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PATHOPHYSIOLOGY AND IMAGING

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INTRODUCTION

The skull and bony structures of the spine and cranium combined with the cerebral and spinal meninges form a relatively rigid construction of protection for the brain, cerebrospinal fluid (CSF) within the subarachnoid space and the craniospinal arteriovenous system. Alterations in the form of an increase in the volume of brain parenchyma, the CSF or the vascular components, or alterations of the bony compartment that contains them can give rise to a progressive increase in intracranial pressure.

The terms intracranial hypertension (ICH) and hydrocephalus do not define the same entity, but rather refer to a variation in the container-content relationship in the first case, and in the second case to an accumulation of CSF with a dilation of the cerebral ventricles; however, in the latter instance, there will also be some degree of necessarily associated ICH. ICH patients usually present the classic sign-symptom triad of headache, vomiting and papillary elevation (7, 9).

The headache has particular semeiologic characteristics, as it usually occurs upon awakening and has a frontal location. It is believed to be caused by the traction exerted on the meningeal neural structures both within the meninges as well as within the related vascular structures. Not only the parietal meningovascular structures are involved, but also innervated are the dura mater of the skull base, the walls of the dural venous sinuses and the bridging veins.

Vomiting is also manifested clinically in the morning, similarly upon awakening, and before the patient has eaten. It is not preceded by vegetative signs such as nausea or ptyalism (excessive flow of saliva) and is almost always associated with headache. The pathogenic mechanism responsible for the vomiting in cases of ICH is still unknown, however it has been observed that the headache subsides partially after the episode(s) of vomiting.

Bilateral ocular papillary elevation is caused by hypertension of the CSF pathways, accompanied by an increase in the CSF pulse pressure transmitted to the optic nerve head. This pulsatile pressure increase is directly broadcasted to the optic nerve trough of the optic subarachnoid space via the meningeal sheaths that coat the entire length of the optic nerve, thereby causing a progressive vascular compression-congestion of the optic nerve head and outward physical bulging of the optic papilla into the rear of the globe.

Minor signs and symptoms of ICH include:

1) Diplopia: Diplopia is caused by a paresis of the 4th cranial nerve as a consequence of the pressure increase, which injures the fragile abducens nerve;

2) Eyesight impairment: Protracted oedema, compression and expansion of the optic nerve head cause regressive phenomena entailing permanent damage and resultant atrophy, ultimately terminating in an impairment of eyesight. In extreme cases amaurosis occurs, a process that is almost always irreversible.

3) Endocrinological alterations: Chronic ICH is associated with a reduction in function of the hypophysis, with resultant alterations in hormone production (e.g., FSH, LH, TSH, ACTH, GH) and changes in the gland's response to physiological and pharmacological stimuli.

4) Disorder of recent memory: This nonspecific sign is almost always present in patients with long clinical histories of minor intracranial pressure elevations. The deficit would seem to affect the recall of recent events, rather than memory of chronological order or long-term memory.

There is a wide range of possible underlying pathological conditions responsible for the ICH, such as neoplastic, vascular, inflammatory, toxic, metabolic, traumatic or hormonal causes. This chapter deals with the physiopathology of intracranial hypertension, cerebral oedema, pseudotumor cerebri and hydrocephalus.

THE PATHOPHYSIOLOGY OF INTRACRANIAL HYPERTENSION

The intracranial space is divided by the tentorium cerebelli into the supratentorial and subtentorial compartments, which communicate with one another via the tentorial incisura. This intracranial space is completely occupied by solid or fluid components, including the brain, the subarachnoid space and cerebrospinal fluid, and the blood and attendant vascular walls; its content is considered uncompressible: variations in intracranial pressure are closely dependent on and correlated with variations in these solid and fluid subcomponents.

Over a wide variety of pathological as well as normal physiological situations (normal health, changes of posture, some forms of illness, etc.) the quantity of blood present in the intracranial space does not undergo significant overall volume alterations. The blood exiting the cranium via the outgoing venous system guarantees sufficient space for the arterial blood that enters the skull with each systolic cycle, thus constituting a self-regulating mechanism of cerebral blood flow. This flow self-adjusts to intracranial pressure changes in order to guarantee steady blood flow to the brain.

Normal intracranial pressure

In order to clearly understand normal variations in intracranial pressure (ICP), a few simple models will be discussed as examples (10). Fig. 3.1a illustrates a patient in the lateral decubitus position, with a spinal tap needle within the lumbar subarachnoid space. The plane of the needle passes through the midsagittal plane of the spine and skull. In the case of the cranium being open to atmospheric pressure, the fluid pressure measured in the spine is equal to the distance between the middle sagittal plane and the uppermost lateral wall of the cranial meninges.

