ORBITAL TUMOURS

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by

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MY PERSONAL INTEREST in the problem of "Tumours of the Orbit"[1] was prompted initially when an ophthalmological colleague referred a 4½-year old female child because of an irreducible unilateral proptosis of three months' duration. The disorder was painless, occular movements were somewhat defective, and ophthalmoscopy revealed gross papillœdema. An X-ray showed a definitely enlarged optic foramen on the appropriate side. The diagnosis, therefore, was that of an Optic Nerve Tumour (probably glioma) extending towards, if not within, the cranium. The precise problem was—could the tumour possibly be removed whilst preserving the eyeball? Even though technically possible, one wondered what effect would result from division of the central artery, when the available collateral circulation must be jeopardised in some measure through requisite operative manipulations and the disturbed neurotrophic connections. In looking into the literature for information on this matter the only important reference was found in Parsons' "Diseases of the Eye"[2] wherein it is stated that "it is possible in some cases of Optic Nerve Tumour to remove the growth while retaining the eye." Thus it appeared that there existed a reasonable chance of relieving the child of her burden with conservation of the eye. At operation the whole optic nerve, from the back of the eye to the optic chiasma, was excised together with the contained tumour. The final outcome was satisfactory although the post-operative period was a little stormy together with corneal ulceration necessitating tarsorrhaphy.

Some months later a similar case appeared, the patient being a young girl of 13 years of age. Treatment was applied along similar lines but an immediate tarsorrhaphy was carried out in view of the previous experience. A satisfactory result ensued without any event of note. In consequence of the upheaval of 1939, the girl was not seen again until 1943, when she appeared with the tarsorrhaphy intact. She was called for follow-up in January, 1951, when, at the age of 25 years, she came to hospital in excellent health. The eye showed a very moderate proptosis of 3 or 4 mm. now reducible, but showing no obvious visible pulsation.

It is no unusual experience of us all to meet two like but uncommon cases within a short space of time. How extraordinary it seems that, up to date, I have not been called upon to treat another case of the type, in spite of an ever expanding series of Orbital Tumours of varied natures.

Orbital Capacity.—The cavity of the orbit is of limited capacity, consequently the formation of an adventitious mass within it requires the

displacement of normal contents. Hence tumours situated primarily within the orbit invariably bring about some degree of displacement of the eyeball, the direction of displacement being determined by the site of the new mass. When the mass develops behind the globe of the eye, the eye is pushed forwards. However, the occurrence of a forward displacement of the eye does not always mean that the orbit is invaded by a new growth of one type or another.

Proptosis in relationship with Cerebral Tumour.—Proptosis, unilateral or bilateral in distribution, accompanies a percentage of intracranial tumours some of which lie far from the precincts of the orbit. In the literature there are recorded cases of proptosis in subdural hæmatoma, tumour of the anterior and middle cranial fossæ, and even tumour of the posterior cranial fossa. Jacques Ley[3] in a paper in the J. Belge de Neurologie, makes the statement that with cerebello-pontine angle tumours, unilateral proptosis is more likely to be found on the contra-lateral side of the head. Different observers record varying incidences of ocular protrusion associated with intracranial tumours. Skydsgaard[4] in 1938 reported this combination in 4 per cent. of cases, whilst Gibbs and Cushing[5] reported an 8 per cent. incidence in a series of 1,545 consecutive cases of intracranial growths. When it is realised that, in the absence of a visible or palpable mass, it is for displacement of the eye that the patients most often solicit surgical aid, we can appreciate the difficulty in certain cases of diagnosis based solely on clinical grounds. Indeed, in my personal experience, most cases of retro-orbital tumour, at one time or other, have been diagnosed as of thyrotoxic or thyrotropic origin.

Were proptosis a sure indication of orbital invasion, well and good, but we are only too well aware that infiltration of the orbital walls can arise out of other than neoplastic disorders. Some generalised forms of bone pathology play their part. Of these more will be said later in the lecture.

Of course, the material implication in the use of the word " tumour " does not restrict us to disorders of purely neoplastic type, but is employed in the wider sense of any new tissue mass.

Relationship to Trauma.—For purposes of completeness it is necessary to comment upon certain effects of injury, as well as structural abnormalities of congenital character, as failure to consider such parts as they may play can confuse the issue.

Thus the clinical diagnosis of " Orbital Tumour " acquires a complexity the analysis of which requires careful assessment of the patient as a whole if accurate conclusions are to be drawn.

INCIDENCE

Doubtless orbital tumours are of infrequent occurrence but their incidence relative to the general incidence of tumours of the body cannot be stated on our present knowledge. My personal experience amounts almost to total exclusion of intra-ocular growths ; perhaps the impression

formed would be changed were these tumours to be included. Godtfredsen[6] of Stockholm recorded that orbital tumours represented about 1 per cent. of admissions into the opthalmological department at the Karolinska Sjukhuset over a period of 15 years.

Age.—No age in the human calendar is free of the possibility of development of a tumour within or encroaching on the orbital cavity. Even obstruction in labour has been recorded as a result of the extensions of an orbital tumour in the child[7]. The author's experience extends over all ages from birth up to and including the eighth decade. The pathological nature of a lesion can be related in some measure to the age of the victim ; e.g., an optic nerve tumour in a child is usually gliomatous, whereas a similarly placed tumour in the adult is meningiomatous.

