

Clinical Aspects of the Commoner Intracranial Tumours in Children

BY

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Brain tumours in children are not uncommon. An average sized neurosurgical unit is supplied with a steady flow of suspected cases to be proved or disproved—some from the general practitioner direct, but the majority from a paediatrician who has been consulted in the first instance. Most practitioners, at least once in a professional lifetime, will encounter a case, and on their shoulders the responsibility for the recognition of the serious nature of the symptoms rests. It is with these early clinical manifestations that this paper is concerned. Once sus-

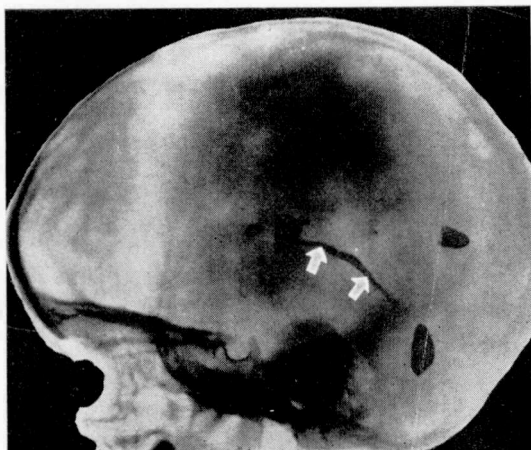


Fig. 1—Brainstem glioma (myodil ventriculogram). Dye outlines part of the third and fourth ventricles. The Aqueduct of Sylvius is elongated and pushed upwards and backwards by a large glioma of the pons (see arrows).

pected, verification is not generally a difficult task with the aid of modern diagnostic tools—electroencephalography, ventriculography with air and opaque media, angiography and diagnostic exploration (Figs. 1 and 2).

It seemed worthwhile to investigate whether any significant delay actually occurs in the recognition of these tumours and whether any lessons might be learnt from their modes of presentation. With this object in mind the case

histories of 50 pathologically proved cases of intracranial tumour, selected at random, were examined, particular note being taken of the clinical findings, history and eventual outcome.

It was soon evident that no such delay in diagnosis occurred, and that no correlation

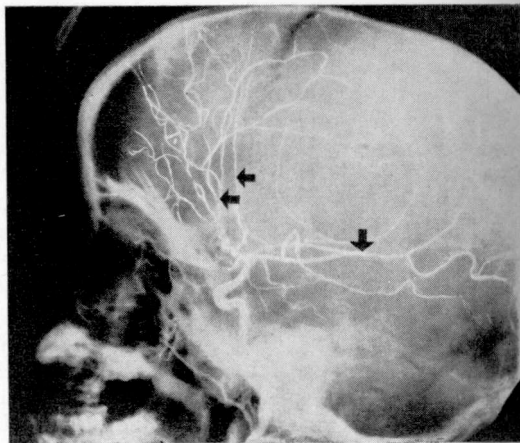


Fig. 2—Glioma of the left cerebral hemisphere. Percutaneous left common carotid angiogram. Arrows indicate the displaced branches of the anterior and middle cerebral arteries.

could be established between long delay before diagnosis and increased mortality or morbidity. The great majority of cases were "text book" in their presentation: the symptoms were urgent and obviously significant. Furthermore, a constantly recurring paradox was the massive size of the tumour as seen at operation or autopsy, compared with the shortness of the period in which clinical manifestations were evident. Yet the few errors which *were* noted were in those conditions which generally offer the best chances with effective surgery—the cerebellar and cerebral gliomata.

On evidence culled from this series the salient clinical features of the commoner intracranial tumours will be reviewed and illustrated by brief case reports. Some aspects of particular interest will be enlarged upon and finally important conditions causing confusion discussed.

GENERAL CONSIDERATIONS

Several characteristics govern the symptomatology of these tumours.

(1) The majority take origin from midline structures—the vermis cerebelli, fourth ventricle, brain stem and structure about the third ventricle. For this reason neurological signs are frequently absent until the growth is far advanced.

