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Cosmetic management of the sequelae of thorium X radiotherapy for naevus flammeus

Naevus flammeus is a benign cutaneous angioma which often appears on the face causing cosmetic concern. Earlier this century, radiotherapy was frequently employed to try to remove the lesion. Thorium X, an alcohol-based paint was a favoured radiotherapeutic agent. Unfortunately, thorium X therapy has side effects including carcinogenesis.

The case of a 62-year-old woman who suffered gross neoplastic dermal disease as well as the full gamut of ophthalmic radiation related disease after thorium X radiotherapy for naevus flammeus, is presented. The management of this patient included extensive plastic surgery and prosthetic contact lens fitting.

Discussion

The history of radiotherapy for naevus flammeus and other angiomata leaves a striking message about the risks of long-term sequelae of new treatments. Current treatment for naevus flammeus is also controversial. This case demonstrates the ocular and dermal sequelae of radiation injury and how it can be managed. The process of prosthetic contact lens fitting is considered in detail. The outcome of this process is a dramatic improvement in the patient's self-confidence and quality of life.

Introduction

Naevus flammeus, or port wine stain is a congenital cutaneous angioma that appears as a flat red to purple lesion. The lesion is due to vascular ectasia and does not spontaneously regress. Naevus flammeus most commonly appears over the distribution of the first and second divisions of the trigeminal nerve.

In this location, naevus flammeus, if associated with leptomeningeal angiomatosis with calcification, is part of the Sturge-Weber syndrome (encephalotrigeminal angiomatosis).

The cerebral lesions may cause epilepsy, hemiparesis, and hemianopia and up to 60% have a degree of mental deficiency[1]. No relationship exists between the size of the facial naevus flammeus and clinical neurologic impairment[1]. Naevus flammeus alone is often unilateral, but may be bilateral in up to 30% of cases[1]. The lesion may extend across the mid-line and down to the upper trunk or arms.

This condition is usually of interest to eye care practitioners because 30% develop glaucoma and 40% exhibit choroidal angioma[2]. In this case, it was the treatment of naevus flammeus, which has led to the need for eye care.

Figure 1
Facial view showing extensive scarring and recurrent carcinoma

Figure 2
Facial view after several excisions and autografts

Radiation has been used to treat many benign vascular dermatoses since the turn of the century[3,4]. Numerous sources of radiation were experimented with including X-rays, radium and thorium[5]. Thorium X had won favour for the treatment of naevus flammeus and similar conditions by the end of the fourth decade[6]. However, concerns were soon being raised about its safety[7]. By the end of the seventh decade thorium X treatment was discontinued due to poor results and concern about neoplastic side effects[8,9].

Presented below is a case report of a patient who suffered the disfiguring atrophic and neoplastic effects of thorium X radiotherapy used in the treatment of naevus flammeus.

Case report

This is the case of 62-year-old woman born with unilateral (left) naevus flammeus. She was first treated, between the ages of three and five, at Great Ormond Street Hospital in London with "snow". This was either liquid CO_2 or liquid nitrogen. Treatment was interrupted by the Second World War.

In 1945, treatment with thorium X was started. Over the subsequent nine years, the patient had more than 50 "paintings" with thorium X. The frequency of these treatments reduced until they were stopped in 1954. The patient recalls trying to be convinced it was helping, but in hindsight she feels the thorium X "did not help at all".

The patient emigrated to Australia in 1972. Little then happened until the late 1970s, when small bleeding sores appeared on her left cheek. These were removed, but other lesions soon followed. Over the subsequent 15 years the patient had dozens of lesions excised. These were later diagnosed as basal cell carcinoma (BCC).

By 1995, carcinomas were growing as fast as they could be removed. The patient had a "patchwork" appearance of lesions and excision scars as well as atrophic telangiectatic skin (**Figure 1**). This caused poor cosmesis on the left side of the face and the need for a more permanent solution. Total excision of the damaged area with autografting (stomach skin for the cheek and neck for the eyelids) was commenced (**Figure 2**).

