COMMUNICATIONS

PERI-ORBITAL TUMOURS*

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I FULLY realize the honour the Institute of Ophthalmology has paid me in asking me to give this address to-day. I wondered how a general surgeon could interest listeners who have devoted so much of their time to the study of ophthalmology. Many years ago I was ophthalmic house surgeon to Mr. Cargill and Mr. Lyle, and it was during that time that I first became interested in peri-orbital tumours, a subject which still is full of curiosity as far as I am concerned, and is one which embraces a wide field of pathology, coming within the spheres of the ophthalmologist, general surgeon, neurosurgeon, and dermatologist.

The commonest types of tumour around the eye are those which occur in the skin or subcutaneous tissues. The basal and squamous-cell carcinomata need little introduction, being well known by reason of their frequency, and differing pathologically in no respect from those occurring elsewhere.

RODENT ULCERS.—These grow by predilection in the skin of the lids, the supra-orbital region, both canthi, and the skin of the cheek, when compared with their incidence in other parts of the body (Figs 1 and 2). They are estimated roughly to be ten times more frequent than squamous-cell carcinomas. They arise from the deeper layers of either the epidermis or the sweat glands and present the characteristic microscopical appearance of

FIG. 1.—Rodent ulcer at the inner canthus in a man aged 68. This ulcer was excised.

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FIG. 2.—Rodent ulcer in a woman aged 88 with ulcer present for 12 years. No treatment prior to excision.

ramifying and often pointed processes which invade the dermis and subcutaneous tissue. Sometimes in microscopical sections the connection with the basal layer of the epidermis can be made out, while in others continuity with the pilo-sebaceous organs is demonstrable. The peripheral layer of cells resembles columnar epithelium, while the more central cells are irregular and contain a considerable number of mitoses. The amount of stroma is variable, being relatively large and fibrous in the more chronic type and embryonic in the more malignant ones.

Rodent ulcers tend to occur after the age of 40 years and like epitheliomata are often preceded by hypertrophic changes in the affected area of epithelium which results in the formation of a prerodent keratosis, firmer and flatter than that of a squamous epithelioma. The reason for this predilection for the face is no doubt due to its exposure to a number of factors believed to be connected with carcinogenesis, such as trauma, and exposure to the weather and to the sun's rays, and in some cases to products of coal tar. All forms of solar or senile keratosis should be regarded with suspicion and treated as precancerous.

The natural history of a rodent ulcer varies from case to case. The first-noticed lesion may be a pimple or a small red nodule. Over a period of from 1 to 30 years the lesion grows slowly. For some years there is no ulceration, but eventually the epithelium over the lesion breaks down and, after abortive attempts at healing, a chronic ulcer remains with a heaped-up, rolled-over edge which is indurated and even hard to the touch. The clinical appearances are not always uniform. A useful classification of the different types is that by Sequeira into superficial cicatrizing, non-cicatrizing or the typical rodent ulcer, and the terebrant type with rapid spread.

TREATMENT.—This has been a subject of controversy, the choice lying between surgical excision and irradiation. Both may play a part but certain points demand consideration.

Site.—Almost all rodent ulcers can be excised, many quite simply with a margin of healthy tissue, others requiring a variety of plastic repairs including free skin grafts or flaps, but all within the scope of the general surgeon (Fig. 3). In some regions around

the eyelids surgical treatment may be difficult, but treatment by irradiation demands care to neighbouring vital structures such as the eye.

Surface Area of Lesion.—Small lesions are naturally the more readily excised, but these are also more suitable to treatment by irradiation. Skin defects left by surgical excision of large tumours are readily closed by graft or plastic repair. On occasion the cosmetic effect may be better from irradiation of large areas, but greater exposure with its attendant disadvantages is necessary.

Depth of Penetration of Lesion.—The depth of penetration is always difficult to determine accurately. The more superficial a lesion the more accessible it is to irradiation by either radium or x-rays. But superficial lesions are also readily treated by surgery. Surgical excision is always the treatment of choice where penetration into the bone or the tarsal plates is deep.

Age and State of Health of Patient.—Advanced age is not a contraindication to surgery. If the patient's state of health is poor, local analgesia can almost always be used.

