Neoplasm of mixed mesenchymal and neuroepithelial origin of the optic nerve

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SUMMARY A case is reported of a 19-year-old male having right proptosis for 4 years because of a mixed meningioma and astrocytoma of the ipsilateral optic nerve. The sheath of this nerve is analogous to the leptomeninges, and neuroglial cells constitute the stroma of the nerve. Both meningioma and astrocytoma therefore can arise primarily in the optic nerve, and they may be combined in the same mass, forming a neoplasm of mixed mesenchymal and neuroepithelial origin.

Several instances have been recorded of neoplasm of mixed mesenchymal and neuroepithelial origin of the neuraxis but none involved the optic nerve (Shuangshoti and Panyathanya, 1974). Subsequently a unique case was encountered of an optic tumour of mixed mesenchymal and neuroepithelial type consisting of combined meningioma and astrocytoma. The tumour arising in the same optic nerve and producing progressive unilateral proptosis is now described.

Case report

A Thai male aged 18 years first noticed progressive exophthalmos associated with blurred vision of the right eye for 4 years before admission to hospital. Two years later the same eye was painful intermittently. One year before admission this eye was blind, but the ocular pain disappeared spontaneously. At the time of admission to hospital the patient complained of frontal headache.

EXAMINATION

The body temperature was 36.6°C, pulse 72, respirations 20, and blood pressure 120/80 mmHg. The right eye showed proptosis, conjunctival congestion, and central corneal opacity. The right pupil, 6 mm in diameter, was not reactive to light; the left pupil, 5 mm across, reacted well. The visual acuity of the left eye was 6/9, but there was no light perception in the right. In the right eye the retina was atrophic and the optic disc pale, but the left eye was normal.

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Routine studies of the blood and urine yielded normal findings. X-rays of the orbits showed on the right side an enlarged optic foramen and thinning of the orbital walls in comparison to the left (Fig. 1). A right carotid angiogram revealed stretching of the intracranial portion of the internal carotid artery. The clinical impression was a retrobulbar tumour with intracranial extension.

OPERATION

The roof of right orbit was excised by an ipsilateral frontal craniotomy one day after the patient was admitted to hospital. A circumscribed retrobulbar mass, 2 cm in greatest dimension, arising in the right optic nerve and extending intracranially through the right optic foramen was almost totally removed. The left optic nerve was thin.

COURSE

Postoperatively the patient gained consciousness spontaneously. Nevertheless the left eye became blind, and diabetes insipidus occurred. A large volume of urine of low specific gravity (1.000 to 1.010) was recorded—as much as 6000 ml on some days. However, the diuresis was controlled by the administration of pitressin. The patient was eventually discharged four months after admission and did not return to the hospital.

PATHOLOGY

The specimen (Siriraj Hosp. S-17-5145) was fixed in 10% formalin and embedded in paraffin. The sections were stained with haematoxylin and eosin. Wilder's stains for reticulin fibres and Mallory's phosphotungstic acid haematoxylin (PTAH) stains for neuroglial fibres were also used.

Fig. 1 Orbital x-ray showing greater size of the right optic foramen (arrow, right) than of the left (arrow, left). Thinning is also noted of the orbital wall on the right side

Fig. 2 Portion of meningioma in the combined tumour from the right optic nerve showing: A. Sheet of meningiocytes, two psammoma bodies at the right upper corner, and a calcific focus toward the left margin of the photomicrograph. Note syncytial appearance and occasionally vacuolated nuclei of meningiocytes (H and E. $\times 100$); B. Whorl of meningiocytes, a diagnostic hallmark of the meningioma (H and E. $\times 100$)

Fig. 3 Portion of astrocytoma in the mixed tumour from the right optic nerve illustrating: A. Neoplastic astrocytes with numerous processes (H and E. $\times 100$). B. More detail of neoplastic astrocytes. Note angular or elongated outlines with coarse processes, glassy cytoplasm, and vesicular and ovoid nuclei. Some astrocytes are multinucleated (H and E. $\times 400$). C. Stromal blood vessels with considerably thick and hyalinised walls (H and E. $\times 100$)

Microscopically there were two different types of neoplastic cells forming the mass. One type was meningocytic (Fig. 2). The cells were plump and syncytial in appearance. The ovoid and vesicular nuclei were occasionally vacuolated. These meningocytes were arranged mainly in sheets and whorls within the space between the sheath of the optic nerve and the dura mater, with minimal infiltration into these two membranes. However, they did not extend beyond the dura mater or into the underlying optic nerve. A few psammoma bodies were scattered in this part of the neoplasm. Many coarse collagen fibres, a few reticulin fibres, and a moderate number of small thin-walled blood vessels composed the stroma of the tumour.

