Orbital rhabdomyosarcoma in an infant

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Rhabdomyosarcoma is a rare primary neoplasm in childhood which usually occurs at the age of 7 to 8 years (Jones, Rees, and Kraut, 1966). The youngest patient so far recorded was 2 years old (Lederman, 1956).

Case report

A 1-year-old male Hindu infant was brought to the Eye Out-patient department of Irwin Hospital, New Delhi, on December 14, 1968, with a history of a swelling in the left orbit and protrusion of the eyeball for one month. According to his mother the swelling had first appeared in the lower nasal quadrant of the left orbit and rapidly progressed to the upper nasal quadrant and displaced the eyeball. No history of trauma or any intercurrent infection was available.

Examination There was an eccentric proptosis of the left eye which was displaced forwards, upwards, and outwards (Fig. 1). A soft ill-defined swelling measuring 2×1.5 cm. was palpable on the inner aspect of the eyeball along the medial margin of the orbit. The overlying skin was free but the swelling was attached to deeper structures. The movements of the eye were markedly restricted. Cornea, anterior chamber, iris, and pupillary reactions were normal.

FIG. 1 *Appearance at first examination, showing proptosis*

Fundus examination showed blurring of the disc margins, hyperaemia of optic disc, and increased tortuosity of the retinal vessels in the left eye. Ear, nose and throat examination revealed that the left side of the nose was displaced inwards.

Laboratory investigations Hb 8 g. per cent., leucocyte count 3,100/cu. mm. (polymorphonuclears 64 per cent., lymphocytes 30 per cent., eosinophils 6 per cent.).

X-ray examination of the left orbit showed soft tissue shadow but no bony destruction. The optic foramina were of normal size.

Treatment Tetracycline 125 mg./6 hrly was given by mouth. During the next few days the swelling and proptosis progressed rapidly and the cornea became hazy. Tarsorrhaphy was done, but the stitches gave way and the cornea became ulcerated and perforated. The orbit was explored on

December 18 through a curved incision over the swelling. A soft ill-defined mass was found to extend along the medial wall of the orbit. Part of the tumour was removed and sent for histopathological examination.

Histology The tumour consisted of groups of round to polyhedral cells arranged in an alveolar pattern (Fig. 2). The malignant cells varied in size and had generally a scanty acidophilic cytoplasm which tended to merge with the septa (Fig. 3) and a relatively large hyperchromatic nucleus.

FIG. 2 Section showing alveolar pattern of tumour. Reticulin. $\times 100$

FIG. 3 Section showing thin septa with the tumour cells closely applied to them. The cytoplasm of tumour cells at many places merges with the septa. Haematoxylin and eosin. $\times 450$

A few multinuclear giant cells were discernible (Fig. 4) and a few mitotic figures were present. The tumour had invaded the vascular channels (Fig. 5). No definite cross-striations could be demonstrated. The histological picture was characteristic of the alveolar type of rhabdomyosarcoma.

Progress Radiotherapy with cobalt 60 (200 r for 3 min. 11 sec. daily 5 days a week for 8 weeks). The swelling regressed considerably (Fig. 6). Exenteration was contemplated but the parents refused and took the child away against medical advice. They returned, however, after 5 months and examination revealed no further change in the clinical appearance.

Comment

This patient is the youngest so far recorded. The tumour more often involves the right orbit and the most common site is the upper nasal quadrant. In our case the tumour arose from the lower nasal quadrant of the left orbit.

These tumours are considered to be radioresistant (Reese, 1951; Duke-Elder, 1952); Lederman (1956) found them highly radiosensitive though rarely radiocurable. In this case radiation given as primary therapy was followed by regression of the tumour (Fig. 6).

FIG. 4 Another field of section in Fig. 3 shows a multinucleated giant cell surrounded by several cells showing pleomorphism. Cross-striations could not be demonstrated. Phosphotungstic acid and haematoxylin. $\times 450$

FIG. 5 Section showing invasion of a vascular channel. Phosphotungstic acid and haematoxylin. $\times 450$

FIG. 6 Appearance after radiotherapy

Summary

A case of rhabdomyosarcoma in a 1-year-old child responded to radiotherapy.

References

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