RHABDOMYOSARCOMA OF THE ORBIT*

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FEW cases of orbital rhabdomyosarcoma, a rare neoplasm of high malignancy, have been described in the literature. The following case report records some interesting features of this type of tumour not hitherto reported.

Case Report

A boy aged 16 years developed a rapidly progressive and painless proptosis of the left eye in November, 1954.

Examination.—The proptosis was axial in nature and the ocular movements were restricted in all directions but especially downwards and outwards. The left pupil was dilated, reacting sluggishly to light. The fundus showed venous engorgement and some papilloedema. In December, 1954, the visual acuity was reduced to counting fingers at 2 ft. All investigations, including skiagrams of the orbital region, were negative. By the end of December, 1954, the proptosis had increased and there was marked chemosis of the conjunctiva. The patient had become very weak on account of continued anorexia and vomiting.

Treatment.—Antibiotics were given for a fortnight without any improvement. Aspiration of the orbit was attempted in vain. By the middle of January, 1955, the patient had developed left-sided headache and insomnia, and showed primary optic atrophy causing total blindness in the left eye (Fig. 1).

A course of deep x-ray therapy was started on January 18, and after the first exposure there was an improvement in all the subjective symptoms. By March 15 a course of 15,000r (three fields) had been completed; there was a complete regression of the proptosis (Fig. 2), and all the subjective symptoms had disappeared, though the chemosed conjunctiva continued to prolapse in the lower part.

FIG. 1.—Before deep x-ray therapy, January, 1955.

FIG. 2.—After deep x-ray therapy, March, 1955.

FIG. 3.—Recurrence before orbitotomy, July, 1955.

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First Recurrence.—On May 1, 1955, the proptosis reappeared and increased rapidly.

A second course of deep x-ray therapy, given between June 1 and 13, resulted in slight improvement only.

First Operation.—The patient first came under observation in July, 1955 (Fig. 3), and a Kronlein's lateral orbitotomy was performed on July 23. While exploring the orbit we found a firm neoplasm of rubbery consistency in the lower part of the orbit near the apex between the optic nerve and the origin of the inferior rectus muscle. With finger dissection the neoplasm could be mobilized in all directions except at its very root near the optic foramen. This pedicle-like root, which cut like a fibrous structure, was divided with scissors and, as far as could be ascertained at the time of the operation, the neoplasm had been removed *in toto* (Fig. 4). All other structures in the orbit were left intact.

Biopsy Report of Primary Tumour (courtesy of Dr. Norman Ashton, Institute of Ophthalmology, London):—"Sections show a highly malignant, densely cellular tumour undergoing focal necrosis in the areas more remote from the thick-walled blood vessels with which it is richly supplied. Much of the growth is undifferentiated and the cells exhibit a striking degree of pleomorphism; the predominant cell is pale and polyhedral, and occasionally shows two or three nucleoli within its nucleus. Other cells show a markedly vacuolated eosinophilic cytoplasm, a few racquet-shaped cells may be seen, and scattered giant cells are present. In some areas there are numerous fibrils and irregular oval or ribbon-shaped eosinophilic masses which show longitudinal or cross striations and resemble fragmented muscle fibres. The histological picture is typical of orbital rhabdomyosarcoma." (Fig. 5).

FIG. 4.—Neoplasm removed on July 23, 1955. $\times 10$. FIG. 5.—Section of neoplasm shown in Fig. 4. $\times 280$.

(Courtesy of Department of Pathology, Institute of Ophthalmology, London.)

Second Recurrence.—On August 20, 1955, there was increased pain in the left orbit, with headache, anorexia, and chemosis of the lower bulbar conjunctiva.

A third course of deep x-ray therapy was started on August 23, and by September 15 a total of 2,000r (two fields) had been given. The patient was discharged completely symptomless on September 30.

Third Recurrence.—On November 4, 1955, he was re-admitted with the same evidence of recurrence as before.

Second Operation.—On November 7, an exenteration of the left orbit was performed. At operation there was a severe haemorrhage from the cut stump despite clamping of the apex, which was apparently incomplete. This was controlled by tight packing, and a blood transfusion was given. Thiersch grafting had to be postponed to a later occasion. After regaining consciousness the patient was discovered to have developed a total right-sided hemiplegia.

