Primary choroid papillomas in the cerebello-pontine angle

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In the great majority of cases choroid papillomas develop inside the cerebral ventricles whereas extraventricular growths of this kind are almost always either a direct extension of a primary intraventricular papilloma or are due to seeding along the cerebrospinal fluid pathways. These tumours may, however, occur primarily outside the ventricles, though very seldom.

There are very few cases of primary papilloma unconnected either with the choroid plexus or with the ventricles, examples being the case reported by Skala (1956), in which the tumour was inside the right frontal lobe, and the three reported respectively by Greene (1951), by Robinson (1955), and by Kahn and Luros (1952), in all of which the tumour was inside a cerebellar hemisphere. For these exceptional occurrences it is reasonable to assume, in Greene's opinion, that the tumour developed from an ectopic remnant of choroid tissue.

There are other extraventricular papillomas that are neither seedings nor outgrowths of an intraventricular tumour and, as they develop from a normal choroid structure, they cannot be regarded as truly ectopic either. These are the papillomas that develop from the small choroid tuft which normally projects outside the foramina of Luschka into the cerebello-pontine angle. These primary extraventricular papillomas sometimes have an outgrowth inside the fourth ventricle also. Hence they have, or may have, points in common with the papillomas that actually originate in the fourth ventricle and then develop, sometimes predominantly, in the lateral recess and eventually, through the foramen of Luschka, they expand outside the ventricle and spread around the cerebellum and bulb. This is not an uncommon occurrence, as may be seen from many reports, the most recent being that of Bohm and Strang (1961). What distinguishes these two topographic varieties clinically is that in papillomas with an intraventricular origin, which early on in the course of the disease occlude the cerebrospinal fluid pathways, intracranial hypertension is generally the first symptom, whereas in the case of solely or mainly extraventricular papilloma the first or pre-

dominant signs are those of lesions of the nervous structure of the angle and thus they simulate the clinical features of the tumours most commonly found in this site.

Having had occasion to observe two cases of these primary papillomas of the angle, as a matter of curiosity we searched the literature for accounts of similar cases published to date and found that in some of the few existing accounts there are observations on the symptoms or clinical and radiological signs that are in conflict with other observations. We felt therefore that it might be worthwhile to reconsider this particular aspect of intracranial pathology right from the beginning, bearing in mind our own observations and those of other workers.

REVIEW OF THE LITERATURE

Cushing (1917) was the first to report a case of primary papilloma of the cerebello-pontine angle in his monograph on tumours of the acoustic nerve. The history of the patient (case 35 in the monograph) covered several years and so cannot be summarized briefly. It is therefore quoted here in some detail.

When the patient was examined for the first time at the age of 22 he had been suffering for four months from pain in the right temple on stooping, for six weeks from clouding of vision and for one month from dizzy spells, diplopia, and some subjective disturbances of taste; for some time he had had persistent suboccipital headaches. The clinical findings were: bilateral choked disc of 3 diopters with haemorrhages and exudates; slight nystagmus on looking to the left; a positive Romberg test with falling to the left; nothing in the trigeminal; definite weakness of the left abducens with diplopia; definite left facial weakness; no obvious lesion of the acoustic nerve. On a diagnosis of subtentorial tumour, presumed to be cerebello-pontine, the patient was operated on for the first time on 17 July 1908 for a palliative left subtemporal decompression. The diplopia disappeared but the patient was readmitted three months later owing to incomplete subsidence of the choked disc and suboccipital discomfort. At clinical examination there was no change. At the second operation (8 October 1908) the posterior fossa was explored and a lateral cistern was found to be