A few additional points of contrast between surgery and irradiation may be made. Whereas one operation is required when surgery is practised and the extent of the necessary excision is gauged, for irradiation, multiple attendances are often necessary. Owing to uncertainty of depth of involvement, irradiation may fail to destroy the deeper layers of cells, and some cells may be resistant to irradiation. The peripheral extent of the growth may not be appreciated without surgical excision and thus an adequate area of tissue may not be irradiated.

The complications of surgical excision of rodent ulcers are negligible. Irradiation, however, may produce dermatitis followed by telangiectasis and atrophy or damage to vital structures such as the eye.

By surgical excision confirmation of the diagnosis is obtained. It has been shown that 13 per cent. of skin carcinomata diagnosed clinically as basal-celled epitheliomata contain prickly cells. These mixed or basal-squamous epitheliomata are more radio-resistant than pure basal-cell tumours.

Recurrences occur in about 5 per cent. of cases treated with x rays, but with adequate surgical excision there should be no recurrence. With regard to time lost from work, the advantage is with surgery; in many cases rodent ulcers may be treated in the Outpatient Department under local anaesthesia. With surgery there is no danger of failure to complete the treatment as there is with the repeated visits often necessary with radiation.

Lastly, in the treatment of recurrences after failure of irradiation, surgery is the only satisfactory choice, recurrence tending to be more radio-resistant than the primary growth.

PAPILLOMATA.—These may occur in the para-orbital region and if not removed are likely to become malignant. Such papillomata consist of projecting papillae undergoing proliferation and frequently branching to form secondary papillae. If the epithelium undergoes keratinization as in common

FIG. 3.—Large rodent ulcer which required excision and a plastic operation to cover the defect left after excision.

warts, these become hard and may constitute horn-like outgrowths. Haemorrhage from such a papilloma is a sign of malignant invasion and always demands radical excision. It is pathetic to see these papillomata allowed to progress towards malignant disease before radical treatment is entertained. It is surprising that members of the public will tolerate large papillomatous masses in the para-orbital region (Fig. 4).

FIG. 4.—Papilloma of the inner canthus.

FIG. 5.—Melanoma of the outer canthus.

SQUAMOUS-CELL CARCINOMATA.—Epithelioma is a new growth of squamous cells often occurring close to the lid margin in contradistinction to rodent ulcer. It infiltrates and destroys local tissue and so forms a malignant ulcer. The constituent cells are large and of irregular shape, forming a variable number of cell nests according to the grade of differentiation and therefore of malignancy. Fortunately, epitheliomata in this situation are not often of a high grade of malignancy and glandular metastases are therefore infrequent except in neglected cases. The treatment is in many respects similar to that for rodent ulcers with a choice of excision or radiotherapy. If radiotherapy is used, it is important to perform a biopsy in order that the diagnosis may be confirmed and the grading known. Surgical treatment is in many cases more satisfactory. The treatment of the lymphatic glands depends upon their clinical state when first seen and upon the age of the patient. Should they be thought to be involved, then a block dissection is considered by many at the present time to be of greater curative value, but irradiation is less disturbing to the patient, particularly in the elderly.

MALIGNANT MELANOMATA.—The third type of malignant tumour of the skin is the malignant melanoma. This may occur around the eye just as in other parts of the body. Bleeding, increase in size, or pain occurring in a previous mole are ominous signs (Fig. 5). It can hardly be stressed too often that any local interference with a pigmented naevus is dangerous for fear of stimulating actual malignant degeneration. When this has occurred no biopsy should be performed, but instead a wide excision should be made to include the tumour, a good margin of healthy tissue, and the deep fascia. These tumours are radio-resistant; they tend to spread by the lymphatics

first but also widely by the blood stream at a variable period of time after onset. Although the prognosis is generally regarded as bad a few cases live for many years, especially when adequately treated. There seems to be no way of distinguishing these and the histology is of no help.

BENIGN LESIONS.—Of benign lesions in the peri-orbital region there are only a few which call for brief attention. Simple papillomata occur frequently in the skin of the eyelids, and are formed by several layers of squamous epithelium covering a vascular stroma. Excessive keratinization forms one variety of cutaneous horn and malignant degeneration is rare. Xanthoma palpebrarum, a deposit of yellow lipid in large polyhedral cells beneath the skin, is sometimes large enough to draw the patient's attention. Both these conditions are easily excised.