Sex.—There seems to be a degree of predilection for the female, but this may prove to be pure surmise arising out of a limited series. This has been noted especially in the inflammatory states and in cases of giant cell tumour.

SYMPTOMATOLOGY

The majority of the patients claim attention on account of occular protrusion, visual disturbance or the formation of a mass, discernible either visibly or on palpation. Other reasons for patients seeking surgical help are :—restricted ocular mobility, diplopia, pain, epiphora, swelling of the eyelids, chemosis and conjunctival injection etc. Perhaps more intimate reference could be made to these several symptoms with advantage.

Proptosis.—Proptosis is the most regular feature of encroachment on the orbital space either from within or from abutting structures. As already indicated, however, we must not infer that the presence of proptosis always means that the orbit is occupied by some adventitious mass. Some of the greatest degrees of protrusion are related to intracranial arterio-venous fistulæ.

Not infrequently the deformity fails to attract a patient's attention until some accidental occurrence reveals its existence. Usually friends, and perhaps relatives, are shy of calling attention to the fact. On the other hand, the patient may become aware of a quite limited protrusion, as in one of my cases. This patient was so certain of his eye becoming increasingly prominent that he instituted a " card test "—repeatedly he placed a card in contact with his cheek and his forehead, and showed an increasing difficulty in closing the affected eye. Should the invading mass be totally incompressible then the deviation of the eyeball is irreducible ; but exuberant vascular channels of naevoid or arterio-venous character, when present, can be evacuated of their contents to allow of compression, and, therefore, permit of a reduction of the protrusion. The occurrence of a rapid increase of protrusion on the adoption of a dependent posture suggests the presence of such vascular channels. In lesser measure, haemangiomatous tumours sometimes show a variable

amount of ocular prominence on changes of posture, crying or other emotional reactions. (Lindsay Rea[8].)

Varying degree of Proptosis.—A variable degree of proptosis is not an uncommon feature in quite a number of conditions, hence more than one explanation for this becomes necessary. Vascular engorgement of distensile nature, hyperaemia and oedema of inflammatory origin, vascular changes of endocrinal source, retention of lachrymal secretions, or free communications between the orbital lesion and the cerebro-spinal fluid spaces (meningocele or encephalocele) are some of the possible factors. So prominent may an eye become that the eyelids retract behind the globe. So much oedema may occur as a result that spontaneous replacement of the lids by the patient becomes impossible—a state comparable with paraphimosis being assumed.

Occasionally the protrusion of the eye is said to be almost precipitate in its development, or a marked sudden increase occurs. Such rapid increases result probably in the former as an outcome of haemorrhage into the tumour, and in the latter from thrombosis of larger venous channels. Should this thrombosis extend, then the outlook becomes serious.

Of all the causes of proptosis, unilateral or bilateral in distribution, the primary lesions are much more often of extra-orbital origin.

Nevertheless, it is not essential that proptosis should accompany every invasion of the orbit by tumour. A glioma of the optic chiasma can extend through the optic canal into the orbit and fail to push the eye forwards, as the amount of bony erosion probably permits of adequate natural decompression.

One may wonder what is the minimal degree of protrusion before its very presence can call for surgical relief. Realizing, of course, that proptosis may occur without orbital invasion, and that orbital invasion can occur without proptosis, it is obvious that in establishing a diagnosis much depends on other symptoms. Rundle and Wilson[9] concluded that 6·0 mm. may be taken as the approximate upper limit of asymmetry in Graves' Disease. In a series of cases of orbital tumours which they investigated, 80 per cent. of the tumours had produced proptosis of amounts greater than 6·0 mm.

Deviation of the Eye.—Deviation of the eye in directions other than forwards is present in the more anteriorly placed tumours, and such displacements are accompanied by a visible or palpable mass.

The situation of such a mass may allow of certain deductions as to likely pathologies owing to the deployment of the various anatomical structures from which tumours are known to arise. But the presence of a visible or palpable mass does not infer one restricted to the orbital cavity, as the following paragraph explains.

Visible and Palpable Tumours.—Many patients appearing because of the intervention of proptosis present a boss in the frontal or temporal region. One encounters these cases giving a history of the presence of

a boss perhaps for several years. Indeed it is no unusual feature for the primary swelling to have been submitted to exploration and biopsy.

My personal experience has been that a generally accepted label of " sarcoma " has been applied, and the patient has had one or more courses of treatment by deep X-ray. More often than not the sequence of events finally shows the disease to be " meningiomatous." Some of my cases of the grossest degrees of protrusion are of this nature.

Visual Disturbances.—Almost as many patients seek aid on account of visual defects as appear through protrusion of the eye. Even then, it is most striking that defective vision in an eye can reach the stage of blindness only to be revealed by some accidental observation. Believe it or not, the author has seen a patient who was unaware of any disability until he found his eye to be blind on closing the other eye in attempting to sight a rifle.

Lesser forms of visual defect may cause but minor measures of inconvenience. Visual disturbance may occur in different forms :—

(1) A careful and observant ophthalmologist finds in comparing serial refractions, an increasing hypermetropia brought about by distortion of the eyeball.