When the patient is in the vertical position (Fig. 3.1b) the hydrostatic column of CSF approximately equals the distance between the point in which the spinal tap is positioned and the top of the cranial meninges if it is open to the influence of atmospheric pressure. However, if the skull is excluded from the direct action of atmospheric pressure but instead is imagined to be completely closed, there is no change in pressure on the passage from the horizontal to the vertical position (Fig. 3.1c), because in this case the spinal portion of the system alone is subject to the action of atmospheric pressure.

Fig. 3.1d illustrates a normal person in a sitting position with the same lumbar subarachnoid needle in place. One can observe how the measurement of pressure reaches that expected of the cervico-thoracic junction level, thus showing to be lower than it would have been if the intracra-

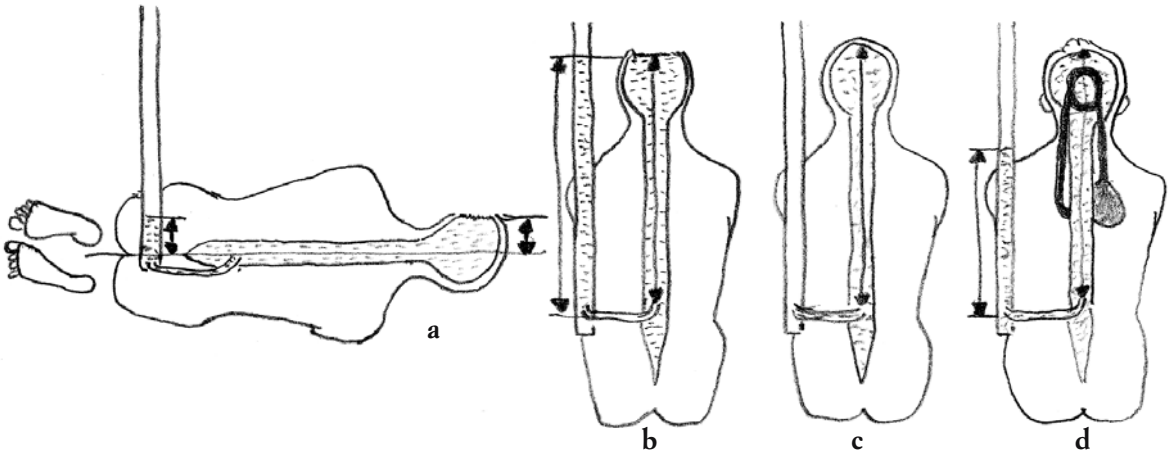


Fig. 3.1 - See text.

nial space had been open to the direct influence of atmospheric pressure, and greater than it would be were the system completely closed.

This series of experiments demonstrates that the intraarachnoid craniospinal system cannot be considered a completely closed system, as it is in contact with atmospheric pressure through the craniocerebral blood vessels. For the most part this pressure effect is transmitted through the veins that lead from the skull base or from the peridural venous plexus at the level of the foramen magnum.

When measuring CSF fluid pressure, the values recorded at lumbar level are greater than those obtained from the cervical area, which are, in turn, higher than those recorded directly at an intracranial level. In normal subjects, the balanced hydrostatic point is situated at the cervical-thoracic junction: it remains the same when measured in either the horizontal or vertical position. The zero pressure point is situated craniad to this in the cervical region of the spine. Normal intracranial pressure values fall between 9 and 13 mm Hg, and therefore values higher than 15 mm Hg are considered pathological.

The pressure-volume ratio

The various phenomena that occur in the volumetric alterations of the intracranial components are linked to physical laws expressed by the pressure-volume ratio (4).

Intracranial hypertension is the result of an increase in the total absolute intracranial volume of the contained soft tissue and fluid components. This volume increase may be caused by expansions in the volume of the cerebral parenchyma, circulating blood or CSF, either singularly or in combination. ICH does not appear immediately in response to a gradual, minor increase in volume of one of these constituents, as the other components of the system are able to compensate to a certain degree, thus maintaining the total volume at a constant level.

The Monro-Kellie doctrine establishes that because the volume within the skull is not able to increase, there is a fixed relationship between the various intracranial constituents. Therefore, if normal intracranial pressure is to be maintained, an increase in volume of one or more of these components or the development of a space-occupying lesion can only take place if there is a reduction in the volume occupied by the others.

