Primary intracranial rhabdomyosarcoma producing proptosis

SAMRUAY SHUANGSHOTI AND CHARE PHONPRASERT

*From the Departments of Pathology and Surgery (Division of Neurosurgery),
Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand*

SYNOPSIS A case is reported of a primary rhabdomyosarcoma occurring in the right subfrontal region of a 16 year old girl. The patient suffered from dull frontal headache and proptosis for three months before hospitalization. The circumscribed and demarcated neoplasm involved the dura mater, and invaded the frontal sinus and roof of the orbit on the same side. The pathogenesis of the tumour is thought to be related to aberrant differentiation of unstable mesenchyme. A suggestion is made that the 'medullomyoblastoma' should be classified as a type of neoplasm of mixed mesenchymal and neuroepithelial origin.

Primary neuraxial sarcomas are rare. Butler and Netsky (1973) in reviewing nine reported series containing 27 487 intracranial neoplasms found only 423 primary sarcomas (1.5%). Shuangshoti and Panyathanya (1974) encountered 16 primary sarcomas among 1028 intracranial tumours in Thailand (1.6%). Most cases described are of meningeal sarcoma, sarcomatous (malignant) meningioma, fibrosarcoma of the meninges, cerebellar or circumscribed arachnoidal sarcoma, sarcoma of lymphoreticular origin (lymphoma, including microglioma and Hodgkin's disease), liposarcoma, and chondrosarcoma (Berger, 1928; Bailey and Ingraham, 1945; Zimmerman *et al.*, 1956; Kernohan and Uihlein, 1962; Zülch, 1965; Raskind and Grant, 1966; Kothandaram, 1970; Shuangshoti *et al.*, 1970; Rubinstein, 1972). The 'monstrocellular sarcoma or circumscribed sarcoma of the blood vessels' (Zülch, 1965) and 'giant cell sarcoma' (Kernohan and Uihlein, 1962) have been considered to be neoplasms of mixed mesenchymal and neuroepithelial origin (Shuangshoti and Netsky, 1971; Shuangshoti, 1973).

Primary intracranial rhabdomyosarcoma is extremely rare and the same tumour is uncommon elsewhere. Only one of 16 primary sarcomas among 1028 intracranial tumours reported by Shuangshoti and Panyathanya (1974) was a rhabdomyosarcoma, a proportion of 0.1% of intracranial neoplasms. This case of cerebellar rhabdomyosarcoma was reported separately (Shuangshoti *et al.*, 1968). An additional example is now described of a primary subfrontal rhabdomyosarcoma producing exophthalmos in a girl.

CASE REPORT

A 16 year old girl was admitted to the ENT service because of dull frontal headache and progressive proptosis on the right side for three months. The abnormal findings on examination consisted of exophthalmos, conjunctival chemosis, and limited movement in all directions of the right eyeball. The equal pupils were normal in size and reactive to light. The visual acuity was 20/40 in the right eye, and 20/30 in the left. The visual fields were full. Papilloedema was present. Radiographs of the skull and tomograms of the orbits revealed destruction of the orbital roof and of the frontal and ethmoidal sinuses on the right side. There was density of the soft tissue in the region of these sinuses.

Address for reprints: Dr. Samruay Shuangshoti, Department of Pathology, Faculty of Medicine, Chulalongkorn University, Bangkok 5, Thailand.

(Accepted 7 January 1976.)

A mucocoele of the frontal sinus with invasion of the right orbit was suspected, leading to the exploration of this sinus *via* a bifrontal scalp flap on the third day of hospitalization. However, a vascular mass was disclosed in the sinus, and a frozen section was reported as malignant.

Four hours later, the patient was in deep coma, and showed decerebrate posturing on painful stimulation. The fully dilated pupils were not reactive to light. After the neurosurgical consultation, bilateral carotid angiography revealed elevation of the anterior cerebral artery, a prominent ophthalmic artery, and vascular staining in the orbital and subfrontal regions on the right side (Fig. 1). The findings were interpreted as consistent with the presence of a mass lesion in the subfrontal area which was confirmed at right frontal craniotomy. A demarcated subdural tumour was firmly attached to the thick dura mater. It invaded the frontal sinus and roof of the orbit, and partially protruded into the orbital fossa. Portions of the mass within the cranium and frontal sinus were extirpated with the dural covering. A small part was left within the orbit.

