

UNUSUAL CASE OF DOUBLE PRIMARY ORBITAL TUMOUR*

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THIS case is reported as being of interest from three points of view:

- (1) Multiple primary malignant tumours are themselves rare in children.
- (2) Two primary tumours of different pathological nature occurring in the same orbit at different times are very rare.
- (3) New primary tumours occurring in areas treated by radiotherapy for a different type of tumour are rare.

The case concerns a schoolgirl who died at the age of 10 years from a meningioma arising in the roof of the orbit 6 years after surgical removal of a tumour of the optic nerve, gliomatous (astrocytoma) in nature. The orbit was treated with deep *x*-ray therapy after removal of the first tumour.

Case Report

History.—A little girl, born in 1942, was noticed at the age of 3 to screw up her left eye when looking at anything. She was taken to the Princess Margaret Hospital for Children and approximately a year later a tumour of the left optic nerve was removed through a Krönlein approach, the eyeball being left *in situ* and the tumour being apparently excised completely. Although the tumour was thought to have been completely removed and was reported by the pathologist as an astrocytoma, deep *x*-ray therapy was given to prevent possible recurrence. Deep therapy was begun in April, 1946, over a 7 × 7-cm. field including the whole orbit and optic foramen. In 2 weeks a dose of 3,300 r spread over ten applications had been given, with an estimated tissue dose of 2,500 r at the optic foramen. During May, June, August, September, October, and December, 1946, and January, February, and April, 1947, single monthly doses totalling 6,500 r (tissue dose 5,500 r) were given. A large *x*-ray burn, involving the cornea and conjunctiva and producing telangiectases of the lids, resulted. She was treated for the corneal ulcer which healed with obliteration of the fornices, adhesion of the lids to the globe and keratinization of the corneal epithelium. Eyebrow and eyelashes were permanently destroyed.

Recent Developments.—She was seen for the first time by one of us (P.C.Y.) in 1950, and at intervals until June, 1951. She remained well until this time when her mother noticed a swelling above the left supra-orbital margin. She was readmitted to the Princess Margaret Hospital and a portion of the swelling was removed for biopsy. The section showed a meningioma, and she was transferred to the Neuro-Surgical Unit of the Royal Perth Hospital. When presented to this Unit the child was suffering from a meningioma arising from the dura over the left frontal lobe. The tumour had invaded and destroyed a considerable portion of the adjacent frontal bone and presented as a soft swelling immediately beneath the skin above the left supra-orbital ridge. Tomographic studies demonstrated a large, roughly circular hiatus in the supra-orbital margin, and the anterior part of the frontal bone.

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Operations (J. P. A.).—A large meningioma (1½ inches in diameter) was found arising from the dura mater immediately above the left supra-orbital ridge. The dura, the tumour, and a portion of the frontal bone were excised in one piece, and the tumour appeared to have been completely removed together with a surrounding area of uninvolved tissue. Removal of the tumour presented no difficulty and it was widely removed together with at least ½ an inch of apparently uninvolved dura mater and a similar margin of uninvolved bone (Fig. 1). The extent of removal was not limited by involvement of any important structure and recurrence of the tumour was not expected. The operation left a defect of bone (a considerable portion of the left frontal bone, the supra-orbital ridge, and the anterior half of the orbital plate), and of dura which was repaired with fascia lata. An impression (in dental Stent) was taken and the skin flap closed. The wound healed. Fig. 2 shows the appearance of the child at that time. The flattening of the orbital margin and the old *x*-ray burn of the lids and eyeball can be seen.

A plastic operation (I.M.) was performed in February, 1952, with the object of freeing the lids from the scarred globe so that a prosthesis could be worn, but the skin of the upper lid was found to be firmly adherent to the dura and only partial mobility was attained.

A further operation (J.P.A. and I.M.) to free the dura and to insert a contact shell covered with a split thickness skin graft was performed but the outer canthus broke down later and the shell extruded.



FIG. 1.—Tumour removed in June, 1951. (Scale in inches).