As alterations in the volume of the intracranial nerve tissue can only be very slight, it follows that the larger compensatory alterations take place at the expense of the volume of the CSF and/or the circulating blood within the craniospinal system. It is estimated that the supratentorial structures can absorb approximately 50% of such volume alterations, the subtentorial structures 20% and the spinal compartment 30%. Any pathological process

that alters the volume of these compartments reduces their total compensatory power.

Generally speaking, the CSF compartment in particular has a relatively high capacity for absorbing increases in intracranial pressure. However, this capacity is contingent on the condition that the pressure increase takes place gradually and slowly. In part this is due to the high resistance of the arachnoid villi to CSF filtration; when this CSF-venous drainage rate limit is reached, the intracranial pressure will rise. The system's fluid reabsorption capacity can at the most guarantee a CSF drainage rate of 1 ml/per minute. It therefore follows that an expansion, for example, of a subdural haematoma may not precipitate signs of intracranial hypertension if it forms sufficiently slowly.

The opposite is true for the vascular compartment, which has very little reserve space without circulatory insufficiency arising within the brain. However, what reserve there is adapts very rapidly, thanks to the direct connection of the cerebrovascular network with the systemic circulation.

Fluctuations in cerebral blood flow are governed by a number of factors such as carbon dioxide levels, adenosine, potassium, prostaglandins and anaesthetics. Numerous medicines, such as xantine, hypertonic saline solutions and hyperosmotic solutions can also increase cerebral blood flow. After a certain point is reached, an increase in cerebral blood flow causes an increase in intracranial pressure.

In healthy subjects, intracranial pressure appears to be controlled and maintained within a relatively narrow range from moment to moment by minor alterations of CSF volume. However, when intracranial hypertension is long-standing and considerable (e.g., cerebral oedema, intracranial space-occupying lesion, etc.), the resulting hypercapnia induced by these conditions causes an increase in intracranial pressure in part due to the reduced amount of fluid available for reabsorption.

To summarize, the abovementioned compensation mechanisms therefore have a varying role in intracranial pressure homeostasis. The intracranial volume occupied by pathological alterations can be obtained at the expense of the CSF volume, the amount of endovascular blood or, to a lesser extent, the brain's intracellular and interstitial water content. However, whereas prolonged mass effect upon the brain can produce a reduction in the amount of brain tissue water, variations in the parenchymal cellular tissue component are practically negligible. Moreover, although intraparenchymal blood volume can represent a small buffer reserve, realistically it is the intracranial CSF fluid content that is the most important buffer volume when intracranial alterations occur.

Intracranial pressure waves

Temporary intracranial pressure readings are not accurate in revealing the real characteristics of pressure waves. However, measurements recorded over a long period of time show a minor pulse-type pattern, due primarily to the effects of breathing and cardiac activity (5).

Three types of pressure wave alterations are described in patients with intracranial hypertension. The smallest waves, termed subtypes B and C, are accentuations of physiological phenomena: the C wave represents the breathing component, whereas the B wave is an expression of cardiac activity. The A wave, the only one of pathophysiological importance, can be divided into two forms:

1) rhythmic pressure fluctuations, with intervals of 15-30 minutes;

2) plateau waves, which last for longer periods of time.

The former have a frequency of 2-4 cycles per hour, they begin spontaneously from an average or moderately high pressure base and reach levels of 60-100 mm Hg, before returning to their baseline values. The latter often exceed a value of 100 mm Hg and represent, like rhythmic waves, a serious prognostic index of imminent serious cerebral consequences.

The relationship between intracranial pressure and CSF production

The CSF produced by the choroid plexuses is not a mere plasma filtrate, but rather an active fluid body tissue (e.g., certain electrolytes

have higher concentrations than does the systemic blood) that functions in part via the action of carriers and pumps.

Increases in intracranial pressure cause a reduction in the cerebral perfusion pressure, which in turn results in a reduction of superfiltrate production by the choroid plexuses by hindering the activity of the active transport carriers and fluid pump. CSF production ceases at an intracranial pressure of 22-25 mm Hg.

The consequences of intracranial hypertension on cerebral circulation

Intracranial hypertension has a direct effect upon cerebral perfusion. An increase in intracranial pressure would cause a halt of the blood cerebral blood flow when arterial and intracranial pressure become equal, were it not for the intervention of two compensatory mechanisms: arteriolar-capillary vasodilatation and an increase in systemic arterial pressure.