Postoperatively, the patient remained permanently unconscious, although the pupils returned to normal size and the decerebrate posture disap-

peared. She finally died three months after the craniotomy. Necropsy was not permitted. However, while she was in hospital, no other tumours were discovered elsewhere on repeated examination of the patient.

The specimen (Chula. Hosp. No. S-18-5688) consisted of a firm mass weighing 60 g and having a finely trabecular and grey cut surface (Fig. 2A). Multiple blocks of the formalin-fixed tissue were embedded in paraffin, and sections were stained with haematoxylin and eosin (H and E), phosphotungstic acid haematoxylin (PTAH), Masson's trichrome, and Wilder's method for reticulin fibres.

Microscopically, numerous pleomorphic cells were arranged in trabeculae (Fig. 2B), papillae (Fig. 2C), sheets, and incompletely formed alveoli. The following types of neoplastic cells were noted: rounded cells with either homogeneous or granular brightly acidophilic cytoplasm (Fig. 2D) or cytoplasmic vacuoles, tadpole-shaped cells (Fig. 2D), and elongated strap cells with polar or central nuclei (Fig. 2D at arrow, and Fig. 2E). A few elongated cells had distinctive cross-striations (Fig. 2E). The tumour cells with eosinophilic perikaryon in H and E stained sections were purple in Masson's trichrome stains, and blue in PTAH

FIG. 1 A subtraction photograph of the right carotid angiogram showing elevation of the anterior cerebral artery (upper arrow), prominent ophthalmic artery (lower arrow), and vascular blush in relation to a mass lesion in the subfrontal and orbital regions.

FIG. 2 A. Bisected specimen of the tumour removed from the right subfrontal area demonstrating circumscribed border (lower half) and finely trabecular cut surface (upper half). The considerably thickened dura mater (arrows) is incorporated in the mass. B. The pleomorphic tumour cells are arranged in trabeculae separated by a vascular stroma. The arrow points towards an elongated cell which is further illustrated in E. H and E, $\times 75$. C. A papilla consisting of pleomorphic rounded neoplastic cells around the connective tissue core containing a blood vessel is exhibited. H and E, $\times 300$. D. A tadpole-shaped cell is surrounded by rounded tumour cells. Some of the latter contain cytoplasmic eosinophilic granules (arrow). Masson's trichrome, $\times 880$. E. An elongated neoplastic cell indicated by the arrow in B has unequivocal cross-striations. H and E, $\times 880$.

preparations. They were interpreted as rhabdomyoblasts. Neuroglial fibres were not observed. The reticulin and collagen fibres were confined mainly to the fibrous septa and their immediate vicinity. They were few in the cellular aggregates.

The diagnosis was primary alveolar rhabdomyosarcoma of the right subfrontal region involving the dura mater and invading the ipsilateral frontal sinus and orbital fossa.

DISCUSSION

The microscopic features of this tumour, particularly the presence of cells with strongly

acidophilic perikaryon and unequivocal cross-striations, are diagnostic of rhabdomyosarcoma. However, the possibility of metastasis must be excluded. The proptosis suggested that the rhabdomyosarcoma might be primarily retrobulbar in location with subsequent intracranial invasion. Nevertheless, the fact that the main part of the tumour was subdural and attached to the adjacent dura mater indicated its primarily intracranial position. A primary intraorbital tumour with intracranial extension should be chiefly situated epidurally. Failure to find any tumour outside the neuraxis in the

examination and investigation of the patient, six months after the onset of frontal headache and proptosis, is sufficient evidence that this rhabdomyosarcoma was primarily intracranial.