(2) Equally, the cerebral hemispheres in childhood have not yet attained full specificity of function, nor is dominance of one hemisphere over the other as obviously in evidence as it is in the adult. A young child, moreover, does not have the capacity nor the inclination to describe his sensations and disabilities. Tumours which are situated in these areas are consequently surprisingly silent.

(3) *The Syndrome of Increased Intracranial Pressure (ICP).*—Encroachment upon cerebrospinal fluid pathways usually marks the beginning of the clinical illness. Whether this is in the fourth ventricle, aqueduct of Sylvius, foramen of Munro or impaction at the tentorial opening, the same picture of raised intracranial pressure will result. Inevitably there is vomiting and headache, the vomiting constantly in the early morning on rising and often neither forceful nor facile; the headache frontal, universal or radiating down the neck, often worsened by stooping and often excruciating. Yet if the process is slow enough, sutures will separate and the skull expand, compensating to some extent for the increased volume of the dammed-up fluid, and delaying the onset of symptoms. Examination of the optic fundi will reveal engorged veins or papilloedema and there may be visual impairment. Kinking of the sixth cranial nerve results in squint and double vision. As the pressure increases, acute constriction occurs at the tentorial opening and the foramen magnum—medulla and cerebellar tonsils descending well down into the cervical canal. Neck stiffness and head retraction are frequent at this stage. Impairment of consciousness, decerebrate rigidity and disturbance of vital processes with sudden respiratory failure are terminal signs.

(4) Sudden aggravation of symptoms or worsening of condition may be referable to an increase in size of a cyst within the tumour, haemorrhage into or from its surface, surrounding oedema or some subtle alteration in dynamics. No matter how short the history, therefore, the possibility of a tumour should always be borne in mind when confronted with the syndrome of increased ICP. Though anaplastic tumours progress more quickly than mature ones, no reliance can be placed on the duration of symptoms in predicting the histology of the growth. This is often the subject of dispute between neuropathologists themselves.

(5) Epilepsy is not a common manifestation of tumours in childhood, whether above or below the tentorium.

Full neurological examination is obviously not possible in young children, yet a great deal can be learnt from unhurried observation of the child while walking and playing. A slight tilt of the head, lack of co-ordinative movement in an arm, and an inclination to look for support from fixed objects, a broad based gait or a stagger on turning sharply—all may be evident on close scrutiny.

The importance of clinical examination of the fundi cannot be stressed sufficiently. Of the 50 cases investigated, only six did not show papilloedema. These were all brain stem gliomata in which absence of raised ICP is a diagnostic feature. The pupils should be dilated where there is any difficulty.

It is worth stressing that careful head measurements may be of value. Repeating the readings after a few days may reveal increase in head size not otherwise apparent. In this regard the average head circumference at 12 months is 18.3 in., at two years 19.3 in. and at five years 20 in. (Nelson, 1954).

INFRATENTORIAL TUMOURS

Thirty-six of these tumours (72 per cent.) were situated in the restricted confines of the posterior fossa. The bulk were formed by the diffuse glioma of the brain stem (Fig. 3), the medulloblastoma, the ependymoma of the fourth ventricle and the cerebellar astrocytoma.

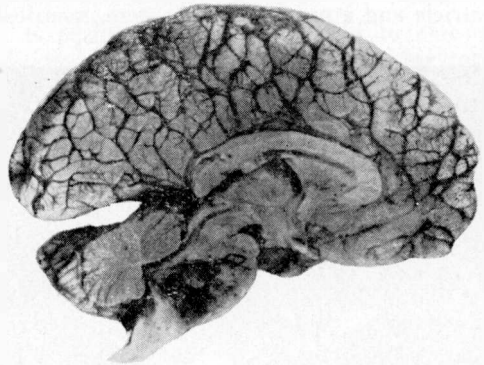


Fig. 3—Diffuse pontine glioma with haemorrhage. Autopsy specimen.