HAEMANGIOMATA.—Haemangiomata are the commonest tumours to occur in the peri-orbital region in childhood, being congenital in origin but tending to increase in size during the first few years of life. They are not, therefore, strictly tumours, with the exception of the less common hypertrophic or angio-endotheliomatous type, but congenital malformations of embryonic vaso-formative tissue. They are presented for medical attention because of the disfigurement they cause, being either of the superficial capillary type or of the deeper and more spongy cavernous type.

Superficial.—It is common knowledge that in a proportion of superficial angiomas, and particularly, in the "strawberry birthmark", spontaneous thrombosis occurs if left long enough. In order to initiate or promote this curative phenomenon a variety of procedures such as cauterization with CO₂ snow, electrocoagulation, and irradiation with x-rays, radium, or radon seeds is in current use. In some cases surgical excision is always more satisfactory being quickly performed without fear of complication and leaving a thin linear scar. These methods are as a rule ineffective in dealing with the deeper subcutaneous and cavernous angiomas and particularly with the advanced type which has acquired an extensive blood supply and now amounts to a cirroid aneurysm. These methods are not only ineffective but also better avoided, because of the importance of maintaining an unbroken epithelial covering, since loss of viability may lead to sloughing, infection, and haemorrhage with serious risk of grave consequences.

Cavernous.—The useful methods of treating cavernous angiomas are surgical excision, ligation of feeding vessels, injection of sclerosive chemicals, and coagulation by injection of boiling water. In many cases simple surgical excision is effective provided that the angioma is suitably situated, certain types requiring subsequent plastic repair. The size of large angiomas, especially those with deep ramifications, may be reduced by preliminary injections of irritating solutions such as 40 per cent. saline or glucose, ethamolin, or boiling water. Reduction in size from thrombosis follows and surgical excision may then be facilitated. The same reduction in size or even complete thrombosis can sometimes be effected by the implantation of radon seeds.

Ligation of the vessels feeding the angioma is never necessary in the absence of pulsation but is sometimes useful as an adjunct to sclerosis and where surgical

excision is impossible as when both eyelids are involved. It is necessary to remember that the vessels feeding a haemangioma arise from the blood vessels supplying the part concerned, but that the blood passing through the angioma is a circulation by itself and independent of adjacent tissue. When the angioma is arteriovenous in type the shunt of blood may on occasion be so great that the quantity available for nourishing neighbouring tissues is reduced even to the extent of producing ischaemic lesions.

INVOLVEMENT OF NEIGHBOURING TISSUE.—The subject of haemangiomas is a suitable one by which to pass from the more common superficial tumours to the less common deep ones involving the orbital walls or neighbouring tissues. Angiomas of the bony walls of the orbit are uncommon but well known, and occur in capillary and cavernous forms. The typical radiographic appearances of radiating trabeculations give considerable help in making a firm diagnosis.

Although uncommon in themselves, angiomas may be reckoned among the more common causes of exophthalmos by the creation of a space-occupying lesion posterior to the globe. The following types are recognized:

- (a) Capillary.
- (b) Cavernous.
- (c) Angioblastic or hypertrophic.
- (d) Racemose or cirroid.

The angioblastic type is firm, being composed of a solid mass of endothelial cells with a minimum of small patent vessels, the lumina becoming obliterated by the proliferation of embryonic epithelium.

The racemose type is composed of a pulsating mass of dilated, thickened, and tortuous vessels. They occur in adults and have a relatively sudden onset. They may represent an ordinary cavernous angioma which has established a communication with a neighbouring artery or arteriole. After this communication has been established the mass becomes locally destructive by reason of constant arterial pulsation. These tumours may erode the walls of the orbit and surrounding structures and even lead to fatal haemorrhage.

A characteristic of haemangiomas, except those of the cirroid type, is that they do not show unlimited growth but tend to reach a certain size and then remain stationary. This is an important point when considering treatment in a difficult case.

Two noteworthy characteristics of this type of 'tumour' in the orbit are:

(1) The mobility of the eye is unaffected.