(2) Displacement of the globe of the eye or restriction of mobility due to muscle infiltration induces diplopia. However, diplopia is by no means as frequent as one would suppose. According to Parsons[2] diplopia is one of the commonest complaints, in fact he states that it occurs in a large proportion of cases owing to interference with mobility of the eyes. Neither the series of Handousa[10] nor my own agrees with Parsons. Handousa found double vision in two of twenty consecutive patients, and about 25 per cent. of my own patients had noted diplopia at one time or another during the course of the disease.

(3) Defects of the visual fields are rarely factors of complaint; indeed they are quite unusual findings on clinical examination. In intra-orbital growths few alterations in the visual fields are to be seen; scotomatous defects occur occasionally; alternatively some peripheral constriction may be encountered. When tumours extend into the cranium the optic tract may be subject to pressure; under these circumstances a homonymous hemianopia may appear. The import in such a disclosure is, of course, that it is proof of the extension of the tumour intracranially.

(4) Diminution of visual acuity is a very common occurrence that proves an important factor for the escape of the patient from the inconvenience of double vision, and may be partially responsible for unobserved proptosis. One may well ask to what extent encroachment on the optic canal must take place without damage to the optic nerve. In a paper by Clegg[11] there is stated that a canal measuring less than 2·6 mm. in one of its diameters cannot contain a normal optic nerve. He states also that skull deformity with optic nerve symptoms is fully twice as frequent in eye practice as are optic nerve tumours.

Pain.—The lack of pain, in the mind of an average patient, affords a sense of well-being; consequently it is usually difficult for the patient to acknowledge the existence of a growth. Pain is not a feature of orbital tumours of innocent pathology. The reason behind this is possibly the particularly slow rate of growth of the majority of these neoplasms. As a prominent symptom pain is indicative of malignant disease although some of the chronic inflammatory states are associated with considerable discomfort. In association with optic nerve tumours pain, in my very limited experience, is not a factor of note. Humphrey Neame[12] says that pain is a more likely feature of a meningioma than a glioma. Usually discomfort or pain from an orbital growth is referred to the precincts of the orbit or possibly the back of the eye. Should pain in a case of proptosis be located to the distribution of the trigeminal nerve, especially the second division of that nerve, it is important to exclude malignant disease of the maxilla. The only innocent new growth associated with pain, in my experience, is the neurofibroma. In two of my patients with neurofibromatous tumours pain was quite a prominent complaint, yet other cases of like pathology were painless.

Diminution of Ocular Mobility.—Most innocent new growths cause little interference with ocular movements until late in the clinical period, even with growths of the optic nerve. Infiltration is the mechanism of early restriction. Hence the liability of limitation of movement in inflammatory states and malignant tumours. The infiltration affects the muscles in particular, but the nerves appear to escape, although they may traverse masses of inflammatory tissue.

Epiphora.—Excessive lachrymation is a feature of many cases. Although the fluid production is undoubtedly increased, it is possible that abnormal apposition of the lids with the eyeball interferes with the proper drainage of tears. No special significance is attached to the symptom.

Other Symptoms.—Yet other complaints are made by patients but those symptoms already described represent the picture of so large a proportion of cases as to make other symptoms of secondary importance. Oedema of the eyelids, ptosis, conjunctival injection, chemosis and irritation are some of the more frequent ones.

PATHOLOGY

Not one of the several recorded series of orbital tumours affords any relative incidence of available pathologies[13], [14], [15]. The differences in the series are partly accounted for by the practice of the individual observers, for we are studying the activities of ophthalmologists, rhinologists, neurosurgeons and others with varied surgical interests. On the other hand, the material is of so limited an incidence as to preclude a true cross-section of available pathologies in the hands of any one observer. To read some reports gives one the impression that malignant disease is extremely common, although my own experience would not

warrant the suggestion. Handousa[15] has seen so relatively large a number of malignant growths of the ethmoid as to favour a high incidence. Some surgeons have suggested that metastatic malignant disease is frequent, yet my series affords but three cases, the primaries of which were uncertain in two cases, probably lung and suprarenal, whilst the third is thyroid in origin.

Undoubtedly lack of verification of pathology may give cause for suspicion. Perhaps this is exemplified in Dandy's cases of Schüller-Christian Disease.[13] My one experience of a case so labelled proved interesting; the patient showed a temporal swelling associated with proptosis, oedema of the eyelids, and slight chemosis X-ray revealed a defect of the lateral orbital wall with a circular margin. A biopsy of the temporal mass had been carried out elsewhere from which the pathological report read as follows :—

> " I would suggest that this is some sort of granuloma. Whether it is associated with a generalised metabolic disorder such as Schüller-Christian's Disease one cannot say, although the history might guide one."

Unfortunately, the biopsy was followed by infection which led to a persistent sinus through the upper eyelid ; it was in this condition that the patient came under my care. The chronic infection precluded operative intervention by the regular route. Exploration of the swelling through a vertical incision in the temporal region encountered a cystic mass containing inspissated squamous material. A histological section of a portion of the wall is typical of dermoid tissue.

From personal experiences, the most frequent lesion to be found in the orbit is the chronic granuloma of infective nature. Tubercle and syphilis account for two cases, one is sarcoid in type, one is described as an eosinophil granuloma, and all the remainder are simple granulomatous lesions in various stages of organization.

