CASE NOTES

DICTYOMA*

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THE term medullo-epitheliomata is applied to a group of tumours arising from the medullary epithelium of the primitive neural tube and its derivative, the primitive retinal epithelium. Medullo-epitheliomata of the brain are extremely rare (Biggart, 1949). Bailey and Cushing (1926) mention but two examples in their series, both arising from the region of the third ventricle, where the floor plate and roof of the primitive neural tube persist into adult life.

In the eye, tumours of this group arise usually from that portion of the derived medullary epithelium which borders the more highly differentiated retina, the pars ciliaris retinae. A number of tumours arising from this part of the eye was analysed by Fuchs (1908). He considered them to be medullo-epitheliomata, but divided them into two main groups. One, consisting of four cases, he termed dictyomata; these tumours mimicked embryonic retina. The other group consisted of a variety of tumours which did not go so far as to produce retinal elements, but showed various epithelial formations resembling other portions of the derived medullary epithelium of the optic vesicle. The tumours of the first group, the dictyomata, were collected from four authors who had previously published accounts of their pathology. Badal and Lagrange (1892) had considered their specimen to be a carcinoma; Emanuel (1900) had suggested the term 'glioma pars ciliaris retinae'; Verhoeff (1904) had coined the word 'teratoneuroma' to describe his example; Kuthe and Ginsberg (1905) called their specimen a 'malignant epithelioma'.

Since the publication of Fuchs's classic paper on the subject, further instances of the occurrence of this tumour have been recorded (Velhagen, 1919; Redslob, 1923; Satanowsky, 1928; Soudakoff, 1936; Satanowsky and Cramer, 1938; Schepkalowa, 1938, 1941; Klien, 1939; Czukrász, 1941; Imre, 1941; Rubino, 1941; Andersen, 1948; Fralick and Wilder, 1949; Gasteiger, 1950). The total number of recorded cases is probably less than a score. Andersen (1948) considered that seven of his 22 cases should probably be excluded. Descriptions vary considerably and some cases *e.g.* those of Greeves, 1911) might be more profitably included in Fuchs's second group. Other possible cases (Stanworth, 1951) are unfortunately devoid of pathological details. A general picture of the nature of this tumour can be obtained from a study of the more detailed reports in the literature (Klien, 1939; Imre, 1941).

*Received for publication March 14, 1955.

Dictyomata have been described with the following features:

- (1) Cellular membranes resembling embryonic retina.
- (2) Membranes closely resembling the ciliary epithelium.
- (3) Areas of rosette formation, as in neuro-epithelioma and retinoblastoma.
- (4) Pigmented epithelium.
- (5) Areas of neuroglia.
- (6) Islets of cartilage.

The tumour is rare, and the following example is recorded here for this reason and also because it demonstrates well the varied histological features and reveals a hitherto unreported degree of malignancy.

Case Report

One morning in 1951, the patient, then a child aged 3 years, awoke to find her left eye swollen, bloodshot, and tender. The swelling at this time did not affect the cheek. The child retched several times during the next few days. She was admitted to hospital with a provisional diagnosis of angioneurotic oedema. The symptoms and signs gradually diminished and a fortnight later the patient was discharged with her eye 'almost normal'. Since that time she had had attacks of pain in the left eye at intervals of about 6 months, each lasting a few days. Her mother thought the child had little or no vision in the eye at this time. A more persistent attack of pain began in November, 1953, and the patient was referred for neurosurgical opinion to the Royal Victoria Hospital, Belfast.

On examination the left eye was slightly depressed and abducted, the upper and lower eyelids were swollen, and the eyeball seemed slightly larger and softer than normal. On ophthalmoscopy a pigmented mass was seen on the lower anterior part of the retina, the anterior margin could not be seen, and the posterior margin did not reach the disc. Tortuous dilated veins and dilated arteries were visible to the left of the tumour, and the disc was partly obscured by a retinal detachment. There was vague perception of light, and slight proptosis of the eyeball. Upward and outward movements of the eyeball were limited. A faint diffuse bruit was heard over both sides of the forehead. X-rays of the skull and orbit revealed no abnormality. The ocular lesion was considered to be an angioma with possibly a further orbital angioma causing the proptosis and bruit.