dilated. The patient was operated on a third time 13 months later owing to aggravation of the cerebellar symptoms (onset of dysmetria and hypodiadokinesia of the left arm): the left cerebello-pontine angle was explored again and a multilobular arachnoid cyst was evacuated. A year later the patient was readmitted and the findings were: positive Romberg test, some degree of incoordination of left limbs; no choked disc; left corneal areflexia; slight left facial weakness; no tinnitus or deafness; labyrinth tests normal. The patient underwent a fourth operation and at exploration of the angle another arachnoid cyst was found and evacuated. A year later there was increased ataxia and unsteadiness; the left facial deficit was as before; left corneal areflexia; occasional diplopia; loss of taste on the left half of the tongue; recurrence of papilloedema; tinnitus of recent onset. The patient was operated on for the fifth time on 24 October 1911, that is three years seven months after the onset of the original symptoms. On opening the posterior fossa, the left cerebellar hemisphere was incised and at a depth of 1 cm. a circumscribed roundish nodule was found (6 × 5 × 5 cm.). It was almost completely removed, only a small fragment of the growth being left in the angle. The patient died six days later from bronchopneumonia.

We have traced eight other cases, which are summarized in Table I. Two of them, Tola's (1951) and

Willis's (1953), were reported very briefly. In the case of Kernohan, Woltman, and Adson (1948) the diagnosis was papillary ependymoma but it is arguable that it was a case of choroid papillomas. Indeed, apart from the fact that Kernohan classified choroid papillomas as ependymomas, in the case in question, a photomicrograph of which is given in Kernohan and Sayre's atlas (1952) (Fig. 45), Zülch (1956) suggested that it was a choroid papilloma. This case is included in the Table but it will not be considered in the discussion of the symptoms of papillomas of the angle, because of the doubts as to its histological classification and because the period of observation was too short for a thorough clinical study.

While on the subject of histological diagnosis, it must be pointed out that choroid papillomas must be distinguished from a particular variety of ependymoma which, according to Zülch and Kleinsasser (1957), develops only in the cerebello-pontine angle and whose general texture closely resembles that of papillomas. This kind of ependymoma has a trabecular-papillary structure and in the midst of the cellular strands there are here and there capillary vessels with an epithelial investment, in the main

TABLE I

SUMMARY OF CASES FROM THE LITERATURE					
Author	Sex and Age	First Symptom (1)	Following Symptoms (2)	Interval between 1 and 2	Duration of case before death
Cushing (1917)	M. 22	Headache	See text	10 wk.	10 mth.
Devic and Puig (1927)	F. 22	Headache, blurring of vision	Vomiting, visual impairment, tinnitus, unsteady gait	6 mth.	10 mth.
Guillain <i>et al.</i> (1932)	F. 47	R. hearing impaired	Diplopia, headache, vomiting, R. lateropulsion	14 mth.	2 yr.
Montanaro and Hanón (1935)	F. 29	R. hemicrania and vomiting	Dizziness, R. tinnitus: weak R. leg	1 wk.	1 mth.
Kernohan <i>et al.</i> (1948)	F. 59	Awkwardness of hands, syncope	Ataxia and speech disturbance	1 yr.?	1 yr.
Tola (1951)	F. 22	L. deafness immediately following parotitis		4 yr.?	6 yr.
Kahn and Luros (1952)	M. 36	R. deafness	Dysarthria, dysphagia, ataxia, blurring of vision	4 yr. and 6 mth.	5 yr.
Willis (1953)	M. 57	Not reported	Weakness of limbs, ataxia, falling to R.		2 yr.
Polizzi (1955)	M. 20	Headache, diplopia, vomiting	R. tinnitus, complete blindness	1 yr.	2 yr.
Morello and Migliavacca (case 1)	F. 35	L. hearing impaired	Deviation to L. and headache	20 mth.	2 yr.
Morello and Migliavacca (case 2)	F. 31	R. hearing impaired and paracusis	Headache, dizziness, vomiting, deviation to R. in walking, neck stiffness	9 yr.	10 yr. 21 mth.

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monostratified. What distinguishes these ependy-momas from papillomas is, according to Zülch and Kleinsasser (1957), that in papilloma two papillae never unite transversally whereas in ependymoma there are slender trabeculae which merge together in twos and threes and so circumscribe cavities. Further, the connective tissue of the papillar stroma is looser and more scanty and the epithelium is taller than in papilloma.