Diffuse Brain Stem Glioma.—This tumour is seen fairly frequently; there were six examples in this series (12 per cent.). Though often mature in histological appearances, it is unequivocally inoperable, diffusely infiltrating the pons, medulla or midbrain, and deep X-ray therapy is of doubtful value. Though symptoms

may be present for prolonged periods (one in this series had a facial palsy for five years), the average duration of symptoms before diagnosis is one to five months. It is extraordinary to compare the degree of involvement of the brain stem at autopsy with the shortness of the clinical duration of the illness. There is no increase in ICP as a rule and the fundi are therefore normal. Multiple cranial nerve involvement is characteristic—disturbance of lateral eye movements, facial weakness and absence of corneal reflex are signs worth stressing—and there are associated pyramidal signs and ataxia. Trauma often seems to initiate the symptoms.

Case Report.—Cynthia S., 2 years 11 months. Development normal until two years, when she gradually developed squint. For five months progressive unsteadiness on walking. For three months shakiness of right hand and dribbling out of the left side of the mouth. There was no vomiting, but occasional pain over the back of the head. On examination, very apathetic and markedly ataxic on standing, tending to fall forward. Fundi normal. Bilateral complete external rectus paralysis. Bilateral facial weakness. Both corneal reflexes absent. All deep reflexes exaggerated. Bilateral extensor plantar response. Myodil ventriculography revealed large pontine tumour pushing the aqueduct and fourth ventricle far backwards and upwards. Died one month later.

Medulloblastoma.—There were 15 of these tumours (30 per cent.). The medulloblastoma is by far the commonest growth in the toddler age-group (Figs. 4 and 5). Growing quickly, it destroys the vermis, fungates into the fourth ventricle and around the brain stem, manifesting

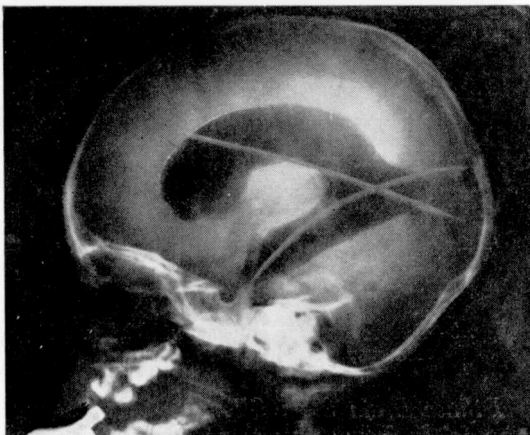


Fig. 4—Posterior fossa tumour (medulloblastoma). Soft rubber catheters have been inserted into the lateral ventricles and the fluid replaced with air. A diffusely dilated ventricular system is outlined. Note separation of skull sutures.

all the signs of a progressively rising ICP. Papilloedema is constant. Localising neurological signs are not characteristic until the disease is far advanced, when they may indicate spinal cord involvement or actual seeding of tumour to distant parts of the nervous system. Deep X-ray therapy offers a chance of lengthened survival, but the long-term results are usually disappointing. The following case is atypical in this respect.

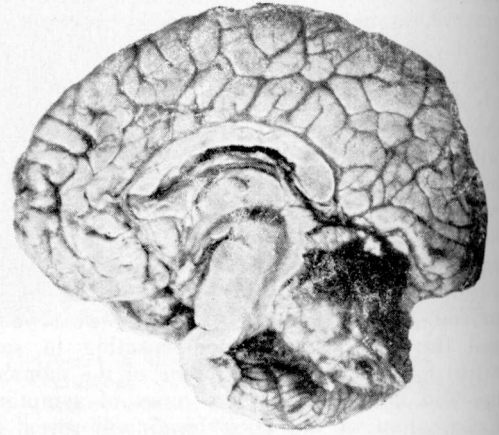


Fig. 5—Medulloblastoma. Note distortion and compression of medulla and pons with great enlargement of the fourth ventricle.