The other type of tumour cells was astrocytic (Fig. 3). They were disseminated within the optic nerve and characterised by their frequently angular outlines, with processes extending from corners and glassy cytoplasm. Some cells were elongated with polar processes. These processes were blue in PTAH preparations and were occasionally attached to the

vascular adventitia. The vesicular nuclei were often eccentrically placed. The stromal blood vessels in this part of the tumour frequently showed thick and hyalinised walls, a characteristic vascular feature in the astrocytoma involving the optic region. Reticulin fibres were absent except for a few round some blood vessels. Foamy and haemosiderin-laden phagocytes were scattered in the lesion.

COMMENT

The diagnosis was a neoplasm of mixed mesenchymal and neuroepithelial origin (combined meningioma and astrocytoma) of the right optic nerve. The terms 'mixed' or 'combined' imply mingling of multiple types of neoplastic cells of both mesenchymal and neuroepithelial origin in the same mass or in juxtaposition.

Discussion

The development of the visual apparatus has been reviewed (Shuangshoti, 1973a). The sheath of the

optic nerve is analogous to the leptomeninges; it contains meningocytes (arachnoidal cells), the mesenchymal derivatives. A meningioma can thus arise primarily in the optic sheath (Shuangshoti, 1973a; Shuangshoti and Panyathanya, 1973).

The retina is derived from the neuroepithelium originally lining the prosencephalon. The retinal ganglion cells yield nerve fibres which extend backwards into the optic stalk and become the optic nerve, chiasm, and tract (Shuangshoti, 1973a). The stroma of the retina and the optic nerve consists of glial cells of both neuroglial and microglial varieties, as in the central nervous system (Friedenwald *et al.*, 1952). The astrocytes and oligodendrocytes constitute the neuroglial interstitium of the retina and the optic nerve. Astrocytoma and oligodendroglioma therefore can arise in the retina as well as in the optic nerve (Jordano *et al.*, 1974). Some optic astrocytomas are associated with phacomatosis, as with tuberous sclerosis and neurofibromatosis (Shuangshoti and Panyathanya, 1972; Jordano *et al.*, 1974), but not in the present case.

In this case a unitary diagnosis cannot be made of either meningioma or astrocytoma because of the mingling of neoplastic meningocytes and astrocytes in the same mass. It is thus appropriate to call the lesion a neoplasm of mixed mesenchymal and neuroepithelial type. This category of mixed tumours comprises 1.6% of our previously reported series of 1028 intracranial neoplasms from Thailand (Shuangshoti and Panyathanya, 1974).

The components of neoplasm of mixed mesenchymal and neuroepithelial origin are diversified in accordance with the multipotential differentiation of mesenchyme or its derivatives (Willis, 1960) as well as commutation among different types of neoplastic neuroglia (Zimmerman, 1955; Shuangshoti and Netsky, 1971). The mesenchymal component in a mixed tumour thus may be a meningioma, rhabdomyosarcoma, neurilemmoma, lymphoma, plasmacytoma, and so on. The neuroepithelial part may be composed of various types of glioma or ganglioglioma. It is therefore not surprising to encounter mixed tumours of meningioma, lymphoma, and glioblastoma multiforme (Shuangshoti and Netsky,

1970); rhabdomyosarcoma, reticulum cell sarcoma, fibrosarcoma, and astrocytoma (Shuangshoti and Netsky, 1971); meningioma, ependymoma, and astrocytoma (Shuangshoti *et al.*, 1971; Shuangshoti, 1973b); lymphoma, plasmacytoma, astrocytoma, and ganglioneuroma (Shuangshoti and Samranvej, 1975); meningioma and oligodendroglioma (Tanaka *et al.*, 1975); and meningioma and astrocytoma as in the present tumour. The mode of occurrence of these neoplasms of combined mesenchymal and neuroepithelial origin has been discussed (Shuangshoti and Netsky, 1971).

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