In March, 1954, the child was admitted for angiography; the eye had then become more prominent, a firm palpable tumour could be felt behind the eyeball, and it was decided to carry out further investigations.

The Wassermann reaction and Kahn test were negative.

The electroencephalogram revealed a normal record. Left carotid angiogram: 'The circulation was rapid and the arterial phase was poorly shown. There was, however, no evidence of an orbital angioma'.

It was therefore decided to explore the orbit, and an operative transfrontal exploration (Mr. Taylor) revealed that the lacrimal gland was enlarged and was attached at its lower end to a firm tumour which seemed to fill the muscle cone. About one-third of the tumour had been resected when it became obvious that the growth was widely infiltrating all the muscles. The operation was left at this stage as a decompression until it was known whether or not radiotherapy would be of value.

The biopsy material included a normal lacrimal gland, a capillary haemangioma, and small fragments of a tumour which was then considered to be probably a mixed tumour of the lacrimal gland and insensitive to x-rays. Orbital exenteration was advised and carried out (Mr. Corkey and Mr. Taylor), but the tumour was found at operation to be infiltrating beyond the orbit.

Recovery from these operations was, at first, uneventful. However, in July, 1954, the patient was seen again, and was then complaining of left frontal headaches and vomiting, these symptoms having commenced some 3 weeks previously. A nodule was noted in the back of the left eye socket, the right disc was slightly swollen, and there was

a suspicion of a right temporal hemianopia. The child was again admitted to hospital. Distended veins became apparent over the frontal region, signs of increased intracranial pressure became marked, and the tumour recurrence in the left orbit increased in size. At the end of July, 1954, to relieve the distressing symptoms of increased intracranial pressure, a Torkildsen's operation (ventriculo-cisternostomy) was performed (Mr. C. A. Calvert).

In the post-operative period there was a distinct improvement in the child's condition, but deterioration soon set in again and over the next 4 months she gradually lapsed into unconsciousness. The orbital recurrence of the tumour grew, ulcerated, and bled, forming a large fleshy protrusion from the orbit; it extended on all sides into the soft tissues and there was tumour present in the left temporal region. The child died on November 1, 1954.

Post-mortem Report.—The body was that of a thin child of about 6 years. There was an ulcerated rounded tumour mass occupying the left orbit. The left cheek and temporal region were swollen. Hydrocephalus was present, there was mild proptosis of the right eye, and in the position of the biparietal burr-holes there were two small, fluctuant swellings, 2 cm. in diameter. The abdominal and thoracic viscera showed no significant macroscopic abnormality.

The skull was thinned by hydrocephalus; underlying it, the cerebral convolutions were flattened, there was herniation of brain tissue through the burr-holes. The brain was difficult to remove as it was bound to the base of the skull by invading, partly necrotic tumour. Most of the anterior and middle lobes were involved basally. The base of the skull showed extensive tumour growth, stemming from the left orbit and invading the ethmoids, the nasal cavity, right orbit, both middle fossae, the sphenoid sinus, and the sella turcica. Tumour was present in the left temporal soft tissues extending directly downwards to involve the parotid gland on that side.

Histology

The tumour shows a variety of features, the most striking being the formation of cellular membranes composed of irregular rows of elongated cells with distinguishable cell borders. These sheets of cells or cell membranes have on one surface a limiting membrane which borders a space (Figs 1 and 5). Mitoses are

FIG. 1.—Cellular membrane lining space which contains hyaline droplets. A limiting membrane is evident. Haematoxylin and eosin. $\times 150$.

FIG. 2.—Showing gradation of cellular membrane into a cuboidal epithelium. Haematoxylin and eosin. $\times 100$.

FIG. 3.—Typical rosettes in dictyoma. Haematoxylin and eosin. $\times 375$.

frequently seen in relation to this limiting membrane. The other surface borders more solid portions of the tumour, loose connective tissue, or structureless eosinophilic material (Fig. 5). These cellular membranes have no regular shape; they are twisted, indented, large and small, merging on the one hand into epithelium of a simpler type only one cell thick and composed of cuboidal cells (Fig. 2) and on the other into small rosettes of the type seen in retinoblastoma (Fig. 3).

