THE BRITISH JOURNAL
OF
OPHTHALMOLOGY
JUNE, 1931

COMMUNICATIONS

TUMOURS OF THE LACRYMAL GLAND

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TUMOURS of the lacrymal gland are relatively rare. Less than 300 have been reported, the earliest record being in 1598 by Fabricius Hildanus in *Observationes Chirurgicae*.

Warthin, in 1901, collected 132 cases and drew attention to the similarity of lacrymal and salivary gland tumours. In 1904 Verhoeff published five cases which had come under his care. Greeves, in 1914, collected 42 cases occurring in the previous 15 years and attempted to make a classification based on the pathological reports as described by the various writers, illustrating various types by description of four tumours which he had examined. Shortly after this publication Lagrange and Birch-Hirschfeld added to the literature in this subject. In 1922, L. L. Lane collected and analysed all the published cases of lacrymal gland tumours. After an exhaustive search she eliminated all duplicates and doubtful cases and was able to find in the literature 112 authentic cases published since Warthin's paper, together with 15 reports overlooked by the latter observer.

I have been able to find a further 17 cases reported in the last eight years, together with the following case which came under my care.

Apart from the pathological nature of the growth this case is of interest as it shows the possibility of recovery of useful vision even

when the optic nerve has been subjected to very considerable stretching over a long period.

Mrs. T., aged 37 years, first came under my care in May, 1926, with a large degree of proptosis of the right globe. She stated that two years previously she had noticed a small lump under the outer part of the right eye-brow, which was freely movable and

PLATE 1.

which could be pressed back "into the eye socket." There was no history of any accident and the family history was good. The lump gradually increased in size without pain and for six months no other symptoms were noticed. About this time diplopia began to develop and the eye was noticed to "stand out." Vision gradually became worse with the increase of proptosis and when she was first seen had decreased to perception of light only. The condition progressed; when she came for advice the exposure of the cornea was causing considerable pain, which she controlled by continuous moist applications, while the increasing size of the growth and the consequent pressure from behind gave her the impression that the eye was about to burst.

When I first saw her (Plates 1 and 2) the right eye appeared to be dislocated out of the orbit. The upper lid was so stretched over the protruding growth that it did not cover the whole of the cornea. The globe was actually pushed over the inferior margin of the orbit medialwards: the medial two-thirds of the lid being tucked up behind it and the inner portion of the bulbar conjunctiva

PLATE 2.

was in contact with the skin of the side of the nose. Reposition by pressure on the eye through the lid was impossible owing to a definite resistance within the orbit.

The eye itself showed marked venous congestion and the exposed lower part of the cornea was hazy and stained deeply with fluorescein. It was covered with an exudate of lymph and the exposed conjunctiva was chemotic.

Viewed from in front the visual axis was directed towards the bridge of the nose, the pupil being displaced 19 mm. downwards and 8 mm. inwards.

The upper lid moved freely over the growth, which came forward out of the orbit from the region of the lacrimal gland. On

attempted movements of the eye the growth moved within small limits over the bony wall of the orbit, while to palpation a smooth fleshy mass was felt relatively fixed. The eye movements were restricted almost completely.

A clear view of the fundus could not be obtained, but the retinal veins seemed to be engorged and the arteries somewhat smaller

PLATE 3.

than normal. There were no glandular enlargements and an X-ray was negative. The vision of the right eye was perception of light only, while that of the left was 6/9 part.

After admission I decided to remove a portion of the growth for pathological examination and under novocaine an incision was made below the brow and over the swelling. An encapsuled tumour was found entirely separate from the skin and bone. Owing to the fact that the eye was practically dislocated out of the orbit it was found possible to follow the tumour backwards without difficulty. It extended some 35 mm. towards the apex of the orbit and could be moved easily in a socket of fat, giving the impression that its removal would be simple. Although at that time no

immediate removal had been entertained, the growth was removed without difficulty: the only attachment being to the conjunctiva of the upper fornix.

The wound was closed and a small drainage tube inserted in the outer angle. This was removed in 24 hours and the wound healed by first intention.

June 11. The eye was then retracted in the orbit, the movements being somewhat restricted in all directions. The upper lid could be raised some 50 per cent. of the normal (Plate 3).

July 2. The eye movements were almost full. The edge of the upper lid was at the level of the pupil and the cornea was almost clear. There was no diplopia and the vision was 6/60, with correction 6/36.

Pathological Examination

The tumour is encapsuled and is roughly spherical, being 35 mm. in its largest diameter. It is solid and cuts like a fibroma.

Under low-power magnification the capsule can be seen to be formed of a delicate fibrous tissue membrane the fibrillae of which run into the tumour tissue itself.

The growth is vascular, the size of the vessels varying in all degrees. Some of these vessels have well-marked walls while in other places the wall is either very thin or absent altogether.