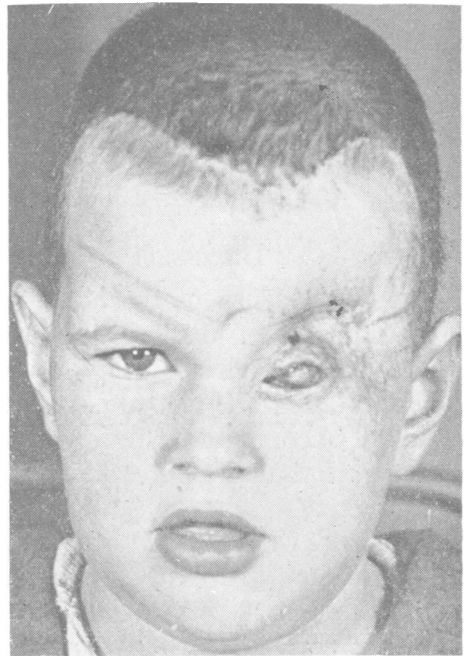


FIG. 2.—Appearance of patient in July, 1951.

Later Developments.—At this time (March, 1952) a smooth painless swelling half an inch wide by three quarters of an inch long appeared in the left lower lid. The appearance of a tumour in the tissues of the lower eyelid at a stage when other evidence of recurrence was absent was totally unexpected. A further tumour also appeared above the left eye and complete exenteration of the left orbit was performed on March 27, 1952. It was

obvious that the meningioma had recurred with extensive involvement of ethmoidal cells and that it could not be completely removed. The wound healed and the child remained well for 3 months.

By July, 1952, the tumour had again recurred and was now the size of a tennis ball. The child had vomited once or twice and there was early papilloedema in the remaining eye. The portion of the tumour mass occupying the left orbit was very vascular and the patient was readmitted to hospital on account of hæmorrhage from it. A month later, during which there had been no bleeding and no deterioration of general health, the tumour had increased in size and had begun to overhang the right orbit and obstruct the visual field mechanically. It had also extended through the left ethmoid and protruded from about half an inch from the left nostril (Fig. 3). This protrusion was curetted away for the comfort of the patient but her condition then deteriorated rapidly. She became comatose and died on September 29, 1952, 15 months after the appearance of the second tumour.

Pathologist's Report

The mass removed from the left orbit and the mass on the left cheek is described as follows: "The supra- and infra-ocular tumours appear to be of the same type. Both show many mitotic figures and nuclear anaplasia. The infra-ocular tumour is the more malignant, yet shows a more typical meningiomatous structure than the upper tumour. Both are malignant meningiomata. The section of the eyeball itself shows absence of retinal structure."

Discussion

The co-incidence of the occurrence of two tumours of dissimilar histological type at an interval of 6 years in a child is sufficiently rare to merit further consideration. The first tumour, a glioma (astrocytoma) of the optic nerve, was itself uncommon. Hudson (1912) found reports of less than 350 cases of optic nerve tumours of all kinds in all the medical literature; of these 118 were gliomata. Between 1912 and 1930, 61 further cases of optic nerve tumour were reported, 52 of them gliomata (Mathewson, 1930). Of the optic nerve tumours, therefore, glioma is the most common; 75 per cent. occur within the first 10 years of life and 88 per cent. before the age of 20. They are more common in females than males. They are slow growing and do not metastasize. If the tumour is completely removed the prognosis is good. Death, when it occurs, is from intra-cranial extension and may be delayed for years (e.g., 26 years in a case reported by Pagenstecher, 1902). *X* rays are considered by Duke-Elder (1940) to be of little value.

Meningioma in children is rare. It usually occurs between the ages of 30 and 50 and is equally common in males and females. The tumours tend to be slow growing, encapsulated, and non-metastasizing. Very occas-



FIG. 3.—Appearance shortly before death of patient in September, 1952.

ionally, sarcomatous changes are present in them and they recur after operation, but usually they can be removed intact with a good prognosis. Recurrence after apparent complete removal, as in the present case, is rare. On the other hand, Myerson (1942) has described the association of gliomata of the chiasma with neurofibromatosis, but the present patient showed no evidence of Von Recklinghausen's disease.

The occurrence of more than one primary tumour in the same individual is not as rare as was at one time thought. It occurs in approximately 1 per cent. of all cancer cases, according to the British Empire Cancer Campaign Survey of Cancer in London (Harnett, 1952). This survey deals with 15,201 cases; the incidence of multiple primaries in the series is shewn in the Table.