The syphilitic lesion was of gummatous type. It resolved extremely rapidly on anti-syphilitic medication; in fact, the reaction had more or less completely resolved by the time serological confirmation came to hand. As far as the eosinophilic granuloma is concerned, the actual process involved remains somewhat doubtful. One pathologist suggests that it is an inflammatory mass in which eosinophil cells exist in rather larger proportions than is usual, whereas another opinion is that the lesion is typical of the form of lipoidosis which precedes the Schüller-Christian Syndrome. The patient was a young boy of nine years of age. A count of his blood showed an eosinophilia of 14 per cent.

Obviously the granulomatous masses already mentioned are uncommon, but the simple granuloma sometimes referred to as the orbital pseudo-tumour[16] is, in my experience, the mass most commonly found in the orbit. Its true origin is uncertain. Some investigators associate the condition with exophthalmic ophthalmoplegia, looking upon both pseudo-tumour and the other disorder as two manifestations of chronic myositis. Dunnington and Berke[17] define chronic orbital myositis as a

non-specific chronic inflammatory disease of the extra-ocular muscles of unknown causation. It is characterized histologically by lymphocytic infiltration, fibrosis and degeneration of muscle fibres. The muscles are grossly enlarged, pale and cartilaginous, and they cut with a gritty feel. However, in the pseudo-tumours the cellular infiltration and oedema are by no means limited to the muscles. The histological appearances vary from case to case, according to the degree of organisation present: some consist of cellular collections of lymphoid and plasma cells, others are very fibrous. Though without doubt of inflammatory origin, one wonders what is the source of infection. The masses appear mainly in the upper and inner part of the orbit, or are related to the floor-sites suggesting the likelihood that the ethmoid sinuses or the maxillary antrum may be likely sources. The author's opinion rather supports the suggestion of Offret[18] that the granulomata are products of chronic endophlebitis. At the present time there is a patient in whom a mass attached to the optic nerve led to excision of the eye and the mass, in the belief that its origin must be of malignant nature. The tissue proved to consist largely of fibrous tissue. This unfortunate man now has a similar lesion of the opposite orbit; consequently his remaining vision is at stake. As there is a shadow on the radiograph of his chest the true pathology is not ascertained, and the relationship between the various lesions still remains unknown.

In order to deal with the neoplasms perhaps some simple classification is helpful. Owing to the innumerable pathological entities reported it is easier to consider the problems in relation to tissue planes. In a series of 125 cases of unilateral proptosis or deviation of the eye, of which about fifteen remain undifferentiated (some awaiting operation), the following pathologies have been encountered:—

I. Tumours of the Optic Nerve and its Sheath:

A. Innocent
- i. Glioma—(a) Astrocytoma.
  (b) Oligodendrocytoma.
- ii. Meningioma.
- iii. Fibroma.

B. Malignant
- i. Retinoblastoma.
- ii. Neurocytoma.

II. Tumours of Orbital Tissues other than the Eye and Optic Nerve:

A. Innocent

Solid arising from:

i. Nerve	— Neurofibroma.
ii. Vessels	— Haemangioma.
iii. Lymphatics and Lymphoid Tissue	— Lymphangioma and Lymphoma.
iv. Gland	— Mixed Tumour of Lachrymal Gland.
v. Fat	— Lipoma.
vi. Muscle	— Rhabdomyoma.
vii. Other	— Xanthoma.

A. Innocent (cont.)
- Cystic:
  - i. Dermoid
  - ii. Retention Cyst of Lachrymal Gland.—Hydatid.
  - iii. Parasitic
  - iv. Simple.
  - v. Haemorrhagic.

B. Malignant
- Primary
  - i. Carcinoma.
  - ii. Sarcoma.
- Secondary
  - iii. Melanoma.

III. Tumours of the Orbital Wall:

A. Innocent
- Solid
  - i. Meningioma.
  - ii. Osteoma—(*a*) Cancellous. (*b*) Ivory.
  - iii. Giant-celled Tumour.
- Cystic
  - i. Osteomatous.
  - ii. Mucocele.
  - iii. Dermoid.

B. Malignant
- Primary
  - i. Carcinoma of Ethmoid.
  - ii. Rodent Ulcer.
- Secondary—
  - (*a*) Carcinoma
    - i. Sympatheticoblastoma.
    - ii. Bronchial.
    - iii. Thyroid.
    - iv. Breast.
  - (*b*) Sarcoma
    - i. Mediastinum.
    - ii. Testis.
    - iii. Suprarenal.

IV. Invasion of Orbit via Anatomical Pathways:

i. Glioma of Optic Chiasma.
ii. Glioma of Temporal Lobe.
iii Meningioma.
iv. Neurofibroma of Gasserian Ganglion.
v. Chromophobe Adenoma of Pituitary.

V. Chronic Inflammatory States:

i. Tubercle.
ii. Syphilis.
iii. Chronic Simple Granuloma—Orbital Pseudo-Tumour.
iv. Eosinophil Granuloma.
v. Sarcoid.

