Medullo-epithelioma (diktyoma) of the eye

M. V. SIRSAT, S. S. SHRIKHANDE, AND M. B. SAMPAT

Department of Pathology, Tata Memorial Hospital, Bombay, India

A medullo-epithelioma (diktyoma) is a rare malignant tumour of the eye which arises from the unpigmented epithelium of the ciliary body; diktyoma of the optic nerve with intracranial extension has also been described (Reese, 1963). The name diktyoma was given by Fuchs (1908) to indicate the net-like appearance of the tumour. We have found fifty examples of diktyoma of the ciliary body, most of them in the German literature. Three cases have been reported from India (Kesavachar and Junnarkar, 1960; Nirankari, Gulati, and Chaddah, 1960; Shivde, Kher, and Junnarkar, 1969). The case reported below is the first to be seen at the Tata Memorial Hospital, Bombay, during the last 30 years.

Case report

A Hindu male child aged 5 years had shown enlargement of the right eyeball for 3 weeks. There was no history of pain or of any past major illness. He was the third child, the other children being healthy and normal.

Examination

The right eyeball was found to be enlarged, with dilated capillaries over the sclera. The visual acuity was diminished. The left eye showed no abnormality. The liver and spleen were not palpable.

Radiology

The bony orbital walls were intact. The right optic foramen was visible. The lungs showed no abnormality.

Operation

Enucleation of the right eye was performed on September 14, 1970.

Result

Recovery was uneventful and the patient was discharged on September 18, 1970. At the last follow-up examination on February 3, 1971, no lesion was detected clinically.

Pathology

GROSS FINDINGS

The specimen received in the laboratory consisted of the eyeball with retro-orbital fat. On opening the eye a glistening white tumour was found; it measured 2.5×2 cm. and filled the entire ocular cavity (Fig. 1).

FIG. 1 *Tumour in cut surface of opened eye*

HISTOLOGY

The tumour consisted of tall columnar cells arranged in rows or forming acini. The cell borders were very prominent. The cells showed a limiting membrane bordering a space (Fig. 2). The spaces showed amorphous acidophilic material and scanty hyaline droplets. At places the tumour showed rosettes similar to those seen in retinoblastoma (Fig. 3).

FIG. 2 *Photomicrograph, showing tall columnar cells with a limiting membrane. Haematoxylin and eosin. $\times 105$*

FIG. 3 *Photomicrograph showing rosettes. Haematoxylin and eosin. $\times 120$*

Scattered areas of glial tissue were seen (Fig. 4). Other characteristic features were the presence of islands of cartilage scattered throughout the tumour with areas of calcification and the scanty stroma (Fig. 5). The tumour was seen to infiltrate the layers of the eyeball, and the retro-orbital fat.

FIG. 4 Photomicrograph, showing areas of glial tissue. Haematoxylin and eosin. $\times 60$

FIG. 5 Photomicrograph, showing areas of cartilage. Haematoxylin and eosin. $\times 60$

Discussion

The most common tumour involving the eyeball in children is retinoblastoma. From this laboratory, Shirkhande and Sirsat (1966) have reported a study of 100 cases of retinoblastoma in Indian children but the case reported above is the first ocular diktyoma.

This tumour arises from the embryonic retina and corresponds to those of the kidney (nephroblastoma) and liver (hepatoblastoma). Like retinoblastoma, diktyoma is more common in children between the ages of 3 and 5 years, the oldest patient was one reported by Soudakoff (1946) in a man aged 28 years. In contrast to retinoblastoma, however, diktyoma is never bilateral or multicentric in origin and there is no evidence of a hereditary factor.

Diktyoma is a slowly-growing tumour and extends locally into the surrounding tissue. It very rarely shows extraocular extension and metastasis, although Malone (1955) reported a case of diktyoma which showed extensive intracranial spread and metastases in the lung. The tumour grows more slowly and has a better prognosis than retinoblastoma (Cardell and Starbuck, 1959).

Many of the structural characteristics are well illustrated in the present case. In diktyoma features of any stage of the developing retina may be reproduced. In its typical form the tumour shows a predominant net-like polycystic structure with cysts lined by single or multilayered epithelium. Rosette formation is common and the solid areas resemble the structure of a retinoblastoma. Other features are the fibrillary structure and the scanty connective tissue structure. Necrotic foci are small and infrequent. Islets

of cartilage are seen in this tumour as in other embryonal tumours elsewhere in the body; the cartilage develops by aberrant or metaplastic differentiation of young mesenchymal cells.

Summary

A case is reported of diktyoma of the eye occurring in a 5-year-old child. It is the first seen at the Tata Memorial Hospital, during the past 30 years.

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