Case Report.—Frank G., 2 years 3 months. He was quite well until nine weeks before admission. Then began vomiting and complaining of headaches. Unsteady on his feet for six weeks and had not walked at all for two weeks. On examination, dehydrated, head circumference 19½ in. Bilateral papilloedema with haemorrhages. Right external rectus weakness. Bilateral extensor plantar response. Ventriculography revealed greatly increased ventricular pressure. Posterior fossa exploration—large fleshy midline tumour 3 in. in diameter removed *in toto* from the vermis and cavity of the fourth ventricle. Full course of deep X-ray therapy. Quite well after five years.

Ependymoma of the Fourth Ventricle.—This tumour is identical in its presentation to the medulloblastoma, but by and large the duration of symptoms before diagnosis is longer. It is less malignant than the latter, but unfortunately involvement of the floor of the fourth ventricle often makes complete removal impossible. There were two examples in the series (4 per cent.).

Cerebellar Astrocytoma.—This slowly growing tumour, commonest in the second half of the first decade, often spreads by infiltration and cyst formation from the midline into the body of the cerebellar hemisphere. To the signs of increased ICP, therefore, may be grafted those

of unilateral cerebellar dysfunction. Intervals of comparative freedom from symptoms may prolong the course of the disease to several years. In contrast to the medulloblastoma, successful surgical extirpations are frequent. There were 12 examples of this growth (24 per cent.).

Case Report.—Michael J., 6 years 2 months. For 18 months before admission felt uncertain of objects held in right hand; tended to drop things easily. More recently—actual tremor of right arm. For nine months staggering to the right on walking. For six months intermittent vomiting and headache first thing in the mornings. On examination, bilateral papilloedema. Coarse nystagmus on looking to left. Scanning speech. Positive finger-nose test and dysdiadochokinesis on right. Occipital burr-hole and air ventriculography revealed diffuse enlargement of lateral ventricles. Apparently total removal of cystic astrocytoma of vermis and right cerebellar hemisphere. After six years, no symptoms. Bats well at cricket, but bowling is weak! Minimal ataxia of right hand. Decompression slack.

SUPRATENTORIAL TUMOURS

Glioma of the Cerebral Hemispheres.—Commencing in the substance of the hemisphere, be it white matter, nuclei or ependyma of ventricle, these tumours are free to spread widely before they produce blockage or encroach on areas where their presence becomes evident. Hence they often reach enormous proportions. Listlessness or an alteration in disposition are often in retrospect the initial symptoms, but the picture is eventually coloured by those of increasing ICP. A varying degree of hemiparesis is frequent, but epilepsy, focal or general, is not seen as often as it is in similar tumours in adults (Ingraham and Matson). Of the six cases in this series (12 per cent.), only one had convulsions. One of these cases proved to be a meningioma—a rare tumour in children. All degrees of malignancy are seen.

Case Report.—Sandra McM., 10 years. Four months previously had slipped in the snow and bumped the back of her head. After this she was irritable and complained frequently of headaches. Was often sick in the morning. Occasional double vision. On examination, intelligence normal. Bilateral papilloedema. Some weakness of right external rectus. Doubtful slight weakness of right lower face. No other clinical abnormalities. Skull X-rays calcification in left frontal region. Huge tumour covering whole convexity of frontal lobe, bulging into lateral ventricle and surfacing on medial aspect of frontal cortex. Microscopy—glioblastoma. Apparent complete removal followed by radiotherapy. Died after 18 months.

Parahypophysial Tumours.—Growths around the pituitary are neither common in childhood nor characteristic in their presentation. The craniopharyngeoma and its classical triad of visual field defects, endocrine disturbance and X-ray changes in the skull, is a tumour of the second or third decade.

There were four tumours in this vicinity in the present series: a glioma of the chiasma, a spongioblastoma of the third ventricle, a glioma of the hypothalamus, and a pinealoma. In all these cases the picture was one of increasing ICP with a varying degree of signs appropriate to their position; for example, in the chiasmatic growth progressive blindness developed, and in the pinealoma there was inability to look upwards.