Biopsy of the exenterated orbital mass showed the tissues to be grossly infiltrated by the sarcoma.

Progress.—On December 5, 1955, the hemiplegia had considerably improved, but on this day the patient complained of a transient total blindness in the right eye. A skiagram of the left optic foramen showed widespread bony destruction. On December 10 there was inspiratory dyspnoea, which increased so much that a tracheotomy had to be performed on December 18, after which the patient was completely relieved.

First Metastasis.—On December 22, a direct laryngoscopy revealed the presence of a firm subglottal metastatic deposit of the size of a cherry, pink in colour and smooth-surfaced. The taking of a piece of tissue for biopsy produced considerable bleeding. The bronchoscope could not be passed beyond the growth which was reported to be an anaplastic sarcoma.

Fourth Recurrence.—On January 30, 1956, there was a slow local recurrence in the orbit, with pain, headache, vomiting, cachexia, and anaemia. By February 9 a more rapid growth had filled almost half the orbit and a skiagram showed destruction of the posterior ethmoidal cells.

Further Metastases.—A skiagram of the lungs showed metastatic deposits in the right mid zone, left base, and left sternoclavicular region. The tracheal growth was now about 2 in. in diameter, and was palpable from outside.

Third Operation.—On February 13 the neoplasm, which now filled the left orbit, was excised with diathermy coagulation. Palpation of the orbital walls revealed total destruction of the floor and medial wall, and 2 days later the right eye showed primary optic atrophy, and the boy became completely blind.

The patient died on March 3, 1956. Autopsy was not permitted, but as far as could be judged clinically there was no evidence of enlargement of the liver, spleen, or regional lymph nodes until the very end.

Discussion

Describing nineteen cases, Reese and Calhoun (1941) gave an average age of 6 to 7 years for the occurrence of an orbital rhabdomyosarcoma, their youngest patient being 2 and the eldest 13 years of age. Our patient was well outside this age group.

This type of neoplasm has been reported to be more common on the right side with a predilection for the superior nasal quadrant of the orbit, but in our case the site of origin was the inferior temporal quadrant on the left side. A tumour originating from the inferior rectus, as in this case, has been previously described only by Dérier (1929).

Rakov (1937) and Jönsson (1938) reported that rhabdomyosarcomata invariably terminated in death, but three cases of permanent cure after exenteration were reported by Reese and Calhoun (1941). The average span of life reported by Reese and Calhoun (1941) was 1 year and 5 months, and our patient died 1 year and 4 months from the time of the appearance of the tumour. There were four local recurrences: after deep x-ray therapy

after removal by lateral orbitotomy, after orbital exenteration, and after local excision by electro-coagulation. Although temporary regression of the neoplasm took place several times, the metastatic tracheal and lung deposits remained unaffected by deep x-ray therapy.

Several features of the present case are of unusual interest:

(1) The axial proptosis and early optic atrophy leading to blindness suggested a possible optic nerve tumour. The proximity of the neoplasm to the optic foramen caused pressure atrophy of the nerve. This is a feature not reported hitherto, although papilloedema was noted in one case by Reese and Calhoun (1941).

(2) The diameter of the optic foramen was enlarged by about half an inch by early bony destruction.

(3) The temporary dramatic response of the primary tumour to irradiation was followed by a lack of response of the recurrent orbital tumour and the tracheal metastasis.

(4) The neoplasm was easily mobilized on finger dissection, and the apparent total eradication of the neoplasm by orbitotomy promised a good outcome, but was soon followed by a local recurrence.

(5) The occurrence of a temporary hemiplegia immediately after exenteration may have been due to a blood clot or an embolism by a cluster of malignant cells in the internal capsule, the latter favouring an early intracranial invasion.

(6) A tracheal metastasis has not previously been reported.

(7) Optic atrophy and total blindness in the other eye has not been reported previously. This could have occurred only by direct extension causing destruction of the basisphenoid and pressure atrophy of the opposite nerve.

(8) The histology of the tracheal growth, which proved to be an anaplastic sarcoma, showed the spindle cells changing to round cells in metastases, as described by Reese and Calhoun (1941).

Summary

A case of rhabdomyosarcoma of the orbit is described. Apart from its typical characters, this case showed certain new clinico-pathological features, which are discussed in light of the available literature.

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