In one place beneath the capsule, but separated from it by tumour cells, are the remains of the lacrymal gland. Parts are normal in appearance but as the remains are followed deeper into the growth atrophic changes are noticed and the gland tubules are surrounded by hyaline tissue. Further towards the centre there are islands of this hyaline tissue where the remains of the gland tubules have entirely disappeared.

The tumour proper consists of a dense stroma of cells, there are no areas of degeneration and it is solid throughout. The cells are elongated and spindle-shaped, being supported by a definite stroma; in places where the cells are not so closely packed this stroma is markedly in evidence and has the appearance of a fibroma. This is a spindle-celled (fibro-) sarcoma.

In all cases of malignant growth the question arises as to whether a more radical operation than the simple excision of the tumour should be performed. In this case I had to consider whether the eye should be excised and the orbit exenterated. As there was a long history with an encapsuled growth I felt that no harm could come from watching the case, especially as the vision had improved

after the operation. Time has now justified this decision and it is now four years since the removal of the growth and there is no evidence of either local recurrence or metastatic formation.

I have recently examined the patient and the present condition is completely satisfactory (Plate 4). The upper lid shows a slight ptosis, but is well clear of the upper limit of the pupil. The patient

PLATE 4.

states that the eye does not run freely with tears during emotion like the left, but otherwise she notices nothing unusual. The eye movements are full in all directions.

The cornea is clear but with focal illumination faint scarring can be seen in the superficial layers, more marked along the upper edge of the area which was previously exposed. The fundus is normal apart from the disc, where there is more connective tissue than on the other side.

The vision of the right eye is 6/36, with correction of - 1.0 D. sph. and - 2.0 D. cyl. axis 105° 6/5 part. The field is full both to white and colours. The vision of the left eye is 6/9 with correction + 1.0 D. sph. and + 0.50 D. cyl. axis 90° 6/5.

In the past considerable confusion has existed in the nomenclature of lacrymal gland tumours. A consideration of the pathological diagnoses made by the original observers shows a diversity of terminology; almost all the existing terms have been used. Warthin's list of cases when classified included 44 varieties of growths. Of the 17 cases published in the last eight years ten were mixed tumours, two, cysts, three, of no very definite pathological diagnosis, one, a fibroma and one, a lymphoma.

Apart from neoplasms lacrymal gland enlargements are caused by cystic dilatations, inflammatory swellings and simple hypertrophy, while secondary enlargements associated with Miculicz's syndrome and the leukaemias are well known.

Tumours, excluding cysts, can be divided into two main classes, *a*. Mixed tumours. *b*. Tumours, the histological features of which suggest sarcoma.

Mixed tumours, endotheliomata, or tumours of mixed mesoblastic tissue as they have been described by different writers form the large majority of growths of the lacrymal gland.

Sections of such tumours show a heterogeneous mixture of tissues in varying combinations: columns of endothelial cells, spindle-shaped cells, connective-tissue elements, myxomatous tissue in varying degrees of degeneration, cartilage, epithelial-like cells with areas of keratinization and prickly cells are found. Greeves classifies them into three types; first, a complex type in which gland-like structures, squamous epithelium with prickly cells and often cartilage are found. Second, the type in which myxomatous stroma is prominent, which contains columns of epithelial-like cells. Third, a cylindroma type resembling adenoma and adeno-carcinoma. Probably many of the cases described as adenoma or adeno-carcinoma belong to this class.

Similar growths are met with in the salivary glands and according to some authorities (v. Ohlen, Kelly and others) have been declared to be endothelial in origin. Warthin supported the endothelial origin of the lacrymal gland growths of this type, at the same time pointing out their dissimilarity to endotheliomata occurring elsewhere in the body. These latter growths are much more malignant and are not composed of such a mixture of tissues. For this reason Collins and Mayou prefer to call the lacrymal growths tumours of mixed mesoblastic tissue.

The endothelial origin has, however, been strongly contested. Wood, in 1904, in his collected cases of salivary gland tumour, stated that 95 per cent. were of the mixed variety and he came to the conclusion that the glandular and prickly cell elements were epithelial in origin. Verhoeff supported this in his study of lacrymal gland tumours and was able to demonstrate to his own satisfaction that these growths always developed from epithelial

cells. He suggested that the origin was partly epiblastic and partly mesoblastic and that the cartilage present was an atypical development of the stroma cells. The later reports of Birch-Hirschfeld supported this view. It has been suggested that the complicated nature of these growths would appear to indicate that they do not develop from normal gland structure, but from cells which have been misplaced in embryonic development. Although the mixed epiblastic and mesoblastic origin of these growths would appear to be generally accepted, some authorities, notably Krumpcher, suggest a purely epithelial origin accounting for the mucoid and cartilaginous tissue by a metaplasia of the epithelial elements.

The stroma of the normal lacrymal gland contains lymphoid tissue (Axenfeld) and it is therefore conceivable that lymphoma or lympho-sarcoma may develop. Lymphomata arising in this position are exceedingly rare; I have been able to trace only one such authentic growth, published by Pfingst and discussed at the American Ophthalmological Society where Verhoeff compared it to a similar case which had come under his care. He (the latter) appeared to have some doubt, however, as to the true lymphoid origin of these growths.

