THE DIAGNOSIS OF BRAIN TUMORS

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The purpose of this paper is to describe briefly the history and physical findings in a few of the more common types of intracranial tumors, namely the gliomas, the meningiomas, the pituitary and the acoustic tumors.

Strictly speaking, the glioma is the only true *brain* tumor, that is, it is the only tumor arising from the brain tissue proper. The meningioma arises from the membranes covering the brain, the pituitary tumor arises from epithelial cells of the anterior lobe of the hypophysis and the acoustic tumor arises from the sheath of the eighth cranial nerve. The glioma, or true brain tumor, however, does not arise from the functioning nervous elements of the brain but from the supporting or glial tissue from which it derives its name. The glioma therefore may be compared to the sarcoma arising from connective tissue. There are four commonly encountered types of gliomas classified according to cell morphology and some twelve more or less common types.

The neurological surgeon is no longer content to diagnose and correctly localize an intracranial tumor, but, with increasing knowledge of the life history of the various types of tumors, he attempts to predict the exact histological structure in each case as it confronts him. It is not merely to satisfy his vanity that the surgeon thus attempts to foretell the histological structure of the tumor with which he is about to deal. This foreknowledge permits him to plan his operative approach more satisfactorily and to deal with the neoplasm more adequately than he otherwise could.

Increased Intracranial Pressure

The symptoms commonly encountered in all types of intracranial tumors are the symptoms of increased intracranial pressure. These symptoms are headache, vomiting which is frequently projectile and unassociated with nausea, failing vision due to choking of the optic discs and in the late stages, a blunting of the intellect. There are, of course, conditions other than tumor which may increase the intracranial pressure and produce this same train of symptoms, the most common being meningitis, brain abscess and traumatic hematoma. However, if these symptoms appear gradually and without a history of a preceding infection or head injury, these other conditions can be eliminated.

There are only two reliable methods of determining the presence of increased intracranial pressure. They are ophthalmoscopic examination and spinal puncture. Edema of the optic discs due to increased intracranial pressure is readily recognized by fundus examination. If this is present, it renders spinal puncture unnecessary. The presence of increased intracranial pressure cannot be determined by spinal

puncture without employing a spinal manometer. The estimation of pressure by observing the rate of flow of the fluid from the needle is absolutely unreliable in most instances.

Increased intracranial pressure with its attendant symptoms, however, is not invariably present in patients with brain tumor. As might be expected, it appears earlier and progresses more rapidly in the more malignant types of gliomas while it comes on later and progresses more slowly in the meningiomas. In pituitary tumors, it is very seldom encountered for the simple reason that, because of the location of these growths, they are apt to cause complete blindness by direct pressure on the optic nerves before they can attain sufficient size to increase the intracranial pressure.

Brain Tumor in Children

Usually the symptoms of increased intracranial pressure in children occur before there are any focal signs to suggest the location of the lesion. Recurring attacks of headache and vomiting without nausea are the rule. These may occur at intervals of weeks or months with completely symptom-free intervals. Recurring bouts of sudden vomiting without headache are not uncommon. The physician's attention, therefore, may be centered on the gastro-intestinal tract for months or until serious visual disturbance points the way to the correct diagnosis. The visual disturbance may be a diplopia due to sixth nerve paresis from the increased intracranial pressure, or this paresis may proceed to an obvious internal squint before the child complains of it. Failing vision also may reach the point where it is evident to the parents before the child is aware of it. The important thing to remember is to make a fundus examination in any child who is having recurring or continuous attacks of headache or vomiting. Another simple, though later sign of increased intracranial pressure in children is the cracked pot sound on percussion of the skull (Macewen's sign). This is due to the separation of the cranial sutures which are not firmly united in children.

The most common tumor in childhood is the glioma of the cerebellum. These are almost always located in the midline between the two cerebellar lobes and are of two varieties. The cystic astrocytoma is the most benign of the gliomas and yields brilliantly to surgery. The other type, known as a medulloblastoma, is composed of highly undifferentiated cells. It cannot be excised completely but responds better to roentgenotherapy than any other brain tumor. These tumors, because of their midline position, soon obstruct the flow of cerebrospinal fluid from the ventricles and thus cause an obstructive hydrocephalus. Local symptoms are frequently lacking or at most may consist of an instability of gait with frequent falling. One ever-present sign in these cases is anesthesia of the cornea. Nystagmus, ataxia and hypotonia, which are the common

signs of most cerebellar lesions, may be very slight or even absent in these midline tumors.