Nevertheless, the present rhabdomyosarcoma must be distinguished from other primary intracranial tumours such as teratoma, embryonal sarcoma (mixed mesenchymoma), and neoplasms of combined mesenchymal and neuroepithelial origin. Elements other than striated muscle were absent in the mass; hence it was not teratomatous. An embryonal sarcoma described by Rubinstein (1972) consisted both of rhabdomyoblasts with distinctive cross-striations and of islands of cartilage. The presence of striated muscular elements alone in the tumour of our patient excludes the diagnosis of a mixed mesenchymoma. Shuangshoti *et al.* (1971) reported a number of neoplasms of mixed mesenchymal and neuroepithelial type. In one of these cases, the demarcated and cystic tumour in the posterior cranial fossa lay over the dorsum of the cerebellum and invaded the medial surface of the right occipital lobe, tentorium, right lateral venous sinus, occipital bone, and scalp. The tumour consisted of dedifferentiated neurilemmoma, meningioma, rhabdomyosarcoma, and astrocytoma. In another instance, a circumscribed neoplasm in the right frontoparietal region contained dedifferentiated meningioma, ependymoma, and astrocytoma. Siqueira and Bucy (1966) also reported a mixed chondroma, ependymoma, and astrocytoma arising in the brain stem and filling the fourth ventricle. The patient described by Goldman (1969) had a mixed fibrosarcoma, rhabdomyosarcoma, and glioblastoma multiforme in the cerebrum. Since the tumour of our patient possessed a papillary pattern of cellular arrangement, it thus might be a mixed rhabdomyosarcoma and ependymoma. However, the presence of connective tissue cores within the papillae, as well as the brightly acidophilic cytoplasm of some tumour cells comprising the papillae, exclude a component of papillary ependymoma in the lesion. A papillary pattern of cellular arrangement was also observed in an alveolar rhabdomyosarcoma occurring outside the neuraxis reported by Enzinger and Shiraki (1969).

Leedham (1972), in reviewing 16 'possible rhabdomyosarcomas' of the neuraxis, noted that six of them were diagnosed as 'medulloblastoma' (Marinesco and Goldstein, 1933; Zülch, 1941; Bofin and Ebels, 1963; Boellaard, 1964; Gullotta, 1967; Russell and Rubinstein, 1971). These tumours arose in the cerebellar vermis, the usual site of medulloblastoma, but contained neuroblasts and striated muscle. They were therefore regarded as variants of medulloblastoma. We suggest that these neoplasms are combined rhabdomyosarcoma and medulloblastoma, and are within the category of tumours of mixed mesenchymal and neuroepithelial origin (Shuangshoti and Netsky, 1971; Shuangshoti and Panyathanya, 1974). Although Rubinstein (1972) has considered such a tumour to be teratoid because of its midline position, we do not agree that a midline neoplasm is necessarily teratoid.

The pathogenesis of primary intracranial rhabdomyosarcoma is likely to be related to aberrant differentiation of the unstable mesenchymal tissue towards striated muscle. Willis (1967) emphasized that rhabdomyosarcomas are rarer in organs containing large amounts of striated muscle than in locations where normally no or only scanty skeletal muscle is present, such as the urinary bladder, prostate gland, vaginal wall, spermatic cord, middle ear, and palate. Willis (1967) thought that rhabdomyosarcomas do not arise from mature striated muscle cells, but are derived from mesenchymal tissue possessing the potency for aberrant differentiation of striated muscle fibres. It is suggested that the present rhabdomyosarcoma arose from intracranial mesenchymal tissue as in the meninges. The dural involvement of the present tumour supports our contention.

We are thankful to Dr. Somsak Dhechakaisaya, Professor and Head of the Department of Pathology, Chulalongkorn University, for permission to report this case.

REFERENCES

- Bailey, O. T., and Ingraham, F. D. (1945). Intracranial fibrosarcomas of the dura mater in children: pathological characteristics and surgical management. *Journal of Neurosurgery*, **2**, 1-15.