The cosmetic improvements were completed with argon laser photocoagulation closure of telangiectatic vessels on the nose and elsewhere on the adjacent host skin.

Figure 3
Left eye after lid reconstruction. The eye is phthisical with cicatrising conjunctival disease, conjunctival hyperaemia, limbal failure and corneal decompensation

Figure 4
Left eye showing gross ocular surface disease including calcification

Figure 5
Facial view at the conclusion of plastic surgery. The main focus of poor cosmesis is now the left eye

During the 1980s, ocular problems began. By 1995, she exhibited the full spectrum of ocular radiation damage: pthisis bulbi, limbal failure, ocular surface disease, chemotic cicatrised conjunctiva and no perception of light (**Figures 3 and 4**). The conjunctival shrinkage and lid reconstruction resulted in abnormal lids with poor blink function and incomplete closure nasally (**Figure 5**). The ocular appearance still gave awful cosmesis despite an improved facial appearance.

The next step was prosthetic contact lens fitting. Keratometry was not possible on the opaque, calcified and irregular left corneal surface. As a starting point, keratometry of fellow eye was measured: 46 (7.3) x 10 47.25 (7.1) x 100. Assuming the two eyes were once the same and fitting slightly steeper to allow for phthisis and lack of need to consider physiology, the following lens was ordered:

7.9/14.00/plano 38% water content with a hand painted opaque tint.

The lens colour was ordered using slit-lamp and external eye photographs (**Figure 5**) as a guide. The size was matched to the right eye: HVID 10.50, pupil 4.00 opaque. Outside the painted iris, the remainder of the lens was painted white to give a scleral appearance over some of the chemotic conjunctiva. The lens is shown in **Figure 6**.

The lens fitted satisfactorily and moved well in all directions of gaze (**Figure 7**). The patient was delighted with the improvement (**Figure 8**). She felt more confident with the lens in and claimed she "would not be without it now" and she "feels like a different person". Particularly, she noted that children did not stare and comment as much as before especially when catching the bus or in other similar situations. Additionally, she reported that while the lens was in, the eye felt more comfortable. This is most likely due to a bandage lens effect preventing desiccation.

Quality of life was measured before and after lens fitting using the Cantrill Ladder[10]. This is a very simple and quick test that yielded a score of 2 before 9 after (Maximum range 0 to 10). This change represents a highly significant improvement in quality of life[10].

Figure 6
Prosthetic contact lens
(Capricornia Contact Lens Company)

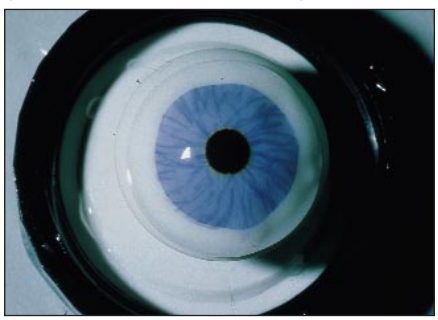

Figure 7
Left eye with prosthetic contact lens in situ

Figure 8
Facial view demonstrating improved cosmesis

Discussion

Although naevus flammeus has long been known to be a benign lesion, its poor cosmesis has driven attempts to remove the lesion. Hartigan was the first to report treatment of naevus flammeus with radiotherapy[3]. He reported a series of patients treated with an average of 39 exposures to unscreened radium[3]. Improvement was claimed in most cases, but most cases also suffered superficial ulceration[3]. Others noted telangiectasia and atrophy as sequelae of radium radiotherapy[11]. Nevertheless, radiotherapy prospered with numerous sources trialled including X-rays, grenz rays and thorium[5]. However, this treatment became very controversial.

In 1946, McKee and Cippollaro wrote: "To cause an obliterating endarteritis in telangiectasia, spider naevus and port-wine mark, requires an amount of treatment that will seriously injure normal tissue"[7]. They went on to suggest that there was no role for X-rays, radium, thorium or other sources of radiotherapy in the treatment of vascular dermatoses[7]. Others felt thorium was a preferred radiation source, because it chiefly produces a-rays that penetrate poorly and do not cause significant radiodermatitis[9].

