*Details of follow-up*
1954	Moore & Iverson	M 50	forehead & temples	thigh	>2 years recurrence free
1959 1972	Woolf et al. Pickrell	M 6	total face (staged)	thigh	recurrence free at 17 year follow-up (by Pickrell)
1962	Fregni & Francesconi	M 22	forehead (twice)	buttock	regrafted after 4 years subsequent follow-up unclear
1963	Hsu & Wu	F 19	cheek chin & mandible	abdomen thigh	>3 years recurrence free 1½ years recurrence free
1965	Martins	M 9	total face (including vermilion & eyelids)	abdomen	3 years recurrence free
1970	Gleason	F 6	forehead & cheeks	?	5 years recurrence free
1975	Kubacek	M 7	total face, back of neck, dorsum of hands (staged)	trunk	11 years recurrence free
1980	Mouly et al.	F 25	face (excluding forehead)	?	recurrences at the periphery of grafts after 10 years

Table 2 Dermabrasion in xeroderma pigmentosum

Year	*Author*	*Patient sex & age*	*Area dermabraded*	*Details of follow-up*
1958	Epstein	8 patients (no details)	not stated	recurrences in 6 months to 4 years
1965	Martins	F 3	face & neck	not stated
1965	Yosipovitch et al.	M 19	face	18 months recurrence free
1972	Epstein et al.	F 26	face	7 years recurrence free
1980	Mouly et al.	F 6 F 11	face cheeks	? 4 years recurrence free not stated

Gleason's (1970) patient by skin grafting. This comparison is unfair, however, as Gleason's patient had already had multiple treatments (including dermabrasion) which had failed to control the disease. Dermatome shaving and dermabrasion must be regarded as prophylactic measures which may be therapeutic only for the most superficial of lesions.

The rarity of XP renders controlled trials of treatment impracticable. Nevertheless it is possible to draw certain conclusions from the foregoing review of reported cases. (1) Dermabrasion, dermatome shaving or excision of skin affected by XP and replacement with split skin grafts appears at least to delay the development of malignant change in these areas. (2) The thighs, abdomen, scalp and buttocks are potential donor sites which in most cases are likely to have had the least exposure to sunlight. In our patient the latter site was the only remaining source of reasonably large sheets of undamaged skin. (3) Many of the reported cases, including ours, continued to develop tumours in adjacent ungrafted areas of the face. On a purely theoretical level this may be taken to vindicate the use of grafts. In practice the patient with continuing tumour formation anywhere is not likely to care that his grafts happen to be tumour-free. The inference must therefore be to attempt more radical and complete resurfacing of exposed skin. There may be an understandable reluctance to graft or dermabrade difficult areas such as the central face and ears as a prophylactic manoeuvre. However, given the near certain risk of malignancy developing at some stage in heavily exposed skin and the notoriety for infiltration of tumours, for example at the canthi and alar bases, this is perhaps a case of spoiling the ship for a ha'p'orth of tar. It is the authors' belief that earlier and more radical excision and grafting of facial skin would have improved the prognosis in the case reported.

A final point needs emphasis. There is a spectrum of severity of XP, as illustrated by the younger sister of our patient who suffers from a much less aggressive form of the disease. Not all patients require the radical surgical approach advocated here, which should be reserved for those who develop a large number of invasive tumours within a short period of time.

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References

Cleaver, J. E. (1968). Defective repair replication of DNA in xeroderma pigmentosum. *Nature*, **218**, 652.

Epstein, E. (1956). Planing for pre-cancerous skin. *Archives of Dermatology*, **74**, 312.

Epstein, E. (1958). Planing for pre-cancerous skin: a follow-up study. *Archives of Dermatology*, **77** 676.

Epstein, E. H., Burk, P. G., Cohen, I. C. and Deckers, P. (1972). Dermatome shaving in the treatment of xeroderma pigmentosum. *Archives of Dermatology*, **105**, 589.

Fregni, A. and Francesconi, G. (1962). Xeroderma pigmentosum. *Minerva Chirurgica*, **17**, 894.

Gleason, M. C. (1970). Xeroderma pigmentosum—five year arrest after total resurfacing of the face. *Plastic and Reconstructive Surgery*, **46**, 577.

Hsu, C-H. and Wu, P-K. (1963). Xeroderma pigmentosum with malignant change treated by excision and free skin graft. *Chinese Medical Journal*, **82**, 763.

Kubacek, V. (1975) Xeroderma pigmentosum. *Acta Chirurgicae Plasticae*, **17**, 3.

Kucan, J. O. and Orgel, M. G. (1985). Xeroderma pigmentosum: Oldest living survivor? *Plastic and Reconstructive Surgery*, **75**, 107.

Lemaitre, A., Romagnolo, J. and Linker, M. (1983). La chirurgie dans le traitement du xéroderma pigmentosum (réflexions à propos d'un cas). *Acta Stomatologica Belgica*, **80**, 121.

Martins, A. G. (1965). The challenge of xeroderma pigmentosum. *British Journal of Surgery*, **52**, 56.

Montandon, D. (1985). Xeroderma pigmentosum: Oldest living survivor? *Plastic and Reconstructive Surgery*, **76**, 797.

Moore, C. and Iverson, P. C. (1954). Xeroderma pigmentosum. *Cancer*, **7**, 377.

Mouly, R., Dufourmentel, C., Banzet, P. and Papadopoulos, O. (1980). Xeroderma pigmentosum. *Annales de Chirurgie Plastique*, **25**, 117.

Pickrell, K. (1972). Follow-up clinic. *Plastic and Reconstructive Surgery*, **49**, 83.

Vaillant, J. M., Flageul, G., Aubiniere, E., Couly, G. and Oger, P. (1979). Un autre regard sur le traitement du xéroderma pigmentosum. *Annales de Chirurgie Plastique*, **24**, 257.

Woolf, R., Kepes, J., Georgiade, N. and Pickrell, K. (1959). Xeroderma pigmentosa. *Plastic and Reconstructive Surgery*, **24**, 214.

Yosipovitch, Z., Sachs, M. I. and Neuman, Z. (1965). Multiple malignant tumour formation in xeroderma pigmentosum. *British Journal of Plastic Surgery*, **18**, 314.

The Authors

G. Ashall, FRCS(Eng), FRCS(Ed), Registrar in Plastic Surgery
A. A. Quaba, FRCS(Plast)Ed, Senior Registrar in Plastic Surgery
M. E. J. Hackett, FRCS, Consultant Plastic Surgeon

North East Thames Regional Plastic Surgery Unit, St Andrew's Hospital, Billericay, Essex.

Requests for reprints to Mr G. Ashall at the above address.

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