VI. Injury, Effects of, including Arterio-Venous Fistula.

VII. Congenital Structural Defects of the Orbit.

VIII. Exophthalmic Ophthalmoplegia.

I. Tumours of the Optic Nerve and its Sheath:

These new growths are rare: such details of them as are available depend greatly on the work of Hudson[19] and Mathewson[20]. Mathewson found that gliomata are about six times as frequent as meningiomata. He states that the portion of the nerve just behind the eyeball is not affected by glioma, whilst the reverse is the case with meningioma. Hudson noted that gliomata occur in children, the average age being thirteen years, while the meningioma is a disease of the adult, the average

age being thirty-five years. Indeed, Hudson commented that 75 per cent. of gliomas of the optic nerve developed within the first decade of life. Humphrey Neame[12] states that circulatory disturbances in the lids and conjunctiva are more marked in meningioma, also that pain is more frequent.

The retinoblastoma is the one tumour of gliomatous nature to metastasize generally within the body. It is a disease of the young, and, as already stated, it may be present at birth. Secondary deposits are to be seen in the liver and kidney on occasion.

II. Tumours of Orbital Tissues other than the Eye and Optic Nerve.

Of these the clinical diagnosis may be helped at times by the situation of the lesion. Neoplasms of the lachrymal gland are relatively frequent. Moreover, most of the tumours developing within the gland are of the "mixed salivary type." These tumours are to be looked upon with suspicion, as they often prove liable to recurrence. But other tumours, in particular Dermoid cysts, appear in the upper and outer quadrant of the orbit.

Haemangiomata are of very slow growth and arise generally behind the eyeball.

Hydatid cysts we do not see in this country. The writer was privileged to see one of Professor Handousa's cases in Cairo. His three patients were aged twelve, ten and nine years, two being girls and one a boy. There appeared to be little that was typical from the clinical aspect but sensitivity tests and blood counts are helpful.

A malignant tumour of primary origin other than of the eye may be carcinoma, sarcoma or melanoma. The carcinomatous disease occurs in the lachrymal gland or an accessory nasal sinus; melanoma is found within the muscle cone, even at times unrelated to the eyeball; and sarcoma in varied forms in the connective or lymphoid tissues. The sarcomatous lesions are usually far the most malignant conditions, and although they sometimes present as defined encapsulated masses, their clinical progress is typical to a degree. Metastatic sarcomatous masses have been described as originating in the mediastinum, testis and suprarenal.

Carcinoma developing within the orbital structures is most likely to occur in association with the lachrymal gland and it is possible that this is the explanation for the rather depressing prognosis attached to new growths of the gland. No doubt an occasional tumour of mixed salivary type undergoes changes involving increased activity, with a corresponding risk of recurrence. The writer's experience of these tumours has been that their gloomy prospect has not been fulfilled unless the existence of evidence of abnormal cellular activity could be recognised in the microscopical sections.

III. Mural Neoplasms and Neoplasms developing outside the Muscle Cone:

Innocent—Osteoma is to be seen in either the "cancellous" or the "ivory

type." Usually the cancellous growth is associated with the cranial vault, where it appears as more of a hypertrophic state of the bone with an ill-defined margin. The ivory osteoma arises in association with one of the accessory nasal sinuses, most commonly probably in the frontal air sinus. Handousa, no doubt as the result of his interest in nasal pathology, has met a considerable number of these tumours.

Dermoid Cysts are one of the more commonly occurring tumours involving the orbit. Usually they form in the upper and outer quadrant of the orbit, sometimes excavating or penetrating the bone; at other times restricted to the orbit. They can be most extensive, and then involve the middle cranial fossa as well as the orbit. One patient, a boy of sixteen years of age, with a marked prominence of the temporal region associated with proptosis, complained also of a discharge of soft flaky material from the anterior nares. He had developed a fistula into the nose through which typical epidermoid tissue was escaping. The clinical history of this boy covered a period of four years.

Reports on individual series give indication of the basic interests of the observer, but even then it is peculiar how common a lesion may appear in one series although lacking in others. Perhaps the most striking instance of this is Handousa's series of *Osteoclastomas of the Ethmoid* (1915). Professor Handousa has collected twelve of these tumours at this site. This is a striking collection, one case of which the author has been privileged to see.

In looking into the literature it is interesting to note the comment of Lord and Stewart[21] that " the skull as a whole, apart from the jaws, is seldom affected by this tumour." Ambrose Paré[22] reported several cases in the maxilla in 1860. Wattles[22] in a paper on " Benign Giant Cell Tumour of the Ethmoid Labyrinth " draws attention to the higher incidence of these tumours in the female, and remarks upon the relationship of the tumour to trauma.

Another growth of interest, found in the wall of the orbit, is of cystic nature. Its true pathology is not determined ; consequently it is referred to as *Osteomatous Cyst*. Of course, other forms of bone cyst occur, as in association with fibro-cystic disease of bone.

Haemangioma of bone is to be found involving the orbit. It appears in two different forms, the cavernous and the capillary. Characteristic radiographic appearances clinch the pathological diagnosis in such conditions. Further reference to this is to be found in the radiological section of the paper.

The most common growth in bone found as the cause of proptosis is the *Meningioma*. Arising in the dura mater of the anterior or middle fossa of the skull, particularly in the form of meningioma en plaque, it pervades the skull, even invading the temporal muscle. Some of the most extreme cases of proptosis arise out of these tumours.