The occurrence of two cases of sarcomatosis of the meninges in this series is a great coincidence. It is a rare condition and will not be discussed here.

The remaining cases were (1) a choroidal papilloma; (2) a cerebellar dermoid; and (3) a cerebral angioma (see below).

DISCUSSION

Of the tumours described, only the cerebellar astrocytoma and the more differentiated of the cerebral growths offer a good chance of permanent survival after complete excision. The place for radiotherapy in the treatment of the less mature tumours is well established, but with present methods permanent cure of the commonest and most malignant tumour, the medulloblastoma, cannot be hoped for. In this small series, 36 children were dead at the end of the first year after diagnosis, 13 were alive and one untraced. It seems unlikely that more than 20 out of 100 consecutive cases will achieve long survival (Bodian and Lawson, 1953).

It is perhaps more encouraging to consider conditions which are often mistaken for intracranial tumours. Space confines one to a simple enumeration of those that have been seen with some frequency.

A full classification would cover the whole field of paediatric neurology. General disorders such as cyclical vomiting, nephritis, hypertensive encephalopathy and hysteria, which sometimes cause confusion, are also excluded.

Intracranial Abscess.—This is suggested by the triad of (1) increased ICP; (2) lateralising signs; and (3) a focus of infection. Common sites of occurrence are the temporal lobe and the cerebellar hemisphere, and neurological signs are, by and large, more obvious than those caused by tumours in corresponding positions. With the universal use of antibiotics, evidence of generalised infection, such as pyrexia and leucocytosis, may be suppressed, greatly increasing the difficulty of diagnosis. The middle ear and mastoid are the commonest sources of the

infection, and the history of a recently discharging ear in a child manifesting signs and symptoms of increased ICP must be viewed with suspicion. Infection may, however, alight in the brain from a distant septic focus and there is an interesting association with cyanotic congenital heart disease.

A well-known variation on this theme is the infected dermoid cyst of the posterior fossa, of which there was one example in this series. Infection spreads into the cyst along a small sinus opening on to the skin over the occiput.

Tuberculous Meningitis.—In this condition premonitory symptoms of vague malaise and irritability are more frequent than in tumours. There is usually a history of contact with a case of tuberculosis and a positive skin test. The diagnosis is confirmed by the demonstration of organisms in the cerebrospinal fluid or gastric washings. A great increase in cells in the cerebrospinal fluid of cases of medulloblastoma and other highly malignant tumours may erroneously suggest this diagnosis.

Subdural Effusion.—This interesting and common disorder in infancy follows trauma (subdural haematoma) and meningitis (subdural hygroma). Birth trauma is the commonest cause, but the first manifestations often appear later (3-6 months). A pouch of fluid of varying extent and with a gradually thickening lining membrane is present over one or both hemispheres. Any or all of the following abnormalities may develop: frank hydrocephalus, intractable vomiting, failure to thrive, anaemia, convulsions (focal or generalised), backwardness or irritability with hypertonia and increase of deep reflexes (Guthkelch, 1953).

Case Report.—Robert H., 5 months. Investigated because of pallor. Normal delivery, but rather short second stage (in hospital). Several malaena stools on third to fifth days. Treated with intramuscular "Synkavit" (10 mg.). Haemoglobin on discharge (9th day) 60 per cent. (Haldane). Rather slow with feeds at home, but weight gain satisfactory and other progress normal. On examination, a bright chubby baby. Skull normal. Moderate exaggeration of all the reflexes of the limbs, which he moved well. Haemoglobin 37 per cent. Bone marrow examination and other investigations not of any significant value. Transfusion given. On tapping the subdural spaces, 5 ml. straw-coloured fluid obtained from each side, containing 1.5 g. protein/100 ml. Daily tappings then performed for three weeks, each producing up to 30 ml. of fluid. As there was no sign of fluid diminishing, bilateral temporal burr-holes were then made. Thick subdural membrane removed on left side after turning anterior temporal bone flap. Progress uneventful. At 2½ years physically and mentally normal.