(2) The variations in size may result in exophthalmos. These variations may be spontaneous and may be due to simple venous stasis from coughing, crying, bending the head down, or compression of the jugular vein.

Sometimes the haemangiomatous nature of the lesion is revealed by an extension forwards rendering it visible from the exterior in a subconjunctival position, or else by the association of a port-wine stain around the eye. When considering treatment, the position, size, and type of angioma must be taken into account and many of the principles discussed for superficial angiomas applied.

DERMOID and EPIDERMOID TUMOURS.—Dermoid and epidermoid tumours of the orbit are common if the para-orbital type is included, but although well-known, intra-orbital dermoids are not frequent. They are found in many situations—external, intra-orbital, intracranial, and in the diploë.

FIG. 6 (a).—External angular dermoid (front view).

FIG. 6 (b).—External angular dermoid (lateral view).

External.—The commonest site is the lateral end of the eyebrow where they are known as external angular dermoids (Fig. 6 *a* and *b*). In this situation they give rise to cysts either tense or soft, which are not attached to the skin but which characteristically form a depression in the underlying skull which may be felt clinically. The depression sometimes amounts to a defect with firm attachment of the epidermoid to the dura (Fig. 7).

Intra-Orbital.—These dermoids occur most commonly in the superior temporal region, causing proptosis and requiring to be distinguished from the less common tumours of the lacrimal gland. In this situation a defect may occur in the bone in the form of either a depression or a foramen which is easily demonstrated in the x-ray.

Other Sites.—Intracranial extension sometimes occurs, and some cysts appear to arise in the diploe of the frontal bone extending into both orbit and cranium.

The treatment for both dermoids and epidermoids is the same and where possible consists of excision. In the deeper and larger varieties of intra-orbital cyst this may not be an easy matter because of the involvement of neighbouring structures and cavities demanding transfrontal approach. In others the cyst may first be incised with evacuation of its contents and then its wall may be removed piece-meal.

A few words are required regarding the terms dermoid cyst and epidermoid cyst. Although they are closely related in origin and behaviour and require similar treatment, much has been written of their differentiation. The term epidermoid has replaced the older cholesteatoma or *tumeur perlée*; the differentiation between the two is perhaps academic, since both are derived from embryonic nests originating from the primitive ectodermal layer, the actual differentiation depending upon either the time at which the cell nest

FIG. 7.—Mid-line dermoid at base of nose.

developed or the degree or depth of inclusion of the original dermal layer. It was Renak in 1854 who first suggested that the 'pearly tumour' of Curveillier originated in ectodermal nests, and this hypothesis has been very strongly supported by subsequent observers. The evidence, however, rests entirely on the histological features of the tumour itself and particularly on the details of the capsule.

The ectodermal layer of the head end of the foetus develops into both skin and neural tissue. The cause for differentiation of this one layer is speculative but it has been suggested that what is known as "organizer influence" is excreted by the mesodermal tissue or cells which later become neural tissue. Jefferson believes that the epidermoids found in the brain substance or within the membrane may possibly be due to failure of this influence to compel groups of ectodermal cells towards neural differentiation.

Dermoids vary in size from that of a seed to that of an apple. They are encapsulated and oval, round, or hourglass in shape. They are soft and usually cystic, the actual consistency of the contents being determined by the presence of sebaceous material, fat, and cholesterol crystals in varying proportions. The presence of hair has been regarded as pathognomonic of a dermoid but intermediate forms have been found. Microscopically the wall of a dermoid resembles skin and one can distinguish from without inwards a fibrous capsule, a connective tissue layer containing blood vessels (especially at the site of the dermoid tuft which represents the original site of the dermal implantation), and an inner epithelial layer. This inner layer varies in thickness but resembles skin, there being a number of layers of cells, the innermost being flattened and without nuclei. The wall nearly always shows some evidence of inflammatory irritation.