Malignant growths, as elsewhere, may be primary or secondary. The commonest malignant growth to be seen is *Carcinoma of the Ethmoid*

Sinus. Indeed these growths are relatively so frequent as to lead to the impression that malignant disease of the orbit is more common than other neoplasms. Similar types of carcinomatous disease giving rise to proptosis are to be seen; they arise from the nasopharynx or the maxilla. Rarely is the orbit invaded by epitheliomatous growths of surface origin. This the author has seen from carcinomatous changes in skin treated by deep therapy, and from a rodent ulcer.

Metastatic Malignant Disease may be found in the bone or soft tissues but the clinical diagnosis, though suspect, is never certain, unless characteristic changes are to be envisaged on X-ray. These lesions have played only a small part in the author's pathological material. The only personal case originated in a sympatheticoblastoma, and I have operated on one intra-orbital secondary of unknown origin. At the present time I have a case awaiting treatment in whom there exists an undoubted secondary of thyroid origin. The suprarenal, the breast, and other sources have been reported.

IV. Invasion of the Orbit via Anatomical Pathways

Some intracranial tumours actually traverse anatomical channels to invade the orbit, but this manner of spread is uncommon. Glioma of the optic chiasma shows a tendency to extend via the optic nerve and is likely to expand the optic foramen. Extension of the tumour into the orbit is not of necessity accompanied by proptosis. The author has seen a case in which a temporal astrocytoma extended into the orbit via the optic canal. In one patient a neurofibromatous growth originating in the gasserian ganglion has invaded the orbit through the foramen rotundum.

In the author's collection there are two patients in whom a chromophobe adenoma of the pituitary has invaded the orbit.

No doubt other forms of new growth must involve the orbit from time to time.

V. Chronic Inflammatory States

No further reference is required at this juncture as adequate description has already been applied to these disorders.

ANCILLARY METHODS OF INVESTIGATION

A. Ophthalmoscopic Examination

Appearances of the ocular fundus vary in accordance with the site of the tumour and with its nature. With tumours of the optic nerve, papilloedema or consecutive atrophy is the regular appearance, but direct invasion of the disc by growth may prove deceptive. Glioma of the nerve is said not to encroach on the eyeball—if this be true then infiltration of the optic disc by tumour would indicate the presence of a meningioma of the nerve or, of course, a retinoblastoma.

Malignant growths give rise to papilloedema of variable height, no

matter what their plane of origin. Innocent tumours within or outside the muscle cone do not affect the optic disc until later in their course when a primary optic atrophy is seen. The inflammatory states vary —they more often give rise to minor degrees of papilloedema, but can induce a primary atrophy. With disease arising outside the orbit, the changes to be envisaged in the optic disc are those accompanying the basic state, though primary optic atrophy is the most likely change.

A flaring type of papilloedema with haemorrhages is seen very occasionally in cases of exophthalmic ophthalmoplegia, but disc changes do not occur regularly in this disease.

B. Radiological Examination

Those tumours restricted to the orbit ordinarily show little change on X-ray, unless they are of very long standing, when the whole orbit may be enlarged in comparison with the other side. According to Handousa this is a frequent finding. My own experience does not concur in the matter of frequency. Of course soft tissue shadows may be seen, or a density of the affected side be increased owing to increased vascularity or oedema; particularly is this the case in orbital pseudo-tumour. Generally speaking, unless the disease implicates the orbital walls, or is of orbito-cranial distribution, radiographic evidence is unlikely. Mucoceles show pathognomonic changes. as the outline of the responsible sinus usually shows enlargement whilst being deficient simultaneously where the mass extends into the orbit. Conditions producing changes in the bony structure may be recognised where typical changes occur, as in the meningioma, osteoma and sclerosing conditions and haemangioma. The radiating trabeculations typifying the cancellous form of haemangioma of bone cannot fail to be recognised. Capillary metastatic deposits are commonly multiple; hence other lesions may be found in the skull or other parts of the skeleton.

Cystic cavitation of bone produces characteristic outlines.

When a mass extends from the orbit into the cranium, the channel of continuity will produce an associated outline. Should the communication be via one of the anatomical channels, i.e., the optic canal or the sphenoidal fissure, then that channel becomes enlarged in comparison with the corresponding structure on the other side. The question of symmetry in normal channels must be considered in order that one may not be misled. According to Clegg[11] symmetry exists in but 45 per cent. of skulls. Nevertheless, the variations are such as not easily to be mistaken. On the radiograph, a normal optic foramen is 5 to 6 mm. in diameter. In the presence of a tumour the canal is distended in the two diameters.

Stereoscopic radiographs are useful at times but they are required rarely in the elucidation of clinical problems.

Arteriography.—There is little scope for arteriography in the diagnosis and treatment of orbital growths. The recognition or verification of

arterio-venous fistulae or aneurysms would cover the application of the method in regular practice.

C. Laboratory Investigations

First of all every patient's blood is examined for the W.R. or Kahn test or both.

Skin sensitivity tests are applied in cases wherein hydatid disease is suspected.

Blood counts are applied on clinical grounds for information referable to anaemia, leukaemia and eosinophilia. Coagulation tests and platelet counts are applied should any question of abnormality be suspected. Occasionally in active infective states and malignancies, the blood sedimentation rate is of value.