Thorium X was applied as an alcohol-based paint usually using a cotton bud (hence, the term "paintings")[12]. Thorium X was used for the treatment of vascular naevi of many types for 30 years until it was discontinued due to poor results and concern about neoplastic side effects[8,9].

The long-term neoplastic potential of medical irradiation is now well established[9,13-15]. The legacy of treatments used earlier this century remains one of the most striking lessons in medicine on the risks of the unknown. However, a-Rays have a relatively low carcinogenic risk. Nevertheless, several cases of BCC as a late complication of thorium X treatment for naevus flammeus can be found in the literature[15-19].

The current treatment of naevus flammeus is still controversial, although perhaps not as it was in the 1950s. Laser, particularly the flash

lamp pulsed tunable dye laser rather than argon or ruby lasers, has become the intervention of choice[20]. However, many do not advocate intervention if it is possible to avoid it. Opaque make-up, such as Covermark is very effective, tattoo camouflage less so. Other possible interventional approaches are dry ice (as used initially in this case), cut and suture, excision with free flap or autograft (using the patients own tissue) repair[21].

This case not only demonstrates the neoplastic, angiogenic and carcinogenic dermal sequelae of radiotherapy, but also the ophthalmic effects of radiation injury. The left eye exhibits every possible sequelae to radiation insult: phthisis bulbi, limbal failure, corneal decompensation, cataract, ocular surface disease, chemotic and cicatrised conjunctiva all resulting in no perception of light. A suitable case for cosmetic improvement using a prosthetic contact lens.

The case also demonstrates some interesting aspects of fitting contact lenses to diseased or damaged eyes. In such eyes, keratometry or corneal topography often cannot be performed due to the state of the corneal surface. Often prior to the disease or injury, both eyes were the same dimensions. This can be checked with a thorough history.

If both eyes were the same, then the fellow eye can be measured for corneal shape. Adjustments to this result can be made to allow for the impact of the disease or injury:
eg flattening over a penetrating injury or, as in this case, steepening due to phthisis bulbi. Trial lens fitting can follow this starting point.

Prosthetic contact lenses should often be fitted a little steeper than normal in eyes without vision and any corneal oxygen supply needs. A tighter fitting lens is less likely to give the strange appearance of a dislocated lens and will centre better in different positions of gaze. Of course, in cases of sighted eyes with high corneal physiological requirements such as a motor vehicle accident victim who is left aphakic and aniridic, the lens should be fitted more loosely.

This patient could have been fitted with several other modalities which are not necessarily inferior to a painted hydrogel lens. It could be argued that a painted scleral lens or even an artificial eye style scleral shell would give a superior cosmetic result since both would cover more of the chemotic conjunctiva replacing it with a white scleral appearance. However, both types, especially the latter, would not move as well during gaze movements. Moreover, cicatrising disease and lid reconstruction had left this patient with minimal fornices thus making lens retention extremely difficult. Hence, a hydrogel lens was chosen. However, other modalities should always be considered in such cases.

This case demonstrates the rewarding nature of prosthetic contact lens fitting. Coupled with plastic surgery, this woman has overcome a
life-long affliction. This has caused an enormous change in her psychological well-being and quality of life - an outcome which, in turn, has enhanced this practitioner's quality of life also.

Conclusion

This case illustrates the interdisciplinary management in a case of facial disfigurement following thorium X radiotherapy for naevus flammeus. The optometrist, ophthalmologist and plastic surgeon all have an important role in the management of this case. Following extensive skin excision and autografting surgery and lid reconstruction, a prosthetic contact lens was fitted. This process resulted in an improved cosmetic appearance with a consequent improvement in patient confidence and quality of life.

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About the Author

Konrad Pesudovs is has recently taken up a post doctoral appointment funded by the NHMRC through a Sir Hamilton Fairley Fellowship in the Department of Optometry at the University of Bradford. He is the first optometrist ever to win this prestigeous fellowship.
He was previously a clinical researcher in the Department of Ophthalmology at Flinders Medical Centre, Australia.

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