SOME DISEASES INVOLVING THE EYE AND THE SKIN

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SUMMARY

Diseases involving the skin and the eye are so numerous that only certain selected aspects can be considered here. For a complete list the reader is referred to a review by **Beerman** et al.[1] *Those abnormalities to be discussed here, and which may present to ophthalmologists, are divided into two broad groups*: (*a*) *skin conditions that may be limited to the eye and eyelids, and* (*b*) *systemic diseases with skin and eye manifestations.*

SKIN CONDITIONS THAT MAY BE LIMITED TO THE EYE AND EYELIDS

These conditions can be grouped in the following categories: (*i*) congenital naevi—vascular, melanocytic, epidermal; (*ii*) traumatic—bites and stings and lice; (*iii*) inflammatory—bacterial (boils, styes, impetigo), viral (warts, molluscum, herpes simplex, zoster), granulomatous (tuberculosis, sarcoid) or non-specific dermatitis (contact, seborrhoea, rosacea); (*iv*) neoplastic—benign (seborrhoeic warts, skin tags, syringoma, kerato-acanthoma) or neoplastic (basal, squamous, melanoma); (*v*) miscellaneous—alopecia areata.

1. Congenital Naevi

Vascular naevi present at birth, or soon after birth, and are of two broad types. There is the naevus flammeus, which will be discussed later, and the capillary cavernous haemangioma. This latter continues to enlarge until about the age of 7 years when it then naturally involutes. If left untreated, therefore, it can occlude the sight of the eye, resulting in a blind eye. If, however, this lesion occurs elsewhere, it is best left alone, but when it interferes with vision, then treatment (X-radiation or surgery) becomes imperative.

Melanocytic naevi. The melanocyte is derived from the neural crest and arrives in the epidermis between the 3rd and 4th month of foetal life. The lids divide into upper and lower lids between the 4th and 6th month of foetal life and therefore any naevi (excessive collections of

melanocytes) that are in this position become divided—hence the term divided naevi.

In the naevus of Ota there is a hold-up of melanocytes in the dermis and it is composed of deeper bluish and more superficial brownish elements. In the eye the affected sclera is blue and the conjunctiva brown. The areas involved are the eyelids, the bulbar and palpebral conjunctiva and the sclera and the fundus of the eye, the cheeks, forehead, scalp and the mucous membrane of the mouth. The distribution seems to be restricted to the areas innervated by the 1st and 2nd division of the trigeminal nerve. The discoloration persists throughout life (unlike the Mongolian blue spot) and only very rare instances have developed into malignant melanoma.

Epidermal naevi. These are hamartomatous overgrowths of the superficial epidermis causing warty lesions which often show linear configuration. They may appear on the face and eyelids. Unfortunately there is no efficient method of treating these disfiguring lesions.

2. Traumatic

Bites and stings, especially of mosquitoes and wasps, can lead to oedema and ulceration of the upper eyelid—occasionally causing severe destruction. The nits of the body louse can collect on the eyelashes, causing irritation and infection.

3. Inflammatory

The infection of the skin and hair follicle with the streptococcus and staphylococcus leads to boils and styes and impetigo. Viral warts and molluscum contagiosum involvement are likewise common. The lesions of the latter are typically umbilicated smooth-edged papules, which give a cheesy material when expressed. They can be removed by scraping with a curette. The lesions are superficial, no anaesthetic is necessary and no bleeding occurs. However, because of its infectious nature, new lesions are liable to occur.

Herpes simplex—particularly of the recurrent variety—is liable to occur on the eyelids and affect the underlying conjunctiva. Typically these are grouped vesicles on an erythematous background. However, on the eye these are usually atypical and look like an impetiginized eczema. These lesions are infectious.

Herpes zoster, when it affects the trigeminal nerve, can cause involvement of the eyelids and the eye. With involvement of the 1st division, involvement of the forehead, upper eyelid and the conjunctiva may occur. Not all the branches need be involved at the same time, but it is important to know that when the nasociliary branch is involved—and this may on occasion be the only branch involved—then the vesicles and ulceration of the tip of the nose are evident. Conversely, when the tip of the nose is involved, then the ciliary body is also involved. Herpes zoster is an infectious disease, due to the presence of the chicken-pox virus in the posterior nerve root and in the terminal nerves in the skin lesions. Paralysis of the ocular muscles occurs in about 7% of cases of ophthalmic herpes zoster. Further, while herpes zoster in general is benign, it may have serious complications when it occurs in the elderly patient or when it involves the eye. Early oral corticosteroid therapy is advisable—60 mg daily for the first week and then reducing to 30 mg and then 10 mg in successive weeks.