Lumbar Puncture and Analysis of the Cerebro-Spinal Fluid

In orbito-cranial lesions the information acquired from lumbar puncture may be helpful. The question of how far proximally a gliomatous tumour extends along the nerve may be indicated in some measure from the cerebro-spinal fluid ; if the total protein content is normal, it is a reasonable assumption that the subarachnoid space has not been reached, or is involved to a minimal extent. For purely orbital disease little information would be forthcoming as a result of lumbar puncture. It should be kept in mind that in a case of orbito-cranial tumour with raised intra-cranial pressure, lumbar puncture may cause alarming reactions, so that this procedure should not be applied indiscriminately.

DIFFERENTIAL DIAGNOSIS

The presence of a mass or deviation of the eye from its normal position are sure indications that some adventitious structure exits. Whether it be neoplasm or not must be concluded from the history and such aid as is to be made available by ancillary methods of investigation. A story of injury succeeded by ocular protrusion, disordered mobility, diplopia, visual deterioration, or the occurrence of a bruit may permit certain determinations. Yet a number of tumour cases cite specific injuries following upon which the symptomatology is said to have developed ; this has been the author's personal experience in cases of glioma, osteoclastoma, haemangioma and granuloma. Recently there was under my care a child who received a blow on the head, in whom proptosis was said to follow. Before she came she was looked upon as in a vascular fistulous condition and calcific changes visible on X-ray were interpreted as callus around a fracture ; however, the lesion was found finally to be a haemangioma.

Where unilateral proptosis is the prime complaint it is strange that the majority of purely orbital tumours have been diagnosed as due to dysthyroidism prior to reaching the neurosurgeon. Exophthalmos, unilateral or bilateral, in a patient who has a goitre or who has been submitted to thyroidectomy may be considered as exophthalmic ophthalmloplegia, but a unilateral exophthalmos in a patient without any associated

endocrine disturbance is very much more likely to arise out of an orbital tumour than any other condition. From the clinical appearances alone differentiation between the two conditions may prove extremely difficult. There is of course the basal metabolic rate, which may prove of value. What points of differentiation are to be applied? First of all, the ophthalmoplegic case rarely remains unilateral, and usually associated evidence of thyroid disease exists, and also the clinical course is one of deterioration followed by regression of symptoms; moreover, with tumour, the difference in protrusion between the two eyes is usually more marked. In an investigation of a series of cases by Rundle and Wilson[9] at the Westminster Hospital the minimal difference in tumour cases was found to be 6 mm. Retraction of the upper eyelid is a common feature in ophthalmoplegia but rare with tumour. Most innocent neoplasms in the orbit are of very slow growth. Consequently the case presents a history of progressive change, frequently of many years' duration. Many tumours form in the orbital outlet; consequently they are to be seen or palpated. Under these circumstances, the situation of the mass often indicates the source of origin, and the likely nature may be assumed from this, together with the recorded duration of the lesion. Frequently patients are found to comment on a variability in the amount of protrusion. Should this arise out of posture an excessive vascularity is indicated. Were the origin a carotico-cavernous fistula, dilated venous channels would occur and also an audible bruit on auscultation of the head. With structural abnormalities of the orbit, as in a meningocele or encephalocele, posture brings about a like change, but no bruit is to be found. In both types the proptosis is usually pulsatile. But rapidly increasing proptosis of lesser degree, brought about by bending or accompanying an emotional outburst, occurs in some haemangiomas.

Granulomatous lesions tend to show remissions in symptoms, possibly as the result of varying intensity in the inflammatory reactions. Pain is a factor in diagnosis, for it does not apply to most innocent conditions other than a granulomatous state. As mentioned previously, some neurofibromata give rise to considerable pain. Otherwise one is anxious in the presence of pain less malignant disease be the cause.

Ancillary investigations may be conclusive. The ophthalmoscope is valuable as many appearances in the optic disc and the retina are helpful. Papilloedema is found in optic nerve tumours and granulomata, otherwise one fears its presence as foreboding malignant disease. As already mentioned, it can appear with exophthalmic ophthalmoplegia in severe degrees, but this is unusual.

Typical appearances on X-ray confirm the diagnosis in many cases, certain diseases such as ivory osteoma, cystic states and mucoceles exhibiting pathognomonic appearances. Of the tumours limited to the orbit, no specific appearances are to be visualised, but a relative density of soft-tissue shadows is not uncommon in granulomata and malignant conditions. Distension of the optic canal or the sphenoidal fissure, or

areas of erosion in the orbital walls are positive evidence of the presence of a tumour.

The blood picture does not play any great part in diagnosis although it may be of value in certain cases. Serological tests and sensitivity tests may be helpful in the exclusion of syphilitic disease or parasitic conditions like hydatid disease.

TREATMENT

Appropriate treatment is gauged by the clinical diagnosis, but a positive diagnosis cannot always be reached on clinical data even after the accessory forms of investigation have been applied. Therefore it may be necessary to resort to removal of tissue for biopsy as the initial measure. But this form of investigation should be restricted as far as is possible to the confirmation of malignant disease. Naturally with tumours lying behind the eyeball this cannot be carried out without exploration of the orbit, in which circumstances the best means of exposure must be debated. Some surgeons with experience of granulomatous states condemn biopsy as, in their opinion, a flare in the disease is a very likely outcome. The writer's experiences have led to a decision to employ conservative measures when dealing with malignant disease rather than to resort to drastic surgical intervention.