In places these membranes merge into solid masses of tumour cells with no structural form. Scattered throughout the tumour are numerous areas of glial tissue, mainly of the oligodendroglial type (Fig. 4). This appearance is also present in the orbital portions of the tumour and is an integral part of the neoplasm.

FIG. 4.—Area of neuroglia. Mallory's phosphotungstic acid-haematoxylin. $\times 125$.

Small islands of cartilage are present in different parts of the tumour; these have no relation to pre-existing bone or cartilage and again appear to form part of the tumour (Fig. 5).

FIG. 5.—Nodule of cartilage adjacent to typical cellular membranes. Haematoxylin and eosin. $\times 100$.

No pigment is seen in the sections examined and no iron-containing pigment is demonstrable by the prussian-blue reaction. An unusual feature is the presence of hyaline droplets in some of the spaces bounded by the cellular membranes (Fig. 1). Wide areas of the neoplasm are necrotic. A few small deposits of tumour are present in random sections of lung (Fig. 6).

FIG. 6.—Small metastasis in lung. Haematoxylin and eosin. $\times 100$.

Discussion

The view that dictyomata were relatively benign in their behaviour (Willis, 1948; Friedenwald and others, 1952) was based on a study of the small number of published cases. In the present case there was extensive direct intracranial and extracranial spread and minute pulmonary metastases were present. In most of the reported cases the eye was removed because of the presence of a small intra-ocular tumour. In this instance the patient was seen, apart from the first episode, at a late stage of the disease, when the eyeball had been penetrated and orbital invasion was established. The coincidence of an orbital haemangioma complicated the clinical picture. It is felt that this case, which differs from previous ones in degree rather than in histological detail, warrants the consideration of this type of tumour as malignant.

Fuchs (1908) ascribed the principal histological features of dictyomata to their basic derivation from the medullary epithelium. Since his descriptions, further recorded cases mention a feature less susceptible of this explanation. Böck (1929), Schepkalowa (1941), Klien (1939), and Fralick and Wilder (1949) all described islands of cartilage in the tumour. The appearance of cartilage, or indeed of any mesenchymal tissues, in embryonic tumours is not uncommon; this develops by aberrant or metaplastic differentiation of

young mesenchymal cells and does not imply inclusion of parts of the sclerotome (Willis, 1948). Klien (1939) offers the facile explanation that the cartilage in dictyomata is due to the inclusion therein of chondroblasts.

As Reese (1951) comments: 'A characteristic of these tumours [dictyomata] is that they have no stroma'. The truth of this statement is apparent when we consider that dictyomata are, by definition and general behaviour, to be regarded as brain tumours arising from derivatives of the primitive neural tube. As such, they can have no intrinsic stromal component other than neuroglia. Remnants of invaded connective tissue act as an apparent stroma for the tumour, and the presence of embryonic tumour tissue may influence the normal connective tissue of the affected region to undergo metaplasia, rather than component mesenchyme as in other embryonic tumours.

Another feature noted in this case was the occasional occurrence of hyaline droplets or spheroids in the spaces bounded by the cellular membranes (Fig. 1). This feature, though not described, is beautifully illustrated in Klien's case. As regards the external limiting membrane, the position of mitotic figures in relation to the membrane, and the general cellular arrangement, these features closely resemble the appearance of early foetal retinal tissue. Carrying the analogy with foetal retinal tissue further, the free surface of the cellular membrane which faces the cavity represents the outer surface of the retina which faces the choroid. The other surface, bounded sometimes by amorphous eosinophilic material (Fig. 5) or by loose vascular tissue, represents the surface facing inwards in contact with the vitreous humour. Reese (1951) suggests that the amorphous material on this surface of the cellular membranes represents an attempt to form vitreous humour.

This tumour behaved in some respects as a tumour derived from cerebral tissue, in that there was extensive direct intracranial spread. The final clinical picture resembled the terminal stages of some cases of retinoblastoma with orbital recurrence, extensive local soft tissue spread, and massive intracranial involvement. The metastasis in the lungs is an unusual feature.