Photomicrographs of six of the published cases (those of Cushing (1917); of Tola (1951); of Willis (1953); of Polizzi (1955); of Guillain, Petit-Dutaillis, Bertrand, and Lereboullet (1932); and of Montanaro and Hanón (1935)) show that the diagnosis of papilloma was correct. In two cases (Devic and Puig, 1927; Kahn and Luros 1952) there is no photo-micrographic documentation and in the case of Kernohan *et al.* (1948), as stated earlier, the interpretation is open to question.

PERSONAL CASES

Our two cases represent 1·2% of the primary tumours of the cerebello-pontine angle operated on at the Istituto Neurologico di Milano between September

1946 and 31 December 1962 (145 acoustic neuromas, nine meningiomas, three epidermoids, and one chondroma). Seven cases of choroid papilloma (five of the fourth ventricle and two of the lateral ventricles) in addition to the two under review were operated on during this period.

CASE 1 L.A. (ser. no. 20720/1955), a woman aged 35, about two years before admission to hospital noted transient deafness and left paracusis after diving into the sea. There was subsequently some transient recurrence of these disturbances. Four months before her admission the deafness became persistent, and posture and gait became unsteady. One month before admission she began to swerve to the left when walking, an intention tremor of the left arm appeared, and finally fairly frequent headache.

Clinical findings Ocular fundi were normal. Left trigeminal hypaesthesia; corneo-conjunctival reflex weaker on left; mild left facial paresis. Fitful nystagmus, clockwise in left lateral deviation, anticlockwise in right lateral and upward deviation of gaze. Severe left hypacusis. Labyrinthine reflexes bilaterally weak. Unsteady standing with tendency to fall to the left; leftward swerve when walking. Dysmetria and hypotonia of the left limbs. Left tendon reflexes slightly flabby.

Radiography of the skull Nothing abnormal; left inner acoustic foramen normal. As the clinical signs clearly

TABLE I—continued
SUMMARY OF CASES FROM THE LITERATURE

Clinical Signs	X-ray Studies	Operation	Outcome	Post-mortem Findings and Other Remarks
lema, severe visual ent	Not performed Not performed	See text Fronto-parietal decompression	Death after operation Death after operation	Vegetating tumour in R. cerebello- pontine angle; not in fourth ventricle
lema. R. 5, 7 and 8 cranial ystagmus, ataxia	Not performed	Walnut-sized tumour in right cerebello-pontine angle removed	Cure	
ema. L. 5, 6, 7, and 8 rves, ataxia	Not performed	Not performed	Death following lumbar puncture	Walnut-sized tumour in lateral recess and fourth ventricle, com- pressing middle cerebellar ped- uncle, pons, medulla
sion; coma, papill- xanthochromic C.S.F., lity	Not performed	Not performed	Death after admission	Hydrocephalus: fungating tumour compressing medulla, pons, and cerebellum. Not in fourth ventricle
ulated acoustic a	Not reported	Removal of tumour in L. cerebello-pontine angle	Immediate post- operative death	Arising from tuft in foramen of Luschka
ema. R. 5, 7 and 10 rves	Irregular destruction of R. petrous ridge	Partial removal of haemor- rhagic tumour in R. cere- bello-pontine angle	Cure	Patient never again able to work. Death 2 yr. after operation from unknown cause
is, R. trigeminal sia, R. facial paresis	Not reported	Partial removal of large, hard but friable vascularized tumour in R. cerebello- pontine angle	Death on sixth post- operative day	Large residual growth in R. cere- bello-pontine angle, impinging on dorsal surface of pons; partial ex- tension into fourth ventricle
illoedema R. 5, 6, 7 rves; nystagmus	Anterior displacement of aqueduct	Partial removal growth underlying R. cerebellar hemisphere	Death on second post- operative day	Tumour 4 cm. diameter adherent to arteries; pons and medulla pushed to left. Papilloma in fourth and third ventricle
d 8 cranial nerves,	Plain radiographs normal	Removal of egg-sized encapsulated tumour	Cure	5. yrs. later slight L. cerebellar signs, improvement of hearing
ma, neck stiffness, s, ataxia	Enlarged R. internal ac. meatus (see text)	Total removal of apricot- sized tumour	Cure	4 yr. later: patient well except slight awkwardness of R. hand

indicated a lesion of the left cerebello-pontine angle, the patient was operated on without any further radiological investigation.

