Brit. J. Ophthal. (1975) **59**, 64

Neurilemmoma of the oculomotor nerve

SAMRUAY SHUANGSHOTI
From the Department of Pathology, Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand

At the time of writing, there are 51 intracranial neurilemmomas in our collection of 1,028 intracranial neoplasms; the only one of these which arose from the oculomotor nerve is reported here.

Case report

A Thai woman aged 64 years first experienced sudden severe headache and generalized clonic convulsion of all extremities, leading to coma for 4 hours, about 6 months before her death. After spontaneously regaining consciousness, she had a right hemiparesis. The patient was brought to a hospital where she was found to have hypertension. She was discharged a week later from that hospital without improvement of the hemiparesis.

3½ months later, the patient was admitted to another hospital because of right hemiparesis, intermittent headaches, and diminished vision in the left eye. After symptomatic treatment in this hospital for 6 weeks, the right-sided weakness gradually improved and it had disappeared at the time of discharge, but there was persistent headache and dimness of vision in the left eye. Subsequently, the patient was admitted to Chulalongkorn Hospital for further investigations of the latter symptoms.

Examination

The body temperature was 36·8°C; pulse, 70; respiration, 22; blood pressure, 200/110 mm. Hg. There was mild ptosis of the left upper eyelid. The left pupil, 7 mm. in diameter, was non-reactive to light. The left fundus could not be seen because of a senile cataract. In addition, there was limitation of the inwards rotation of the left eyeball. The right eye was normal. Other neurological findings were intact.

Routine studies of the blood and urine, and determinations of the blood sugar, cholesterol, and urea nitrogen revealed normal findings, as did radiological examination of the skull and chest. However, the left carotid and right retrograde brachial angiograms gave evidence of an aneurysm of the terminal portion of the left internal carotid artery. The clinical impression was paresis of the left oculomotor nerve secondary to the pressure effect of the aneurysm.

Operation

A left fronto-temporal craniotomy was performed on the tenth day in hospital. Two metallic clips were applied on a structure considered to be the neck of the aneurysm.

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

Course

The patient recovered uneventfully, but there was no improvement in the impaired functions of the left eye.

Termination

On the eighth postoperative day the patient suddenly became unconscious and died.

AUTOPSY

The general autopsy findings (Chula. Hosp. A-7782) included severe arterial and arteriolar sclerosis, involving especially the aorta, coronary arteries, pancreas, adrenal capsules, and kidneys; cardiac hypertrophy; chronic passive congestion of the lungs, liver, and spleen; and abundant mucus in the trachea and bronchi.

The brain, weighing 1,110 g., showed severe atherosclerosis of the arteries forming the circle of Willis. A fusiform aneurysm, 1·0 cm. in length and 5 mm. in diameter, was noted at the terminal part of the considerably sclerotic left internal carotid artery. Two metallic clips had been placed on this artery just below the proximal end of the aneurysm. The latter did not impinge upon the left third cranial nerve. The nerve, however, contained a cystic mass, 5 mm. in diameter, at a point 1·5 cm. from its point of exit from the mid-brain. Coronal sections of the brain disclosed no abnormalities.

Microscopically the mass of the third cranial nerve was composed of both dense and loose parts (Figure). The dense portion consisted of many spindle-shape cells arranging mainly in streams and intertwined bands. Occasional nuclear palisades, and a few cellular balls considered as Verocay's bodies were present. The loose part was composed chiefly of connective tissue fibres, and sparse haemosiderin-laden macrophages. Only a few spindle-shaped cells were seen in the loose zone.

Comment

The diagnosis was neurilemmoma of the left oculomotor nerve, an extremely rare type of tumour to arise in this nerve.

Zülch (1965) stated that this tumour might occur in the third cranial nerve, but he did not give an example. A standard textbook on the tumours of

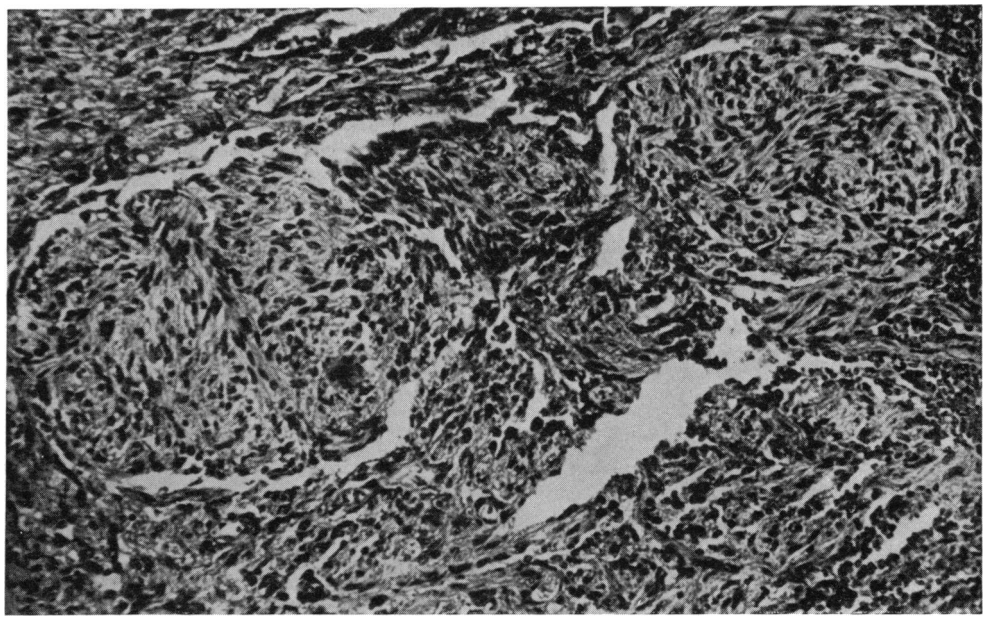

FIGURE *Verocay's bodies consisting of tangled masses of spindle-shaped tumour cells surrounded by connective tissue fibres. Haematoxylin and eosin.* × *100*

the nervous system (Russell and Rubinstein, 1963) did not describe neurilemmoma of the oculomotor nerve.