Summary

The clinical and pathological findings in a case of dictyoma are recorded. Some of the features of this tumour are discussed. In this case, the tumour recurred after exenteration of the orbit, invaded the skull and brain, and caused death.

I should like to express my gratitude to Professor J. H. Biggart for his advice during the preparation of this paper, and to Mr. Corkey, Mr. Calvert, and Mr. Taylor for allowing me access to the clinical records.

The photography is the work of Mr. D. Mehaffy, A.R.P.S.

REFERENCES

- ANDERSEN, S. RY. (1948). *Acta ophthalm. (Kbh.)*, **26**, 313.
 ASBURY, M. K. (1944). *Amer. J. Ophthalm.*, **27**, 1094.
 BADAL, —., and LAGRANGE, F. (1892). *Arch. Ophthalm.*, **12**, 143.

- BAILEY, P., and CUSHING, H. (1926). "Tumors of the Glioma Group". Lippincott, Philadelphia.
- BIGGART, J. H. (1949). "Pathology of the Nervous System", 2nd ed., p. 293. Livingstone, Edinburgh.
- BÖCK, J. (1929). *Z. Augenheilk.*, **69**, 17.
- CZUKRÁSZ, I. (1941). *v. Graefes Arch. Ophthalm.*, **142**, 619.
- EMANUEL, C. (1900). *Virchows Arch. path. Anat.*, **161**, 338.
- FRALICK, F. B., and WILDER, H. C. (1949). *Trans. Amer. ophthalm. Soc.*, **17**, 317.
- FRIEDENWALD, J. S., WILDER, H. C., MAUMENEE, A. E., SANDERS, T. E., KEYES, J. E. L., HOGAN, M. J., OWENS, W. C., and OWENS, E. U. (1952). "Ophthalmic Pathology", p. 405. Saunders, Philadelphia.
- FUCHS, E. (1908). *v. Graefes Arch. Ophthalm.*, **68**, 534.
- GASTEIGER, H. (1949). *Dtsch. Gesundheitswes.*, **4**, 530; *Ext. in Excerpt. med. (Amst.)*, Sect XII, **4**, 762 (1950).
- GREEVES, R. A. (1911). *Trans. ophthalm. Soc. U.K.*, **31**, 261.
- IMRE, H. (1941). *Arch. Ophthalm. (Chicago)*, **25**, 629.
- KLIEN, B. A. (1939). *Ibid.*, **22**, 432.
- KUTHE, R., and GINSBERG, S. (1905). *Beitr. Augenheilk.*, "Festschr. Julius Hirschberg", p. 127. Veit, Leipzig.
- REDSLOB, E. (1923). *Bull. Ass. franç. Cancer*, **12**, 573.
- REESE, A. B. (1951). "Tumors of the Eye", p. 50. Cassell, London.
- RUBINO, A. (1941). *Ann. Ottal.*, **69**, 385.
- SATANOWSKY, P. (1928). *Arch. oftal. B. Aires*, **3**, 636.
- , and BERNASCONI CRAMER, E. (1938). *Ibid.*, **13**, 523; *Abs. in Amer. J. Ophthalm.* (1939), **22**, 1061.
- SCHEPKALOWA, W. M. (1938). *Vestn. Oftal.*, **13**, 95; *Abs. in Zbl. Ophthalm.* (1939), **43**, 381; *Arch. Ophthalm. (Chicago)*, **24**, 415. (1940).
- (1941). *Ibid.*, **18**, 323.
- SOUDAKOFF, P. S. (1936). *Arch. Ophthalm. (Chicago)*, **15**, 680.
- STANWORTH, A. (1951). *Trans. ophthalm. Soc. U.K.*, **71**, 225.
- VELHAGEN, C. (1919). *Klin. Mbl. Augenheilk.*, **62**, 571.
- VERHOEFF, F. H. (1904). *Trans. Amer. ophthalm. Soc.*, **10**, 351.
- WILLIS, R. A. (1948). "Pathology of Tumours", p. 895. Butterworth, London.