Rhabdomyosarcoma of the iris

Report of the first recorded case

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Neoplasms of the iris are uncommon, but a number of authors, on the basis of their own cases, have reported some scores of primary iris tumours (Duke and Dunn, 1958; Rones and Zimmerman, 1958; Reese and Cleasby, 1959; Zimmerman, 1963; Ashton, 1964; Ashton and Wybar, 1966). The most frequent neoplasia of the iris are malignant melanomata, and the next most frequent group includes smooth muscle tumours. Angiomata, neurilemmomata, neurofibromata, and benign melanotic tumours are rare (Reimer and Wolter, 1969). Single neoplasia of the iris with a doubtful histogenesis have also been recorded in the literature (Duke and Dunn, 1958; Bürki, 1961; Hogan and Zimmerman, 1962), but no rhabdomyosarcoma of the iris has so far been described, and it was therefore felt appropriate to report this case of an iris tumour composed of cells showing distinct cross-striations in the cytoplasm.

Case report

A girl aged 4 years was admitted to the Ophthalmological Clinic of the Medical Academy in Szczecin in June, 1966, with the diagnosis of iris tumour of the left eye. In the months preceding her admission, her mother had noticed an increasing growth in the iris of the child’s left eye, but this had not caused any discomfort.

Examination

In the iris of the left eye between 2 and 3 o’clock, a greyish pink profusely vascularized prominence was seen to spread from the pupillary edge to the base of the iris, protruding towards the posterior surface of the cornea but without touching it (Fig. 1, opposite). A round pupil located centrally reacted to light efficiently, but was significantly slower in the vicinity of the tumour.

The intraocular pressure was normal. The filtration angle was open on gonioscopic examination. The media were clear and the fundus normal. The right eye showed no pathological changes. The visual acuity in both eyes was 1.0.

In the differential diagnosis tuberculosis was excluded, as were malignant melanomata and leiomyomata, as being very unlikely in early childhood. Diktyoma was excluded by the absence of change in the area of the ciliary body. Xanthogranuloma juvenile was therefore accepted as the probable diagnosis. This was suggested by the histological examination of subcutaneous nodules removed from the crown of the head and diagnosed as xanthomata.

Treatment

Following the advice of various previous authors (Cleasby, 1961; Gass, 1964; Hamburg and Koten, 1964), ten x-ray treatments of 100 r each (total 1,000 r) were applied. In the course of 3 months,
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the nodule gradually regressed to half its original size, but in the next 3 months, despite cortisone therapy for 2 weeks and a further five irradiations of 120 r each (total 600 r), the iris tumour began to grow and spread in the direction of the filtration angle as well as upwards. Signs of secondary glaucoma appeared in the left eye, and after the increased intraocular pressure had been lowered by daranide, a sectoral iridectomy was performed on January 31, 1967. A histological diagnosis of rhabdomyosarcoma embryonale was then established.

As a result of the histological diagnosis of a malignant intraocular neoplasm and because secondary glaucoma was increasing, the left eye was enucleated on November 20, 1967.

Result

The child has since been observed for 4 years and at present her development is quite normal. During this period there has been no local recurrence of the neoplasm and no metastasis.

Pathological studies

Histologically, after the enucleation, a thickening of the iris was noted, the whole iris being infiltrated by neoplastic tissue composed of closely packed, distinctly outlined cells, with abundant acidophilic cytoplasm (Fig. 2). The nuclei were round or oval with moderately sized nucleoli. The majority of the cells were oval or polyhedral in shape with eccentric nuclei, but some of them formed "straps" with several linearly arranged...
nuclei (Fig. 3). In the cytoplasm of the latter cells, distinct cross-striations were easily identified (Fig. 4A, B). Mitotic figures were not numerous. The stroma was inconspicuous, with a few thin-walled bloodvessels. The inner pigmented epithelium of the iris was preserved, and the ciliary body was not infiltrated by the neoplastic tissue.

A portion of the iris tumour previously fixed in formol saline and embedded in paraffin, upon hydration was after-fixed with 2 per cent. osmic acid, dehydrated, and embedded in Vestopal according to the method of Hubner (1970). The ultrathin sections double-stained with lead citrate and uranyl acetate were examined under the electron microscope.

