

Diktyoma

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Only about fifty cases of this extremely rare tumour arising from the pars ciliaris retinae in young children have been recorded so far (Cardell and Starbuck, 1959). This is the second case to be reported from our hospital; the first was described by Kesavachar and Junnarkar (1960).

Case report

A girl aged 3 years was admitted to the Plastic Surgery Department in December, 1967, with pain and total loss of vision in the left eye of 3 months' duration. There was a history of injury to the left eye and the parents said that the swelling and loss of vision began a few months after the injury.

The right eye was normal, and there was no family history suggestive of retinoblastoma.

Examination

The left eye, which was totally blind, was proptosed and with very swollen lids. The cornea and sclera were completely destroyed and replaced by swelling which was immobile and fixed to the superolateral orbital margin. There was no lymphadenopathy of the pre-auricular lymph gland.

Treatment

The eye was enucleated and sent for histopathological diagnosis with a provisional diagnosis of retinoblastoma.

Pathological report

MACROSCOPICAL The specimen was an irregular oval tumour mass, measuring approximately 45 mm. in the sagittal, 30 mm. in the frontal, and 35 mm. in the vertical diameter. The cornea and lens could not be identified. Externally the tumour was an irregular nodule of greyish-white colour (Fig. 1).

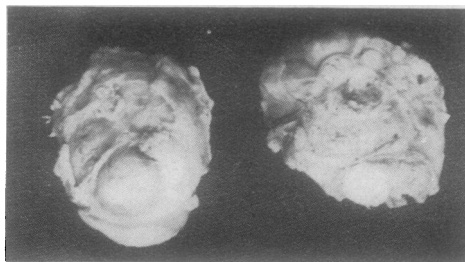


FIG. 1 *External surface of tumour (actual size)*

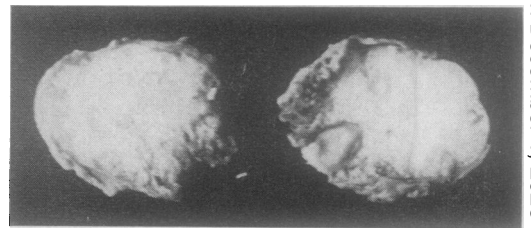


FIG. 2 *The tumour bisected (actual size)*

On bisection the cut surface showed yellowish opaque tumour tissue, with tiny cystic structures containing clear fluid (Fig. 2). The chambers of the eyeball could not be distinguished. On further dissection, a small cyst-like structure surrounded by pigment and measuring about 12 mm. in the

sagittal diameter was identified as the original eyeball which had been pushed to one side after being invaded and surrounded by tumour tissue.

MICROSCOPICAL Sections stained with haematoxylin and eosin and Masson's trichome stains were studied. The anterior portion showed a normal conjunctival epithelium, but the deeper portion was completely occupied by tumour tissue composed mainly of tubules and bands of several layers of columnar cells with prominent oval nuclei. All these tubules and bands showed a distinct basement membrane. Most of the tubules were filled with faint eosinophilic material resembling vitreous substance. Some of the tubules were empty.

In one section there were round cells with scanty cytoplasm arranged in solid sheets, showing a rosette formation in places and resembling retinoblastoma (Fig. 3). In another there was well-formed cartilage surrounded by tubules and bands of columnar cells (Fig. 4). There was glial tissue in most of the sections. The connective tissue stroma as demonstrated by Masson's stain was very scanty. No pigment could be seen in any of the sections.

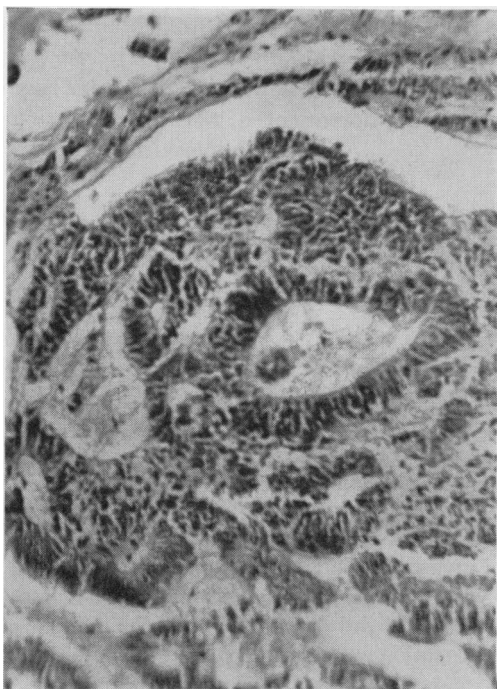


FIG. 3 Tumour tissue composed of columnar cells with oval nuclei presenting a tubular arrangement with distinct limiting membrane. $\times 250$

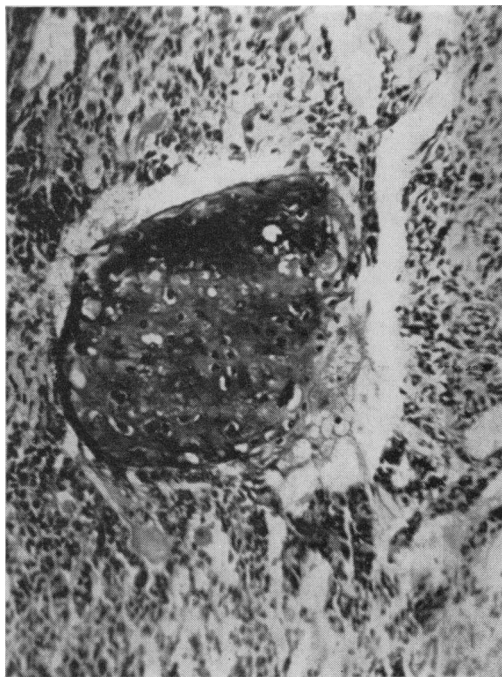


FIG. 4 Cartilage surrounded by tumour tissue. $\times 250$

Diagnosis

These histological findings were considered to be characteristic of diktyoma.

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References

- CARDELL, B. S., and STARBUCK, M. J. (1959) *Brit. J. Ophthalm.*, **43**, 217
 KESAVACHAR, K. R., and JUNNARKAR, R. V. (1960) *Ibid.*, **44**, 693