The first of these mechanisms is a cerebrovascular self-regulation mechanism caused by a parietal vessel reflex that acts on the muscular fibres of the vessel wall, relaxing them when the pressure inside the vessels drops; a local increase in CO_2 and metabolic acids have a vasodilatory effect.

The increase in systemic pressure is a result of a bulbar (i.e., brainstem) reflex triggered by brainstem ischaemia and would only seem to come into play once the cerebrovascular self-regulation mechanisms have failed. The latter are efficient up to intracranial pressure values of 50-60 mm Hg, beyond which passive vasodilatation occurs. Increases in intracranial pressure then take place until complete circulatory arrest ultimately occurs.

Mechanical consequences of intracranial hypertension

By altering the mechanisms that keep intracranial pressure within normal limits, intracranial space-occupying lesions have an effect on cerebral perfusion but my also induce displacements of the cerebral tissues within the cranial cavity.

In childhood, intracranial hypertension takes longer to manifest itself than it does in adults because of the elasticity of the immature skull. The absence of cranial suture closure allows a degree of expansion of the bony structure of the calvaria of the skull when an increase in intracranial pressure occurs, whereas this adaptability no longer exists in adults having a rigid skull.

The matters discussed thus far may suggest that an increase in intracranial pressure is distributed uniformly inside the skull and is therefore borne equally by the various parts of the neuraxis. However, this is not true for two reasons: the majority of lesions causing intracranial hypertension are focal in nature, and, the cerebral parenchyma has mechanical properties that are similar to both those of elastic solids as well as those of viscous fluids.

The nervous tissue adjacent to a mass undergoes deformation, and the pressures are distributed in various ways: they are high in the vicinity of the lesion and gradually diminish with distance, thus creating pressure gradients under which the nervous tissue is displaced. These shifts of the nervous tissue, favoured by the oedema that reduces the viscosity of the parenchyma, may result in internal cerebral herniations. Neoplasia, abscesses, haematomas and other space-occupying lesions may cause these dislocations of the contents of the cranium.

When intracranial hypertension associated with mass formation occurs, initially the most important factor to recognize clinically is not the nature of the cause, but rather the presence or absence of internal cerebral herniation. The manner in which these shifts occur is more or less similar irrespective of their cause, although they vary with the site, size and rapidity with which the space-occupying lesion evolves.

In the initial stages, any expanding lesion exerts uniform pressure on the surrounding tissues. These forces are opposed by others, for example the cerebral tissue itself and the hydrodynamic resistance of the CSF-containing ventricles that oppose resistance to deformation from compressive forces. Another opposing force is the cerebral vascular system and the contained arterial pressure, as these arteries constitute a sort of skeleton for the surround-

ing cerebral tissue. In addition to the arteries, the veins, nerves and meninges also oppose this type of pressure.

It should also be remembered that the falx cerebri and the tentorium cerebelli divide the skull into three compartments and provide further opposition to the dislocating effects of mass lesions. This division of the intracranial space allows displacements of the brain parenchyma under the effect of space-occupying processes only in certain directions, including: within the same compartment, from one supratentorial compartment to another, beneath the falx cerebri, downward or upward through the tentorial hiatus, and from the posterior cranial fossa downward into the spinal canal through the foramen magnum.

In the presence of a space-occupying lesion, a sequence of compensation mechanisms can be described. Initially, the subarachnoid spaces adjacent to the lesion are compressed, with a flattening of the superficial gyri, distortion of the ventricular cavities and a deformation and dislocation of the nearby arteries and veins. This is followed by a second phase in which the volume of the brain tissue involved increases due to oedema, and the CSF spaces are no longer able to compensate for the primary and secondary mass effect. Any further compensation requires a shift of parenchymal tissue from one anatomical compartment to another, with the consequent development of internal cerebral herniation.

Subfalcian herniations are observed in association with dominant hemicranial lesions; the degree of brain dislocation beneath the falx varies according to the original site and size of the mass lesion. For example, masses that originate in the frontal regions are more frequently associated with this kind of herniation as the falx cerebri is less broad anteriorly and consequently the free space below is greater than that posteriorly, where the falx and the splenium of the corpus callosum are in closer proximity. This type of cerebral herniation involves the supracallosal and cingulate gyri, the corpus callosum, the anterior cerebral arteries and their branches, the frontal horns of the lateral ventricles and the midline cerebral veins. The third ventricle is also shifted across the midline.