Operation (11 January 1955) Intratracheal general anaesthesia was maintained with N_2O and ether. A suboccipital flap was made. In the left cerebello-pontine angle a well-encapsulated, pale, straw-coloured tumour, the size of a hen's egg, was found and completely removed. It was not attached to the dura and had no outgrowths in the porus acusticus internus. The acoustic and facial nerve could thus be spared. The trigeminal hypaesthesia swiftly disappeared but cerebellar signs subsided only slowly and incompletely. At follow-up five years later there was still some unsteadiness of gait and lack of precision in the left hand movements; recovery of hearing was fair.

Histology The tumour consisted of an assemblage of ramified papillae which did not anastomose with one another. They were made up of slim vascular connective tissue axes covered with monostratified columnar epithelium. No mitoses (Fig. 1) were visible. Microscopical diagnosis was choroid papilloma.

CASE 2 C.M.T. (ser. no. 28376/1959), a woman aged 31, from the age of 9-10 years experienced hypacusis and at times paracusis on the right side. At 19 years worsening of right hypacusis was followed by the onset of almost continuous headache, dizziness, vomiting, lateropulsion and stiff neck. Admitted to another hospital in 1948, she was operated on for a walnut-sized cystic tumour in the right cerebello-pontine angle attached by means of a small pedicle to the dura at its insertion on the petrous bone. The tumour was apparently completely removed. Histological examination showed that it was a papilloma with a monostratified epithelial investment. After an uneventful

FIG. 1. Photomicrograph of sections of the tumour in case 1. No mitoses can be seen. Haematoxylin and eosin $\times 110$.

post-operative course the patient kept fit, apart from a cranial injury in 1952, until 1958 when the dizzy spells and headache returned and, within a month, vomiting, unsteadiness on her feet, and lateropulsion.

Clinical findings on admission (14 February 1959) The occipital flap was bulging. Tendency to hold the head bowed to the right. Bilateral papilloedema, more marked on left. Eye movements normal; no nystagmus. Trigeminal and facial nerves unaffected. Deafness on right side; vestibular block on right. Right deviation when walking. Coordination tests less precise on right side. Tendon reflexes normal on both sides.

Radiology of the skull Suboccipital craniectomy; decalcification of the dorsum sellae and of the floor of the right posterior fossa, enlargement of the right porus acusticus whose inferior margin was eroded.

Vertebral arteriography was attempted but was unsuccessful. **Right carotidography** disclosed a confluence of arteries in the right cerebello-pontine angle such as to form a pathological circulation, which persisted in the venous phase.

Operation (26 February 1959) Intratracheal anaesthesia was maintained with N_2O and ether. On raising the old suboccipital flap, on the right side, under a thin mantle of cerebellar tissue, was a cyst containing about 20 ml. of brownish fluid. At its base there was an apricot-sized, well-encapsulated, brownish tumour of tough elastic consistency. The tumour was isolated from the cerebellar tissue and then completely removed together with its richly vascularized dural insertion lying on the postero-inferior surface of the petrous bone. The bone was likewise richly vascularized, and electrocoagulation and bone wax were necessary to ensure haemostasis. The post-operative course was disturbed by meningitis, which was treated with antibiotics, but in the end the patient was cured. Quite recently (1963) she wrote to say that she felt well, was leading a normal life, and had married. There is still some unsteadiness of the right hand, especially when writing.

Microscopical examination¹ Tumour with a fairly regular papillar structure. Each papilla had a blood vessel running through it and a fairly substantial framework of connective tissue, which here and there was oedematous or hyalinized. The epithelial investment was monostratified and consisted of columnar or cuboidal cells. There were no mitoses (Fig. 2). Histological diagnosis was choroid papilloma.