Another less common tumor in childhood and adolescence is the tumor of the hypophyseal stalk usually referred to as the craniopharyngioma or Rathke's pouch cyst. This tumor arises just above or within the sella turcica from embryonal cell rests left by the outpouching from the primitive nasopharynx which goes to form the anterior lobe of the pituitary gland. Because of pressure on this gland, these tumors usually produce signs of pituitary hypofunction. These children, therefore, are apt to be chubby and dwarfed and the signs of puberty are lacking. In spite of their location in close proximity to the optic nerves and chiasm and because of their cystic nature and slow growth, the resulting loss of vision comes on late and appears gradually. When the cyst has attained sufficient size to compress the third ventricle and thus produce an obstructive hydrocephalus, symptoms of increased intracranial pressure supervene with rapid loss of the remaining vision. These benign epithelial lined cysts are best approached by a frontal craniotomy. Though their total removal is attended by a high mortality due to damage to the surrounding nervous structures, they may occasionally be dissected out with surprising ease. A partial removal is most commonly employed and usually results in a long symptom-free interval. They are not at all sensitive to roentgenotherapy.

Gliomas of the cerebral hemispheres occur in children but they do not produce a syndrome as characteristic as those described above. The meningiomas and neurofibromas rarely occur before the age of maturity.

Glioma of the Cerebral Hemispheres

In adult life, gliomas of the cerebral hemispheres are encountered more frequently than any other type of tumor. These vary in malignancy from the very rapid growing glioblastoma multiforme to the slow growing benign oligoglioma. A fairly typical case history of the former type of tumor is as follows: A healthy active man of 45 years begins to complain of headache. He ignores it for a few weeks but the headaches increase in frequency and severity so that he stays home from work. It is then noted by the family that his memory is impaired and that he is occasionally mildly confused. The symptoms steadily increase in severity and new ones appear. There is numbness or weakness of one side of the body and some blurring of vision. The patient is hospitalized about three months after the appearance of the initial symptom. On examination at this time, the patient is found to be lethargic and confused and he complains of headache when aroused. There is an advanced choking of the optic discs with recent retinal hemorrhages. The neurological signs indicate the location of the lesion fairly definitely. At

craniotomy, the cerebral cortex appears flattened and the intracranial pressure is very high. An incision through the cortex reveals the soft infiltrating growth beneath the surface. Provided with an adequate decompression, the patient improves temporarily after which the intracranial pressure recurs and he dies about four months after operation. This is the type of neoplasm which has gained for brain tumors a most unenviable reputation. It represents about 15 per cent of all intracranial tumors.

The oligoglioma is at the other end of the scale as far as malignancy is concerned. This tumor may cause symptoms for five or ten years and, after a partial removal, the patient may not succumb for another five or ten years. The initial symptom is most commonly a convulsion, usually Jacksonian in character. The convulsions recur at first at long intervals, perhaps years, but steadily increase in frequency and are accompanied by a gradually increasing weakness and spasticity of the extremities affected by the convulsion.* With the appearance of headaches, the patient is referred to the surgeon. From a detailed description of the attacks, it is sometimes possible to localize the lesion accurately without even seeing the patient. This is especially true if the tumor is in the parietal lobe in which case the patient will describe a numbness of the limb preceding the attack. Other aura of immense localizing value are a sudden stench perceived by the patient, indicating an irritation of the uncinate gyrus, or flashes of light in one half of the visual field indicating the opposite visual pathway. Calcium deposits in the oligoglioma are rather frequently visible on the x-ray film and this constitutes another important diagnostic feature.

At operation, this tumor is found near the surface and, due to its firmness, it is readily palpable. It can easily be dissected out on all sides but one. Here it shades off imperceptibly into the white matter. If this area is radically excised, a permanent cure is possible. If it can be done without too great a sacrifice to the patient, it is better to excise the entire affected lobe with the contained tumor.

Between the glioblastoma on the one hand and the oligoglioma on the other, there are many types of gliomas of varying degrees of malignancy and less clear-cut clinical syndromes.

Pituitary Tumor

The pituitary tumor produces a fairly definite symptom complex. This tumor arises from the cells comprising the anterior lobe of the pituitary gland. Histologically and clinically, there are two commonly encoun-

*Convulsions appearing for the first time after the age of 30 are most frequently due to brain tumor and the burden of proof is upon him who would assume otherwise. If the convulsion is followed by a transitory weakness of an arm or leg, the presence of a tumor is particularly indicated.

tered types. The chromophobe adenoma arises from the chromophobe or neutral staining cells and produces a picture of pituitary hypofunction. The acidophilic adenoma arises from the granular cells which take the acid stain and it produces a mixed picture of hyper- and hypofunction. A third and very rare type has been described by Cushing as a basophilic adenoma. This tumor arises from the granular cells which take the basic stain, but it is still questioned by many pathologists and clinicians.