The ultrastructure of cells from formol-fixed material is usually rather indistinct, but our electron micrographs showed that the cytoplasm of neoplastic cells was filled with delicate fibrils corresponding to myofilaments arranged in myofibril-type bundles, with abortive sarcomere formation and slightly marked Z-striations. Single myofibril bundles lying

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**Fig. 3** Oval and polyhedral cells with eosinophilic cytoplasm and eccentric nuclei. Strap-cell forms are also seen (arrow). Haematoxylin and eosin. × 420

**Fig. 4** Strap-cell forms of rhabdomyoblasts with cross-striations. Haematoxylin and eosin. (A) × 2,000 (B) × 1,200
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close to each other often ran in various directions, so that the bundles were seen side by side longitudinally and cross-sectioned (Fig. 5). These bundles of fibrils appear to be characteristic of the ultrastructure of rhabdomyosarcoma cells (Kroll, 1967; Freeman and Johnson, 1968; Resnik, Nameroff, and Hansen, 1970). They differ from the electron micrographs of leiomyoma cells (Meyer, Fine, Font, and Zimmerman, 1968).

FIG. 5  Electron micrograph of portion of tumour cell. The cytoplasm contains bundles of myofilaments longitudinally (black arrow) and cross-sectioned (white arrow). N = nucleus.  X 21,000

Discussion

Rhabdomyosarcomata grow relatively often in orbital tissues (Horn and Enterline, 1958; Kassel, Kopenhaver, and Aréan, 1965; Jones, Reese, and Kraut, 1966). As far as we know, this type of iris tumour has not previously been described. The normal iris has smooth muscle tissue which, according to some authors, is of neuroectodermal origin (Brown, Kellenberger, Hudson, and Matthews, 1957). The absence of cross-striated muscle in a normal iris does not, however, render impossible the pathogenesis of rhabdomyosarcoma-type neoplasia, since the latter proliferates quite often in organs devoid of cross-striated muscles: e.g. uterus, urinary bladder, and prostate (Willis, 1962), biliary duct (Horn and Enterline, 1958), meninges (Domagala, 1969), and brain (Legier and Wells, 1967).

The tissue in which these rhabdomyosarcomata originate is probably derived from the omnipotent mesenchyme. We must also remember that the sporadic, malignant, non-classified tumours of the iris composed of polymorphous cells may belong to the group of rhabdomyosarcoma-type neoplasia, although there are no traces of cross-striation. In our case the double course of irradiation may have led to some differentiation of neoplastic cells by making the cross-striations evident in both the light and the electron microscope. It is well known that x rays promote the process of maturation and differentiation of certain neoplastic tissue (Friedman and Drutz, 1958; Glücksmann, 1952).
The final problem is the assessment of the malignancy of the tumour described. Classical morphological features and the infiltrating growth of neoplastic tissue in the entire iris support the diagnosis of a malignant tumour of the rhabdomyosarcoma embryonal type. Special care must be taken in estimating the degree of malignancy, since the biology of iris neoplasia differs from that of similar neoplasia in other regions. The oncological literature emphasizes that, in iris tumours composed either of smooth muscle or of melanotic cells, an estimate of malignancy based exclusively on morphological features is doubtful, because some tumours diagnosed histologically as malignant neoplasia of the iris (e.g. leiomyosarcoma, malignant melanoma) show a clinical course similar to that of benign tumours (Rones and Zimmerman, 1958; Reese and Cleasby, 1959; Zimmerman, 1963; Ashton, 1964).

Summary

The first reported case of a rhabdomyosarcoma of the iris was observed in the left eye of a 4-year-old girl. Histologically, cross-striations were easily identified in the cytoplasm of the tumour cells. Ultrastructurally, the cytoplasm of most of the cells contained a collection of myofilaments arranged in bundles with abortive sarcomere formation. Although the histological features seemed to indicate malignancy, no recurrences or metastases developed in the 4-year period of follow-up.

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