Epidermoids are simple in nature, and form a more solid mass which may be described as being made up of two layers: the outer layer, or stratum durum as it is called, is formed of layers of collagen fibres with little or no cellular formation; it is this outer layer which gives the epidermoid its characteristic appearance. The inner layer forms the interior with a depth of from two to ten cells; fine granules of keratohyalin are scattered throughout the cytoplasm of these cells and the interior of the cyst is composed of accumulated products of the desquamated epithelial layer. These tumours are of course quite different from the cholesteatomata that occur in the middle ear; the latter are associated with inflammation and consist mainly of epithelial debris, granulation tissue, leucocytes, and cholesterol crystals which are the accumulated products of excess desquamation and reaction to inflammation.

OSTEOMATA.—Tumours arising in the walls of the orbit may be either benign or malignant, primary or secondary, or due to one of a group of miscellaneous diseases of doubtful aetiology.

Osteomata are described as either cancellous or ivory but the latter term is the better. Ivory osteomata occur in almost any part of the skull including

the orbit. They are often dome-shaped and are formed of very hard, mature bone. They are found in both sexes in later life, and are slow-growing and painless. They sometimes follow trauma and after some years of growth tend to come to a standstill. On the vault of the skull they are usually symptomless but when they occur in the orbit proptosis may follow or in advanced cases blindness may result from pressure on the optic nerve. Their usual form is sessile like a limpet and covered with periosteum, the surface being hard and frequently polished. Occasionally, small compact pedunculated tumours are found, which have a short stout pedicle and a mushroom-like head. They are composed throughout of mature, compact bone continuous with the outer table of the adjacent bone and formed of concentrically-arranged laminae containing small haversian systems.

A special type of these osteomata is that which occurs in either the frontal or the ethmoid sinus or in the antrum of Highmore; many remain small and undetected, but others grow steadily larger and sooner or later produce subjective or objective symptoms. They enlarge in a centrifugal manner destroying the bony walls of the sinus by pressure atrophy. In this position symptoms arise of inflammation or obstruction of the sinus. Later they spread into the cranial cavity, nose, and orbit, causing pressure symptoms, proptosis, and sometimes the formation of mucocoeles or aerocoeles.

The precise aetiology of these tumours is not known. Associated infection is more likely to follow than to cause them. A history of trauma is common but its association is difficult to prove. The ethmoid type is formed in cartilage but those on the frontal bone in membrane. In this latter region however the frontal sinus continues to grow until after puberty and it may be that islands of cartilage are misplaced and stimulated to grow, possibly by trauma. The treatment of these osteomata often involves a complicated neurological operation; in the old days it terminated only too frequently in meningitis and cerebrospinal rhinorrhoea.

The so-called cancellous type of osteoma is usually due to an underlying meningioma and is better known as a hyperostosis (Fig. 8). Amongst the commoner sites are the olfactory groove and the lesser wing of the sphenoid. A meningioma developing in one of these positions, often of the *en plaque* type, invades the underlying bone and causes new bone formation. Hyperostosis of this type is often the cause of exophthalmos, and such a diagnosis may be confirmed by an appearance of fullness in the temporal

FIG. 8.—Hyperostosis of frontal bone due to underlying meningioma.

fossa, or of thickening of the sphenoidal wings, or it may be seen by x-ray or may cause an increase of cerebrospinal fluid protein.

Of malignant tumours of the bone around the orbit, osteogenic sarcoma and fibrosarcoma are rare but secondary tumours are more common. A particular example of this is neuroblastoma—the so-called Hutchison's syndrome. Secondary deposits are also rarely found as the result of some haemopoietic disease, such as lymphatic leukaemia in children and myelogenous leukaemia in adults.

FIG. 9.—Skull of case of leontiasis ossea (Royal College of Surgeons).

omitted a few rare tumours which should have been included, but I have confined my remarks to clinical material that has come under my care during the past thirty years.

In Schüller-Christian disease, alternatively known as xanthomatosis, deposits of lipoid material occur in the membrane, bones, lymphatic glands, and viscera. X rays display the typical punched-out appearance of the bone.

Paget's disease attacks elderly people and occasionally picks out one bone such as the upper jaw. A similar deformity may arise from leontiasis ossea (Fig. 9), a condition of bone overgrowth of uncertain origin, which produces asymmetrical deformity with proptosis of one or both eyes and in late cases optic atrophy.

This survey of peri-orbital tumours, although comprehensive, may have