- Berger, W. (1928). Über einen eigenartigen Tumor (lipoplastisches Sarkom) der Schädelbasis mit Einbruch in das Gehörorgan. *Beiträge zur Anatomie, Physiologie, Pathologie und Therapie des Ohres, der Nase und des Halses*, **27**, 301–324.
- Boellaard, J. W. (1964). Eine Medulloblastom mit quergestreiften Muskelfasern. *Archiv für Psychiatrie und Nervenkrankheiten*, **206**, 228–236.
- Bofin, P. J., and Ebels, E. (1963). A case of medulloblastoma. *Acta Neuropathologica*, **2**, 309–311.
- Butler, A. B., and Netsky, M. G. (1973). Classification and biology of brain tumors. In *Neurological Surgery*, vol. 3, pp. 1297–1339. Edited by J. R. Youmans. Saunders: Philadelphia.
- Enzinger, F. M., and Shiraki, M. (1969). Alveolar rhabdomyosarcoma: an analysis of 110 cases. *Cancer*, **24**, 18–31.
- Goldman, R. L. (1969). Gliomyosarcoma of cerebrum. *American Journal of Clinical Pathology*, **52**, 741–744.
- Gullotta, F. (1967). *Das sogenannte Medulloblastoma*. Springer: Berlin.
- Kernohan, J. W., and Uihlein, A. (1962). *Sarcomas of the Brain*, pp. 1–192. Thomas: Springfield, Ill.
- Kothandaram, P. (1970). Dural liposarcoma associated with subdural hematoma: case report. *Journal of Neurosurgery*, **33**, 85–87.
- Leedham, P. W. (1972). Primary cerebral rhabdomyosarcoma and the problem of medulloblastoma. *Journal of Neurology, Neurosurgery, and Psychiatry*, **35**, 551–559.
- Marinesco, G., and Goldstein, M. (1933). Sur une forme anatomique, non encore décrite, de médulloblastome; médullo-myoblastome. *Annales d'Anatomie Pathologique*, **10**, 513–525.
- Raskind, R., and Grant, S. (1966). Primary mesenchymal chondrosarcoma of the cerebrum: report of a case. *Journal of Neurosurgery*, **24**, 676–678.
- Rubinstein, L. J. (1972). Tumours of the central nervous system. In *Atlas of Tumor Pathology*, 2nd ser., fasc. 6, pp. 191–204 and, 215–234. Armed Forces Institute of Pathology: Washington, D.C.
- Russell, D. S., and Rubinstein, L. J. (1971). *Pathology of Tumours of the Nervous System*, 3rd edn, p. 15. Arnold: London.
- Shuangshoti, S. (1973). Neoplasm of mixed mesenchymal and neuroepithelial origin: liposarcomatous meningioma combined with gliomas. *Journal of Neurology, Neurosurgery, and Psychiatry*, **36**, 377–382.
- Shuangshoti, S., Hongsaprabhas, C., and Netsky, M. G. (1970). Metastasizing meningioma. *Cancer*, **26**, 832–841.
- Shuangshoti, S., and Netsky, M. G. (1971). Neoplasms of mixed mesenchymal and neuroepithelial origin: relation to 'monstrocellular sarcoma' or 'giant-celled glioblastoma'. *Journal of Neuropathology and Experimental Neurology*, **30**, 290–309.
- Shuangshoti, S., Netsky, M. G., and Jane, J. A. (1971). Neoplasms of mixed mesenchymal and neuroepithelial type: with consideration of the relationship between meningioma and neurilemmoma. *Journal of Neurological Sciences*, **14**, 277–291.
- Shuangshoti, S., and Panyathanya, R. (1974). Neural neoplasms in Thailand: a study of 2,897 cases. *Neurology (Minneapolis)*, **24**, 1127–1134.
- Shuangshoti, S., Piyaratn, P., and Viriyapanich, P. L. (1968). Primary rhabdomyosarcoma of cerebellum: necropsy report. *Cancer*, **22**, 367–371.
- Siqueira, E. B., and Bucy, P. C. (1966). Chondroma arising within a mixed glioma: case report. *Journal of Neuropathology and Experimental Neurology*, **25**, 667–673.
- Willis, R. A. (1967). *Pathology of Tumours*, 4th edn, pp. 757–771. Butterworths: London.
- Zimmerman, H. M., Netsky, M. G., and Davidoff, L. M. (1956). *Atlas of Tumors of the Nervous System*, pp. 103–107. Lea and Febiger: Philadelphia.
- Zülch, K. J. (1941). Ein Medulloblastom mit glatten Muskelfasern. *Archiv für Psychiatrie und Nervenkrankheiten*, **114**, 349–352.
- Zülch, K. J. (1965). *Brain Tumors. Their Biology and Pathology*, 2nd edn, pp. 219–225. Springer: New York.