TABLE
INCIDENCE OF MULTIPLE PRIMARY TUMOURS IN 15,201 CASES

Primary Tumours		Previous		Simultaneous		Subsequent	
		Another type	Same type	Another type	Same type	Another type	Same type
All Cases	No.	86	61	65	86	99	21
	%	1		1.1		0.8	
Total (excluding tumours of skin, large intestine, and mouth)	No.	76	17	51	20	92	5
	%	0.6		0.5		0.7	

If double primaries of the skin, large intestine, and mouth are deducted, the percentages are much smaller.

No case of tumour of the optic nerve was recorded in this double series, nor was any case at all found in the 15,201 records.

In another series of multiple primary malignant tumours collected by Stalker, Phillips, and Pemberton (1939), 113 patients out of 2,500 had more than one primary. Their average age was 59.7 years. No example was found in children. Epiblastic and mesoblastic tumours were rarely found together.

American series tend to show higher figures than the London survey. For example, a survey of 2,829 autopsies (Warren and Ehrenreich, 1944) produced an incidence of 6 per cent., and Slaughter (1944), in an analysis of a large series of published surveys, found an incidence of 3.9 per cent. for all cancer cases; none of them were cases of optic nerve tumour or multiple primaries in childhood.

The question arises whether the original radio-therapy given to the orbit after the tumour had been apparently completely removed, could have been in any way connected with the appearance of the very unusually malignant meningioma 5 years after cessation of treatment. In view of the severe x-ray burn of the face involving the orbital margin in the area of origin of the second tumour and the rarity of multiple primary tumours in children, this must

certainly be considered as a possibility. Since gliomata of the optic nerve are benign and of low radio-sensitivity it is not usual to treat the area as a prophylactic against recurrence. We can find no recorded similar case. Indeed, it is not easy to find any cases of second primary tumours arising in irradiated areas. Harnett (1952) quotes the following cases without comment; in two of them at least, the second primary appeared within the area irradiated for the first.

(1) **Female, aged 41.**—Carcinoma of pyriform fossa treated by radiotherapy unsuccessfully. At autopsy a squamous carcinoma was found at commencement of oesophagus.

(2) **Female, aged 62.**—Successfully treated by radium for a growth of the floor of the mouth. 42 months later a squamous carcinoma (from which she died) appeared on the posterior third of the tongue.

(3) **Female, aged 68.**—Rodent ulcers of the skin of the nose and orbital region were successfully treated by radium on two occasions at an interval of 9 years; 13 years later she died of cancer of the lung.

(4) **Female, aged 69.**—A local recurrence following a radical mastectomy was treated by radiotherapy; 6 years later she died of cancer of the pylorus.

(5) **Male, aged 48.**—Rodent ulcers on both ears treated by excision and surface radium; 2 years later he died of carcinoma of a bronchus.

(6) **Male, aged 74.**—Growth of tongue treated by *x*-rays and radium; 1 year later growths appeared simultaneously on upper and lower lips; these were treated again with *x*-rays and radium, but he died of cervical secondaries.

(7) **Male, aged 72.**—Carcinoma of tongue excised, 7 months later treated by radiotherapy, and a rodent of the cheek by *x*-rays; 2 years later a third growth appeared on the tongue. Recurrence cannot be excluded.

(8) **Male, aged 65.**—Epithelioma of tongue treated by excision; 1 year later rodent ulcer of the ear treated by radium; 1 year after this epithelioma of the lip appeared.

Again, in this series, the age is high and the tumours all epithelial in origin. The carcinogenic action of the radiotherapy in some of them cannot be excluded, but is not as likely as in our case, which combines several rare features; namely, the occurrence of multiple primaries in a child, the occurrence of a second primary (meningioma) following a glioma of the optic nerve, the unusually high malignancy of the latter, and the occurrence of the second primary within the irradiated area.

Summary

A case is described in which a glioma of the optic nerve was successfully removed from a patient aged 3 years. Irradiation, which was presumably given to prevent recurrence, led to a wide-spread *x*-ray burn, and was followed 6 years later by a highly malignant meningioma involving the frontal bone, from which the child died.

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