Clinically, the symptoms of pituitary tumor may be divided into the endocrine signs which usually appear early and the pressure signs such as headache and failing vision which appear later. In the most common or chromophobe type, the early symptoms are usually obesity, reduction of libido and in the female, amenorrhea. In the acidophilic type, the symptoms are gigantism if the tumor arises prior to the completion of epiphyseal union, acromegaly, reduction of libido, and, in the female, usually amenorrhea. The former variety is accompanied by a subnormal metabolism and frequently by an increased glucose tolerance. The latter variety is accompanied by an increased metabolism and frequently by elevated blood pressure and decreased glucose tolerance which may amount to a frank diabetes. In neither type is surgical intervention warranted until pressure symptoms intervene. The pressure symptoms are common to the two types and are as follows: Headache which is due presumably to increased pressure within the expanding sella turcica and impairment of vision which becomes manifest when the adenoma has grown upward sufficiently to compress the visual pathways. The most common type of visual loss is a bilateral reduction in visual acuity and a bitemporal hemianopsia due to pressure of the optic chiasm. However, if the lesion expands more rapidly on one side, a unilateral loss of vision and homonymous hemianopsia may result. The optic discs show the pallor of a primary atrophy. In the late stages, somnolence and diabetes insipidus may occur, indicating damage to the hypothalamic structures.

Roentgen examination of the skull in the presence of pituitary tumor always discloses a ballooning out of the sella turcica with the floor pushed downward into the sphenoid sinus. This is not to be confused with the erosion of the clinoid processes produced by tumors arising at a distance from the sella. As stated before, the intracranial pressure is rarely increased in pituitary tumors because serious visual loss usually points the way to the correct diagnosis before the lesions are large enough to cause this condition.

Practically all surgeons agree that these lesions can best be treated by a right frontal craniotomy. The acidophilic type sometimes responds well to roentgenotherapy.

Acoustic Tumor

In the acoustic tumor, a careful chronological history will often establish the diagnosis and almost obviate the necessity for other studies. As these tumors grow on the sheath of the eighth cranial nerve, the initial symptom, as might be expected, is tinnitus accompanied by or followed shortly by increasing deafness. Sometimes there is a brief period of mild vertigo at the onset due to irritation of the vestibular fibers of the eighth nerve. As these fibers are paralyzed by the tumor much earlier than are the auditory fibers, this early vertigo, if present, soon subsides. There then follows a progressive impairment of hearing in the affected ear which becomes complete in a period of months or years. The next symptom is usually a mild unsteadiness of gait which progresses slowly to a marked cerebellar ataxia. During the progression of the ataxia, headache and cranial nerve signs make their appearance. The headache is more commonly suboccipital and frontal and may be confined to the side of the tumor. The symptoms of involvement of the adjacent cranial nerves consist of numbness in the distribution of the trigeminus and occasionally of facial pain which may resemble a true tic doloreaux. Facial weakness as evidenced by a unilateral slowing of the blinking reflex and slight widening of the palpebral fissure is frequently present. As the headache becomes more severe, it is accompanied by forceful vomiting, diplopia and failing vision.

Examination at an early stage in this disease will disclose merely unilateral nerve deafness and lack of response to the labyrinthine tests of Barany on the affected side. Later on, there will appear anesthesia of the cornea, coarse nystagmus especially on looking toward the side of the tumor, Rombergism, staggering toward the affected side, slight facial weakness, internal squint, choked discs and unilateral suboccipital tenderness. Roentgen examination frequently will disclose some erosion of the posterior surface of the petrous bone.

The Meningioma

The meningioma is a benign, encapsulated growth originating in the meninges. It occurs most commonly in the fourth and fifth decades of life and is very rare prior to the third.

Although a preoperative diagnosis of meningioma is not always possible, there are certain findings in the majority of cases which make the diagnosis likely. The following signs and symptoms should lead one to suspect this type of tumor. The onset of symptoms may date back years rather than months as is commonly the case in the more rapidly growing type of lesions. The initial symptom in approximately one-third of the cases is a Jacksonian convulsion. X-ray demonstration of

local thickening or erosion of the skull or actual calcification within the tumor occurs in one-fourth of the cases. A palpable swelling of the overlying scalp occurs in a much smaller proportion. However, the point upon which the differential diagnosis most frequently hinges is the occurrence of a slow growing tumor in a location known to be a favored site of the meningioma. These favored sites in order of their frequency are:

(1) Attached to the lateral wall of the sagittal sinus near the vertex. In this location, Jacksonian convulsions and hemiparesis beginning in the opposite leg are common symptoms.

(2) On the sphenoidal ridge. Here the localizing signs may be meager or absent. The most important evidence is obtained from the roentgen examination which may show erosion of the sphenoidal ridge or dilatation of the middle meningeal groove. Tenderness and fullness in the temporal region is a very suggestive sign.

(3) Over the convexity of the cerebral hemispheres. In this location, the occurrence of Jacksonian convulsions together with roentgen evidence of bony proliferation or erosion is frequent.

(4) From the tuberculum sellae. A meningioma in this location announces its presence when symptoms suggestive of pituitary tumor develop in a middle-aged person, but the roentgenogram fails to show the typical sella picture.

(5) In the cerebellopontine angle. Tumors in this location are difficult to distinguish from acoustic neurofibromas.

(6) In the olfactory groove. Here the story is one of progressive loss of sense of smell, mental symptoms and headache.

(7) On the sheath of the Gasserian ganglion. In this site, the tumor causes trigeminal pain with associated impairment of sensation in the distribution of the fifth nerve.