Granulomatous inflammatory conditions. Tuberculosis involves the eyelid with ulcerated nodules which become confluent and lead to plaques with eventual atrophy.

Sarcoidosis involves the eye and is the presenting sign in about 20% of patients. In the eye itself it can cause a uveitis or iridocyclitis. There may be associated papules and plaques in the skin and these must be searched for. They are of various sizes and shapes but are 'juicy', rubbery, red to brown lesions, and when they occur in the inner canthi of the eyes, they are diagnostic. Associated findings are lymphadenopathy, hepatosplenomegaly, infiltration of the lungs and cystic lesions of the bones of the hands. The tuberculin test is negative and the Kveim is positive.

Non-specific dermatitis. Patients with rosacea complex may present to the ophthalmologist with episcleritis and keratitis. There is a blotchy erythema of the face, with papules, pustules and telangiectasia. The eyelids may be involved.

The eyelids may be involved in contact dermatitis and in seborrhoeic dermatitis. It is important to realize the existence of contact dermatitis of the eyelids and one must be continually in search of the causal agent or agents. These are numerous and the groups that one must remember are the cosmetics (lanolin, nail polish, lacquer, rarely the eyeshadows); medicaments (eye ointments, eyedrops); and plants (primula, smodingium) and metals (nickel). I recently saw a patient with long-standing blepharitis where the cause was lanolin used cosmetically and in the prescribed ointment. The vehicles such as ianolin, paraben, chlorocresols and ethylenediamine used to contain the medicament in the ointment compounding are liable to cause a dermatitis.

4. Neoplastic

All types of growths can involve the eyelids. These can be benign, such as seborrhoeic warts, with their typical warty appearance, yellowish brown and fragile, which can be separated from the skin with the fingernail; syringoma—hundreds of small, soft, slightly yellowish nodules, pinhead in size; and kerato-acanthoma—a dome-shaped nodule with a central crater and smooth edge, sudden in onset and of rapid growth—difficult to distinguish from squamous carcinoma. The lesion is self-healing.

The malignant lesions include basal-cell and squamous-cell carcinoma and malignant melanoma, all of which have the same characteristics as similar lesions elsewhere on the body.

5. Alopecia Areata

This patchy loss of hair of unknown causation usually involves the scalp as well as other areas. When the scalp is involved then the prognosis is good in the first attack. When the hair elsewhere is involved, such as the eyebrow, the chance of recovery is less certain.

SYSTEMIC DISEASES WITH SKIN AND EYE MANIFESTATIONS

The following 6 categories will be discussed: (*i*) the phacomatoses; (*ii*) other heredito-congenital abnormalities; (*iii*) metabolic and endocrine disorders; (*iv*) oculo - mucous-membrane - cutaneous syndromes; (*v*) auto-immune disorders; and (*vi*) atopic dermatitis.

1. The Phacomatoses

This term implies 'spots' of congenital origin in various tissues and includes tuberous sclerosis, neurofibromatosis, naevus flammeus with glaucoma (Sturge-Weber syndrome). This group of disorders is characterized by congenital retinal tumours, all have skin manifestations and all have an autosomal-dominant inheritance (doubtful in the case of Sturge-Weber).

Tuberous sclerosis. These present to the ophthalmologist with retinal tumours, small round, flat growths or mushroom forms at the nerve head. Raised intracranial pressure with optic atrophy has been recorded. The skin lesions are those of adenoma sebaceum, predominantly on the 'butterfly area' of the face—these are red, vascular, telangiectatic papules, and skin-coloured fibrotic nodules which may be polypoid. Further, these may be leathery fibrous plaques (shagreen patches), mainly on the back, periungual fibromas, *café au lait* pigmentation, achromic areas (with ash-leaf appearance) and radiologically nodular cerebral calcifications.