It has been stated that sarcoma of the lacrymal gland does not occur. In later years this diagnosis has rarely appeared, but without an exhaustive search I have been able to find 25 cases published under the heading of sarcoma, lympho-sarcoma or myxo-sarcoma. An analysis of the diagnoses shows that in 12 cases the growths were of small round cells, occurring unilaterally and not associated with swellings of the salivary glands; seven undoubtedly belonged to the mixed tumour class; two were without adequate diagnosis and one was described with minute histological detail as a spindle-cell sarcoma. Considerable doubt has been expressed as to whether these small round-celled growths are true sarcomata. Most of the cases have been called lympho-sarcoma but the clinical findings of the majority do not bear out the diagnosis of malignancy. Their rate of growth is slow and metastases practically unrecorded. Greeves points out that small round-celled sarcomata do occur and cites a case in a child where a rapidly fatal result was recorded. He suggests that inflammatory and secondary enlargements of the gland may account for many of the cases which have been published in this class. Such enlargements due to overgrowth of lymphoid tissue may occur in connection with leukaemia or pseudo-leukaemia, while mumps, syphilis and tubercle have been known to produce enlargement of the gland.

Habermann, in 1918, reported a case of spindle-celled growth. He concluded a detailed histological description by stating that "although in several cases the tumour cells appeared to grow out

of the cells of the vascular epithelium yet in spite of this the growth was a pure sarcoma not a mixed form nor yet a transitional form, for an 'endothelial tube' (which proved that the vessels grew independently of the growth) was always to be seen under closer examination."

It would appear that the great majority of the lacrymal gland tumours are made up of a mixture of tissues of mesodermal and ectodermal origin and therefore should be regarded as mixed tumours. In some cases, however, the stroma is so definitely predominant that the mixed elements may be overlooked and a mistaken diagnosis made. The stroma of the first case in Greeves's series was made up of spindle cells and resembled a spindle-celled sarcoma, but, apart from the presence of prickle cells and epithelial elements, the vascular and mucoid tissues were not present in the proportion as found in the normal gland. This leads to a point of importance in diagnosis, viz., the pathological nature of a tumour cannot be determined by the excess of a specific type of cell only, unless the other tissues are present in the same proportion as in the normal gland. Provided that the parenchymatous elements are present in normal proportion then a tumour, arising from the supporting tissues and having the characteristics of embryonic connective tissue, must be sarcomatous.

Although it is possible that many of the cases of lympho-sarcoma may be inflammatory or secondary enlargements, yet, if lymphoma can develop, it would appear that a primary tumour of this type, which departed from the normal characteristics, would constitute a lympho-sarcoma.

Lacrymal neoplasms occur chiefly in patients past middle-life; when occurring in children they are almost always rapidly fatal. The average history is about two years and pain except in the late stages is rare. The effects on the globe depend on the size of the growth; diplopia is not as constant as would be expected. Defective vision is chiefly due to optic atrophy, hyperaemia of the disc and corneal exposure. The defective vision undoubtedly accounts for the absence of diplopia in many cases. Some authors have stated that lacrymal growths are relatively benign, but a study of the literature cannot support this. Lane recorded 20 per cent. of recurrences and, in 95 cases, 12 per cent. were fatal, seven of these from metastatic formation. Fatal results were recorded in 20 per cent. of the cases in Birch-Hirschfeld's series.

Treatment consists of an early and complete removal of the growth retaining the globe when possible. The case which I have described shows the possibility of recovery of useful vision even when an external examination showed very little hope of recovery.

In the majority of cases the growth can be removed through an incision along the upper edge of the orbit, although, if much

adhesion to surrounding tissues is found, resort may have to be had to the method of Krönlein. Birch-Hirschfeld expressed the opinion that unless the growth is removed complete, the malignancy is increased and, for this reason, the Krönlein method of exposure, offering as it does freer access, is recommended by some authorities.

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A RAPIDLY-GROWING MALIGNANT TUMOUR OF THE LACRYMAL GLAND

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PATIENT, a Chinese, Li Shen-Chuin, a farmer, aged 21 years. Case No. 16216. Seen at Wesleyan Mission Hospital, Paoking, Hunan, China, April 7, 1930.

Clinical History.—"Patient came to hospital on April 2, 1930, for treatment for his eye which was covered by a projecting tumour. This tumour, he states definitely, has only grown during the last 40 days, previous to which he had nothing the matter at all. For the past 20 days it has been getting red and inflamed and for 12 days he has noticed the gland in front of his ear which is painful.

"Present Condition: The patient is a healthy well-nourished farmer boy. Temperature and pulse normal. No sign of any disease beyond his eye trouble. Skin particularly clean and healthy. There is a large swelling of the whole of the upper lid of the right eye, which stands out almost at right angles to the face. The