Discussion

After evaluating the clinical history and the *post mortem* findings, it is concluded that various cerebral symptoms, *viz.* headache, convulsion, coma, and right hemiparesis are features of hypertensive encephalopathy (Peart, 1971). The generalized atherosclerosis is considered as related to the effect of hypertension. The fusiform aneurysm is common in the patient having severe atherosclerosis; indeed, it is described by some authors as atheromatous aneurysm (Brain and Walton, 1969). Aneurysms of the terminal part of the internal carotid and posterior communicating arteries are known to be a frequent cause of oculomotor paresis because of pressure effect upon the nerve; the aneurysm may cause pupillary dilatation before other signs appear (Van Allen, 1969). In the present patient, however, both structures are widely separate; even in the living state, it is less likely that the pulsating small aneurysm will compress the nerve. The ocular and pupillary impairment on the left side, therefore, is considered to be associated with the neurilemmoma of the left oculomotor nerve alone.

The mechanism whereby paresis of the left third cranial nerve occurred in this patient may be understood by reviewing the innervation of the eye. The oculomotor nerve innervates the levator palpebrae, the medial rectus, the superior and inferior rectus, and the inferior oblique muscles. Each oculomotor nucleus is located in the central grey matter of the mesencephalon in the region ventral to the upper part of the aqueduct. Its intramedullary fibres pass forwards and ventrally through the red nucleus and medial portion of the substantia nigra, and emerge from the midbrain through the sulcus on the medial side of the cerebral peduncle. The extramedullary portion of the oculomotor nerve passes anteriorly through the lateral wall of the cavernous sinus, and enters the orbit through the superior orbital fissure to innervate the aforesaid ocular muscles. The oculomotor nerve, furthermore, conveys preganglionic parasympathetic fibres from the Edinger-Westphal nucleus located in the area adjacent to the oculomotor nucleus. After synapsing in the ciliary ganglion, postganglionic parasympathetic fibres arise which pass by way of the short ciliary nerve to the circular muscle of the iris (Davies and Coupland, 1972).

It is suggested that the pupillary dilatation and loss of light reflex of the left eye in the present case are related to interruption of the preganglionic parasympathetic fibres within the left third cranial nerve by the incipient neurilemmoma. The sympathetic fibres, originating in the intermediolateral column of the lower cervical and the upper thoracic segments of the spinal cord and subsequently innervating the radial muscle of the iris (Buchanan, 1957; Van Allen, 1969), are intact; hence, the pupil is

dilated. Ptosis and limitation of inwards rotation of the left eyeball is associated with interruption of the nerve fibres supplying the respective levator palpebrae superioris and medial rectus muscle.

Summary

A case is presented of neurilemmoma of the left oculomotor nerve occurring in a 64-year-old hypertensive woman. The incipient tumour produced ptosis, limited inwards rotation of the eyeball, and persistent pupillary dilatation on the left side. The mechanism of this process is discussed, having regard to the interruption of the fibres innervating these structures by the tumour, and considering the mode of occurrence of the ocular and pupillary impairments. The patient also had generalized atherosclerosis and fusiform aneurysm of the terminal part of the left internal carotid artery. The symptoms of headache, seizure, coma, and temporary right hemiparesis were considered as manifestations of hypertensive encephalopathy unrelated to the tumour.

This work was supported by Rachadapiseksompoj Research Grant of Chulalongkorn University on the Royal Silver Jubilee Anniversary. The author is grateful to Prof. Somsak Dhechakaisaya, Head of the Department of Pathology, for permission to study this case.

References

BRAIN, R., and WALTON, J. N. (1969) "Brain's Diseases of the Nervous System", 7th ed., p. 314. Oxford University Press, London

BUCHANAN, A. R. (1957) "Functional Neuro-anatomy: Including an Atlas of the Brain Stem", 4th ed., pp. 197–200. Lea and Febiger, Philadelphia

DAVIES, D. V., and COUPLAND, R. E. (eds.) (1972) "Gray's Anatomy: Descriptive and Applied", 34th ed., pp. 1139–1141. Longmans, London

PEART, W. S. (1971) "Arterial hypertension", *in* "Cecil-Loeb Textbook of Medicine", ed. P. B. Beeson and W. McDermott, 13th ed., pp. 1050–1062. Saunders, Philadelphia

RUSSELL, D. S. and RUBINSTEIN, L. J. (1963) "Pathology of Tumours of the Nervous System", 2nd ed., p. 244. Arnold, London

VAN ALLEN, M. W. (1969) "Pictorial Manual of Neurologic Tests", pp. 86–96. Year Book Medical Publishers, Chicago

ZÜLCH, K. J. (1965) "Brain Tumors: Their Biology and Pathology", 2nd ed., p. 193. Springer, Berlin