KERATOPLASTY IN XERODERMA PIGMENTOSUM*

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KERATITIS is the chief ocular complication of xeroderma pigmentosum. The severe photophobia, a marked feature of this disease, and the extensive corneal involvement, mainly in the interpalpebral zone, make sufferers from this malady virtually blind, at any rate by day. In a report of seven cases of this disease with ocular complications (Sivasubramaniam and Hoole, 1952), keratoplasty was suggested as a means of treating the corneal condition, though at that time no case had been so treated.

As there appears to be hardly any literature on this aspect of xeroderma pigmentosum, the following account of three patients treated by keratoplasty in the Government General Hospital, Jaffna, Ceylon, may be of interest.

As far as keratoplasty is concerned the important lesions are a diffuse keratitis extending down to the anterior two-thirds of the substantia propria of the cornea, and telangiectases visible to the naked eye at and within the limbus. The depth of the keratitis was only discernible on the operating table as examination with the biomicroscope was precluded by the severe photophobia. Symblepharon may be encountered though this was not a problem in the cases described below.

Case Reports

Case 1, a 28-year-old female, had bilateral keratitis. The right cornea showed a transversely disposed triangular patch of calcification in the interpalpebral zone with the base of the triangle nasally.

Visual acuity in August, 1952, was 6/60 in the right eye and 4/60 in the left.

In August, 1952, a left central 5-mm. penetrating keratoplasty was performed under endotracheal anaesthesia. In order to ascertain the depth of the keratitis a lamella of cornea was first dissected; the posterior third of the cornea was unaffected. As a full-thickness graft had already been cut, a penetrating keratoplasty was performed. The recipient cornea was a little thicker than the graft. Healing was uneventful and the graft remained clear for 2 months, the best vision during this period being 6/60 with -4 D sph. Gradually the graft turned opaque and even the junctional zone between the graft and host was lost in the general haze. Cortisone was not available at the time.

As the vision of the right eye deteriorated appreciably, a central 6 mm. lamellar keratoplasty was performed on this eye under general anaesthesia. The trephine as well as the lamellar keratoplasty knife were felt to grate against the calcified patches on the cornea though the latter in no way impeded the operation. Roughly the anterior

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two-thirds of the cornea were included in the disc of cornea removed and the bed was found to be clear. The graft remained clear from the outset, cortisone drops being instilled from the seventh post-operative day (Fig. 1). Vision, which at the time of operation in August, 1954, was 1/60, improved to 6/36 post-operatively—a level still maintained. This visual acuity could not be improved by glasses.

FIG. 1.—Case 1, right eye after operation. Note calcified patch nasal to graft. The two bright areas along the upper edge of the graft are the reflections of the light used for photography.

Relief from photophobia, irritability, lacrimation, and mistiness of the cornea adjacent to the graft were beneficial results of the operation. The calcified area nasal to the graft persisted, however, and to date has not extended beyond the graft edge.

Case 2, a 24-year-old girl, had keratitis in the left eye, a striking feature of which was the unusually large telangiectasis invading the cornea from the limbus.

The visual acuity was 1/60 in the affected left eye, and 6/9 in the right. The patient sought relief from irritation of the eye, pain, and photophobia.

A keratectomy was done first to rid the cornea of a knotty telangiectatic mass in its centre. Bleeding vessels were touched with a hot probe. In August, 1954, 3 weeks after the keratectomy, a central 5-mm. lamellar keratoplasty was done under local anaesthesia. Threatened vascularization of the graft was controlled by liberal exhibition of cortisone drops. Immediate post-operative vision was 6/24 which steadily improved to 6/9 in about 5 months. The graft is clear at the time of writing.

Case 3, a 20-year-old girl, had bilateral keratitis. The left eye had had a limbal growth which had been excised in 1951; microscopic examination had demonstrated a squamous carcinoma.

FIG. 2 (a and b).—Case 3, left eye before operation.

FIG. 3.—Case 3, left eye after lamellar keratoplasty.

A total lamellar graft was performed on this eye under local anaesthesia. Bleeding was profuse and was arrested by application of hot probes. A 10-mm. lamellar graft was secured in place by overlay sutures. The graft took at first and remained clear (Figs 2 and 3). Visual acuity which was hand movements at the time of operation in March, 1955, improved to 6/60, and all irritative symptoms disappeared. At the end of 6 months the graft became opaque through vascularization, especially at the periphery.

Cortisone and a combination of neomycin and cortisone were used topically, and riboflavin, ascorbic acid, and calcium were given systemically, but the beneficial effects of these were short-lived.

Comment

Keratoplasty was performed in four eyes of three patients with xeroderma pigmentosum. Of nine patients under observation, three had eyes that were shrunk, two refused operative treatment, and one had very mild corneal changes not warranting surgery.

Technically, the operation is not different from keratoplasty in other conditions, though the vascularity and photophobia make surgery more difficult. The vascularity presents a formidable problem, for not only the trephine cut but also the suture points bleed. The latter may be overcome by the use of general anaesthesia.

As the keratitis is confined to the anterior two-thirds of the cornea, only lamellar keratoplasty is ideally suited; apart from satisfying the surgical indication for such a step, it also provides for safety in the conduct of the operation. If the telangiectases could be attacked by x irradiation before and after surgery, the chances of securing clear grafts might perhaps be greater, but this was not available in the institution in which these patients were treated.

The failure of the first operation is attributable to the fact that cortisone was not used post-operatively. The liberal exhibition of cortisone topically in the second eye of Case 1 and in Case 2 seems to have materially improved the post-operative health and viability of the grafts. Cortisone appeared to be mainly responsible for a shrinkage of the telangiectases in the second patient's cornea, though the therapeutic effect of keratectomy and of lamellar keratoplasty on the telangiectases should not be underrated.

The keratectomy in Case 2 was not followed immediately by keratoplasty owing to want of donor material. This situation, though forced on us, appears to have been the desirable course of action, for it is probable that new vessels would have invaded the graft had one been done in the wake of keratectomy.

The opacification of the graft in Case 3 was manifestly due to vascularization which had a greater chance of engulfing the graft, because its edge lay close to the limbus where numerous blood vessels were already present. Cortisone alone could not have kept so many blood vessels at bay.

The symptomatic relief to these patients afforded by keratoplasty and the clearing of the cornea adjacent to the graft are examples of the beneficial effects of keratoplasty, aided perhaps by cortisone. The visual improvement in three of the four eyes was satisfactory, though in one it was not maintained. This, together with relief from pain, photophobia, and lacrimation, has certainly made the lot of these patients happier.

One cannot say whether this improvement will be permanent. As the

graft is replaced by the host tissue, it is probable that the area may succumb to the pathological process. Perhaps a total replacement of the host cornea in gradual stages by donor material would render the 'new' cornea immune to photosensitization. Two of the grafts reported above have remained transparent for 18 months, but with marked corneal vascularization the chances of maintaining transparency are remote. Opacity due to photosensitization may be delayed until the whole graft is replaced by host tissue.

The fact that grafts take well and even remain clear for some time, suggests that there is no neural cause for this condition (Duke-Elder, 1938). Corneal grafts can survive for months before innervation takes place, although in certain states of denervation or anaesthesia of the cornea severe ulceration may occur.

The present results show that there is hope of giving some measure of relief for variable periods of time, and that these could be extended by repeating the operation as long as donor material is available.

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