COMMENT

In connexion with his case, which he saw for the first time in 1908, Cushing wrote in 1917: 'With our present understanding of this latter condition (acoustic fibroneuroma), in the absence of any significant auditory disturbances, the case, despite the involvement of the Vth, VIth and VIIth nerves and the slow progress of the symptoms after the

¹ The peculiar features of this tumour, especially its insertion on the petrous bone, left us uncertain as to the interpretation of the preparations and we are grateful to Professor K. J. Zülch (Köln-Merheim, Germany) for the diagnosis.

FIG. 2. Photomicrograph of sections of the tumour in case 2. As in Fig. 1, no mitoses can be seen. Haematoxylin and eosin $\times 110$.

decompression, would have sufficed to show that we were dealing with a recess tumour of another sort.' This affirmation of Cushing's is wholly acceptable but it may be added that it applies equally well to other rare tumours of the cerebello-pontine angle, such as, for example, epidermoids.

According to Polizzi, papilloma of the angle differs from acoustic neuroma in the chronological order of onset of symptoms, hearing disturbances being slight or late in the former and appearing only when intracranial hypertension has already been manifest for some time. The history of his case accords with this statement but the only other similar case is that of Devic and Puig (1927). In the case of Montanaro and Hanón (1935) the development of the symptoms was swift, as may occur with a malignant tumour of the brain-stem. The account of Willis's (1953) case is too summary to permit of any conclusion. The case of Kernohan *et al.* (1948) is not considered here for the reasons already stated. The five remaining cases conflict with the opinion that in papillomas of the angle the signs of impairment of the acoustic nerve are slight or late. As a matter of fact, impaired hearing preceded all other signs by 14 months in the case of Guillain *et al.* (1932), by four and a half years in the case of Kahn and Luros (1952), by four years in Tola's (1951) case (deafness was immediately preceded by parotitis), by two and a half years in our first case, and by 10 years in the second. In the subsequent course of these five cases there was nothing to distinguish them from acoustic neuromas. To

sum up, on the strength of the experience so far collected it does not seem that there is any clinical feature on which to base a pre-operative diagnosis of choroid papilloma of the angle. On the contrary, this kind of tumour often mimics acoustic neuromas.

Polizzi (1955) held that radiology can supply a negative pointer or even decisive evidence. According to him, the absence of bone changes in the region of the porus acusticus internis is a negative pointer. This does not seem to be a criterion of much value since bone changes are absent in some 15 to 20% of acoustic neuromas according to several broad-based statistics. Further, in two cases of papilloma of the angle, in that of Kahn and Luros (1952) and in our second case, there were lesions of the petrous bone or actually of the porus. Polizzi regarded ventriculographic demonstration of a ragged shadow in the fourth ventricle, due to the presence of a portion of the papilloma within it, as decisive evidence. We do not think that much reliance can be placed on this radiological criterion because in only three cases (Willis, 1953; Montanaro and Hanón, 1935; and Polizzi, 1955) was there an outgrowth of the tumour in the fourth ventricle. In the other six cases collected from the literature there is either no mention of such an outgrowth or it is definitely excluded by the post-mortem findings (Devic and Puig 1927; Kernohan *et al.*, 1948). The fourth ventricle was not explored in either of our cases but it seems unlikely that there could have been any outgrowth of the papilloma inside it since, had there been, it would have given signs of its existence in the long period of time following the operation in both cases.

In conclusion, at the moment there does not seem to be any sign, either clinical or radiological, that permits pre-operative diagnosis of primary papillomas of the cerebello-pontine angle. All that may be said about these tumours is that, with the exception of the case of Montanaro and Hanón, they develop slowly. Their clinical signs and symptoms do not differ from the general signs of tumours of the angle and they may even mimic the features of acoustic neuromas, at times even radiologically.

SUMMARY

Cases of primary choroid papilloma in the cerebello-pontine angle collected from the literature are reported and two personal cases are added with some remarks on their clinical signs and symptoms and radiological findings.

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