In so far as pathologically innocent neoplasms are concerned, if the patient's physical state justifies the undertaking and the disability warrants it, then excision is the method of choice. Our next consideration is the question of operative exposure. Wherever possible, the scope of surgery requires the use of the most direct method affording the optimum exposure. Therefore many tumours at the orbital outlet can be exposed satisfactorily through an anterior incision. But with growths extending behind the eyeball the field for manipulation through an anterior approach is so restricted that the feasibility of total removal becomes problematical. Even the lachrymal gland tumour, although easily discernible to the palpating finger, may be as the iceberg in the sea and project but a ninth of its total capacity. This being so, especially with a tumour of such a fearsome reputation, one is doubtful of the advisability of attempting its excision from the front. Biopsy material can be removed through an anterior incision, but it is not advisable for puncture biopsy to be done.

What then is the best means of approach to give the maximum exposure with the minimum of intervention?

We are all conversant with the Krönlein operation and the Naffzigger method. Neither method is ideal. Other operations have been designed by a number of surgeons but they have not been so beneficial as to warrant their adoption in preference to those commonly applied. The disadvantages of the Krönlein are the limited exposure, a tendency to permanent strabismus owing to muscle fixation, and the poor aesthetic appearance of the final scar in many cases, whilst its adaptability in dealing

with more expansive lesions is unduly restricted. The objections to the Naffzigger operation are its expansiveness and the pulsating exophthalmos that results. Although the cited objections may not be tenable as a whole, it becomes apparent to one who is frequently called upon to treat orbital growth that the disadvantages of these two different operations could be eliminated were an exposure possible whereby the lateral scar may be avoided and the extent of the exposure prove adequate. The author has applied the two methods to common advantage by avoiding the turning of an osteoplastic flap yet permitting a more extensive decompression than applies to the Krönlein exposure. A much wider field of action ensues and a greatly increased area for purposes of decompression is the outcome. For lesions such as exophthalmic ophthalmoplegia or pseudo-tumour this wider decompression has great advantages to offer.

Lesions involving the orbit and the cranium require the more extensive field for manipulation afforded by a transfrontal osteoplastic flap. That removal of the orbital roof leaves a pulsating exophthalmos unsightly and disagreeable to the patient bears limited truth.

Undoubtedly when under an anaesthetic an exaggerated example of this exists, but in the erect posture the pulsation is minimal and is hardly noticeable to the untrained eye.

The malignant tumours, whether epiblastic or mesoblastic in nature, demand special consideration. My personal belief is that, with our present knowledge, radical surgery has little to offer. The only malignant growth to be submitted to radical surgery is the retinoblastoma. To excise a mass, an eye together with an attendant mass, or to exenterate the orbit is apt finally to leave the patient with a foul, fungating herniation that proves unresponsive to any measure of treatment. Should one be able to avoid the pitiful course of a patient harbouring a hideous mass emitting an offensive odour surely this is wise. My own view is that the malignancies, on established proof, should be submitted to deep therapy without further surgical intervention. The radiotherapists however, are not agreeable to intensive teletherapy with the eyeball in situ, therefore enucleation is the common practice prior to X-ray therapy.

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MONTHLY DINNERS

Monthly dinners are held in the College on the Wednesday before the second Thursday of each month. The following are entitled to attend with their guests; all Diplomates and students of the College and Members of the Associations linked to the College through the Joint Secretariat. It is not necessarily intended that guests should be members of the medical profession.

The dinners will be at 7 p.m. on the following Wednesdays: October 10, November 7, and December 12, 1951; January 9, February 13, March 12, April 2, May 7, June 11, and July 9, 1952.

The cost is £1 10s. 0d., which includes cocktails before dinner and wine at the table. Applications for tickets, accompanied by a cheque for the appropriate amount, must be sent to the Deputy Secretary at least a week before the date of the dinner. Cheques should be made payable to "Royal College of Surgeons of England." The dress is Lounge Suit.

DIARY FOR AUGUST

There will be no lectures at the College during August, but the Museum and Library will remain open.

DIARY FOR SEPTEMBER

Mon.	3		Basic Sciences Course for Dental Students re-commences.
Tues.	4	4.0	DR. RALPH M. TOVELL—Frederic Hewitt Lecture—New Horizons in Anaesthesiology.*
Tues.	25	5.0	MR. RAINSFORD MOWLEM—The Use of Bone in Reconstructive Surgery.
		6.15	MR. RODNEY MAINGOT—The Surgery of the Bile Ducts.
Wed.	26	5.0	MR. NORMAN TANNER—Non-Malignant Surgical Affections of the Cardiac End of the Stomach.
		6.15	MR. CHARLES READ—The Ureter in Gynaecology.
Thur.	27	5.0	MR. R. FURLONG—The Painful Shoulder.
		6.15	PROF. G. PERKINS—Pott's Fracture.
Fri.	28		Basic Sciences Course for Dental Students ends.
		5.0	MR. W. C. GISSANE—The Principles of Treatment in Burns.
		6.15	MR. J. B. PENNYBACKER—Obstructive Hydrocephalus.

* Not part of course.