Multiple neurofibromatosis (Von Recklinghausen's disease). This may present to the ophthalmologist with intra-orbital plexiform neuromas, pulsating exophthalmos and optic nerve gliomas, and the dermatological features in such a case help to solve a difficult problem. These are *café au lait* marks—pale yellow-brown macules with a definitive relatively smooth border; the presence of 6 or more, 1·5 cm or more in diameter, is diagnostic. Axillary freckling, too, is a sign of neurofibromatosis. Skin neurofibromas may also be present.

Sturge-Weber syndrome. This consists of angiomas, convulsions, paralysis, mental retardation and visual disturbances. All of these manifestations may not be present in any one patient. There is haemangiomatous involvement of the eyelid and uveal tract, and the patient may present with glaucoma. The skin manifestations are cutaneous angiomatosis—purplish to red macules varying in size from a few square centimetres to very large areas. These angiomata may be widespread over the body or limited to the face only, and even only to the forehead and upper eyelid. There is no relationship between the extent of the naevus and degree of meningeal involvement. X-ray of the skull may reveal sinuous parallel streaks of calcification.

2. Other Heredito-Congenital Abnormalities

Pseudoxanthoma elasticum. The ocular findings may occur independently of skin lesions, and consist of retinal angioid streaks. Vision may be impaired by haemorrhages, choroidal sclerosis, exudates, clusters of dark pigment and macular degeneration.

The skin clues are seen on the face, neck, axillary folds, inguinal folds and umbilical area. The lesions are yellowish, chamois-coloured papules which merge giving a coarse leathery feel, and a roughened 'plucked chicken' or 'lemon peel' appearance. It looks like prematurely aged skin in patches.

Albinism. The ocular findings are lack of pigmentation, nystagmus and high myopia. This lack of pigmentation is reflected in the skin and hair, photosensitivity and the development of carcinomas and melanomas.

Xeroderma pigmentosum. The ocular manifestations of this entity are conjunctival injection, keratitis ectropion and retinoblastoma. The skin manifestations are erythema, freckling and hyperpigmentation progressing through atrophy to carcinoma.

3. Metabolic and Endocrine Disorders

Xanthelasma palpebrarum may be associated with xanthomata elsewhere.

Ochronosis gives a brown fleck in the conjunctiva and is associated with bluish pigmentation of fibrous tissue, cartilage of ears and tip of nose and axillae.

Lipoid proteinosis presents with a hoarse voice from birth, papules on the lid margins and disfiguring scarring skin lesions, especially on the face.

Amyloid disease can present with papules round the eyes and infiltration of tongue and palms—usually associated with multiple myelomatosis.

Hyperthyroidism can present with circumorbital hyperpigmentation.

Menstruation can be preceded by circumorbital cyclic hyperpigmentation.

4. The Oculo-Mucous-Membrane-Cutaneous Syndromes

The erythema multiforme syndromes can present with conjunctivitis and blepharitis and can lead to gross scarring of the palpebral conjunctiva. The typical target-like skin lesions on the body, extremities, palms and hands can follow. When severe, this condition can take on a 'scalded man' appearance.

Reiter's syndrome also presents with conjunctivitis, balanitis, urethritis and arthritis. Occasionally there are skin lesions only, erosions of the mouth and dull red macules (on the skin and soles of feet and palms of hands) which develop into cone-shaped hyperkeratosis (keratoderma blenorrhagica).

5. Auto-immune Disorders

The ophthalmologist may be presented with eye involvement of the following disorders:

Systemic lupus erythematosus or the eye complications of 'antimalarial' drug therapy. These latter are corneal oedema, retinopathy, episcleritis and post-subcapsular cataracts. The skin lesions are numerous, but are all manifestations of a cutaneous vasculitis and are seen over the bridge of nose and cheeks (so-called butterfly area), fingertips and elsewhere.

Dermatomyositis can present with periorbital violaceous erythema and oedema, dermatitis on the knuckles and muscular (girdle) weakness.

6. Atopic Dermatitis

This is often associated with cataracts in the younger age-group, particularly if they have been on oral corticosteroids. The eczematous skin lesions can be widespread, but are particularly seen in the flexures of the elbows and the knees. Usually there is an associated asthma and hayfever.

CONCLUSION

The ophthalmologist will find in examination of the skin many clues in the diagnosis of disorders he is called upon to manage.

REFERENCE

1. Beerman, H., Kirshbaum, B. A. and Cowan, L. K. (1959): Amer. J. Med. Sci., **238**, 491.