Intracranial Venous Thrombosis.—There are three forms of thrombosis:—

- (1) Sagittal sinus thrombosis in marasmic and dehydrated infants.
- (2) Lateral sinus thrombosis following ear or mastoid infection.
- (3) Cortical thrombophlebitis secondary to a near or distant septic focus.

The first generally occurs in young infants and is usually terminal. The second rarely produces the high swinging pyrexia that was characteristic before the advent of antibiotics, and it can only be distinguished from abscess formation by full neurosurgical investigation. The third condition has only been recognised comparatively recently. It is usually of sudden onset, self-limiting and with a good prognosis. Convulsions, starting focally and characteristically spreading to both sides with hemiparesis and coma, are the salient neurological features.

Angiomatous Malformations.—When involving the cortex, these anomalies, varying from capillary telangiectases to huge cirroid arteriovenous connections, are a rare cause of focal epilepsy. The first indication of their presence, however, may be a sudden subarachnoid or intraventricular haemorrhage, manifesting in all degrees of severity up to swift coma and death. Auscultation of the skull sometimes elicits a bruit over the malformation.

Lead Poisoning.—In this important condition misdiagnosis of tuberculous meningitis or tumour may remove a slender but real chance of survival. Lead is absorbed by ingestion of lead-containing paint from toys, cot-sides, walls, etc., or more rarely from contaminated water or the fumes from burnt lead batteries. Coma and intractable convulsions may develop with extreme rapidity—perhaps related to the acidosis of an infection and consequent release of lead from the bones. The majority of the well-known features are illustrated in the following case, but one worth stressing is the increased excretion of glucose and amino-acids in the urine (Wilson *et al.*, 1953). The condition is excellently reviewed by Marsden (1955).

Case Report.—Harry C., 4 years 3 months. Admitted with deepening unconsciousness for 24 hours and twitching for six hours—first the right cheek only, then the left limbs as well. Had not been well for six weeks prior to this; listlessness, frequent vomiting, abdominal pain and pallor. For one year the child had persistently gnawed and chewed painted furniture, doors and skirting boards. On examination, he was unconscious, with eyes widely open and continuous nystagmus. Moved left limbs less readily than right in response to painful stimuli. Retinal veins full, but no frank swelling of the discs. Plantars both extensor. B.P. 140/85. X-ray examination: markedly increased density of wrist

metaphyses and skull sutures separated. Haemoglobin 62 per cent. (Haldane); marked punctate basophilia of red cells. Urine contained reducing substance identified on chromatography as glucose, a trace of albumin and excessive amino-acids. Lumbar puncture—much increased pressure; protein 70 mg./100 ml. Cells less than 1 c.mm. Death took place shortly after admission in spite of sedation and medical and surgical attempts at lowering the ICP. De-leading agents which remove the lead in an innocuous form were not then available. At autopsy, brain engorged and tense with oedema. Intranuclear inclusions in kidneys and liver (Marsden and Wilson, 1955). Lead was demonstrated in the long bones.

Benign Stenosis of the Aqueduct of Sylvius.—

This is one of the causes of congenital hydrocephalus and hydrocephalus in infancy, but there is also a well-defined group occurring in late childhood and adolescence. These children develop signs of increased ICP without other neurological accompaniments, and there is unremitting deterioration because of the relative rigidity of the skull at this age. The condition is of particular importance because of its amenability to treatment. An outlet for the fluid is secured by Torkildsen's operation or a third ventriculostomy. Provided that significant deterioration in vision has not already occurred, the prognosis is excellent.

CONCLUSIONS

An attempt has been made to define the modes of presentation of some of the commoner intracranial tumours in children. By and large the picture is simply that of raised pressure within the skull—increase in size of the head, headache,

vomiting and papilloedema—with other neurological signs relatively late in their appearance. A completely different picture is produced by the brain stem glioma in which pressure signs are late or absent and multiple neurological deficits early and constant.

Seven conditions commonly causing confusion are discussed.

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