Axial herniations take place through the tentorial hiatus in either an upward or downward direction. This type of internal herniation causes a distortion and compression of the brainstem. When downward and the mass effect is sufficiently large, the herniation may also affect the lower cerebellum, which can be displaced through the foramen magnum.

Temporal herniations involve the medial part of the temporal lobe and in particular the hippocampus and the uncus, which can herniate either unilaterally or bilaterally. This herniation stretches the oculomotor nerve, compresses the posterior cerebral artery and can impinge on the cerebral peduncle on one or both sides. These events are followed by secondary lesions including oedema and haemorrhage.

Temporal herniations therefore threaten functions regulated by the brainstem, including vigilance, muscle tone, voluntary motion and vegetative functions. A unilaterally expanding temporal mass lesion is less favourable than a bilateral one because it can cause temporal herniation at an earlier stage in the mass forming process.

Downward cerebellar herniations are observed as a complication of expanding processes in the posterior cranial fossa and may occur in two forms that are often associated. In the first type, the cerebellar tonsils are thrust towards the upper spinal canal through the foramen magnum. In the second type, the upper part of the cerebellar vermis (i.e., culmen) herniates upwards through the tentorial hiatus, thus pushing the lamina quadrigemina and the midbrain forwards. The resulting injury to the brainstem depends in part upon vascular compression and secondary ischaemia of the upper brainstem.

Finally, internal cerebral herniations can obstruct the subarachnoid cisterns, thus preventing the free circulation and proper drainage of CSF. Above the level of the herniation, intracranial pressure tends to increase, whereas below it it is normal or only slightly raised. These differentials in CSF pressure add to the vector of thrust and thus worsen the herniation.

This in part explains why in such cases a lumbar puncture can precipitate a worsening of the clinical status.

The relationship between intracranial pressure and cerebral function

Many patients with obstructive hydrocephalus or pseudotumor cerebri show modest signs of cortical compromise in the presence of high intracranial pressure, the degree of which depends in part upon whether or not the cerebrum in such patients was normal prior to the onset of the pathological event. The situation is somewhat different in patients with pre- or coexistent parenchymal lesions, such as neoplasia or contusions. In addition, an increase in ICP due to the volume of the mass, cerebral oedema and/or vasodilatation secondary to hypercapnia combine with local cerebral hypoperfusion, the function of the brain adjacent to the expanding lesion can be compromised with even relatively low elevations of ICP (e.g., 15-25 mm HG).

In summary, an increase in ICP causes malfunction of cerebral function through four related mechanisms: a generalized reduction of cerebral blood flow, a compression of the tissue surrounding the focal mass with local cerebral microcirculatory compromise, brainstem compression and an internal herniation of brain tissue.

PATHOPHYSIOLOGICAL CLASSIFICATION

The general causes of intracranial hypertension can be summarized as an increase in volume of one or more of the intracranial soft tissue components: the parenchyma, the CSF volume and the blood volume.

ICH Resulting From Cerebral Oedema

Cerebral oedema (13) is defined as an increase in the volume of the encephalon caused by an increase in its water content. This content may be focal or generalized. When widespread and severe it can be associated with neurological signs and may ultimately result in internal cerebral herniation. Cerebral oedema can be divided into a number of different types, including: vasogenic, ischaemic, cytotoxic and interstitial related to hydrocephalus. Cerebral oedema is usually accompanied by intracranial hypertension, but there are exceptions, especially when the degree is minor.

On CT scans cerebral oedema is characterized by an area of hypodensity as compared to the parenchyma. On MR oedema is hypointense on T1-weighted images, more intense than CSF and less than the parenchyma. On T2-weighted scans, the relative hyperintensity of oedema varies, and, depending on the protein content, it can appear more or less intense relative to CSF.

Vasogenic oedema

Vasogenic oedema is the most common form of cerebral oedema and is typically associated with neoplasia, abscesses, intraparenchymal haematomas and traumatic contusion. It is caused by an increase in the permeability of the blood-brain barrier and usually affects the white matter with a resulting increase in density/intensity between white and grey matter on medical imaging. These alterations are due to an increase in the volume of the extracellular fluid.

Vasogenic oedema is easily discernible in the white matter as it generally spares the grey matter and exerts mass effect on the ventricular structures (Fig. 3.2). After IV contrast medium administration, a curvilinear or gyral pattern of enhancement can often be observed also related to the increase in blood-brain permeability.

Ischaemic oedema

Ischaemic oedema is the result of a cerebrovascular accident. The pathological process involves both white and grey matter with a loss of differentiation on imaging between the two

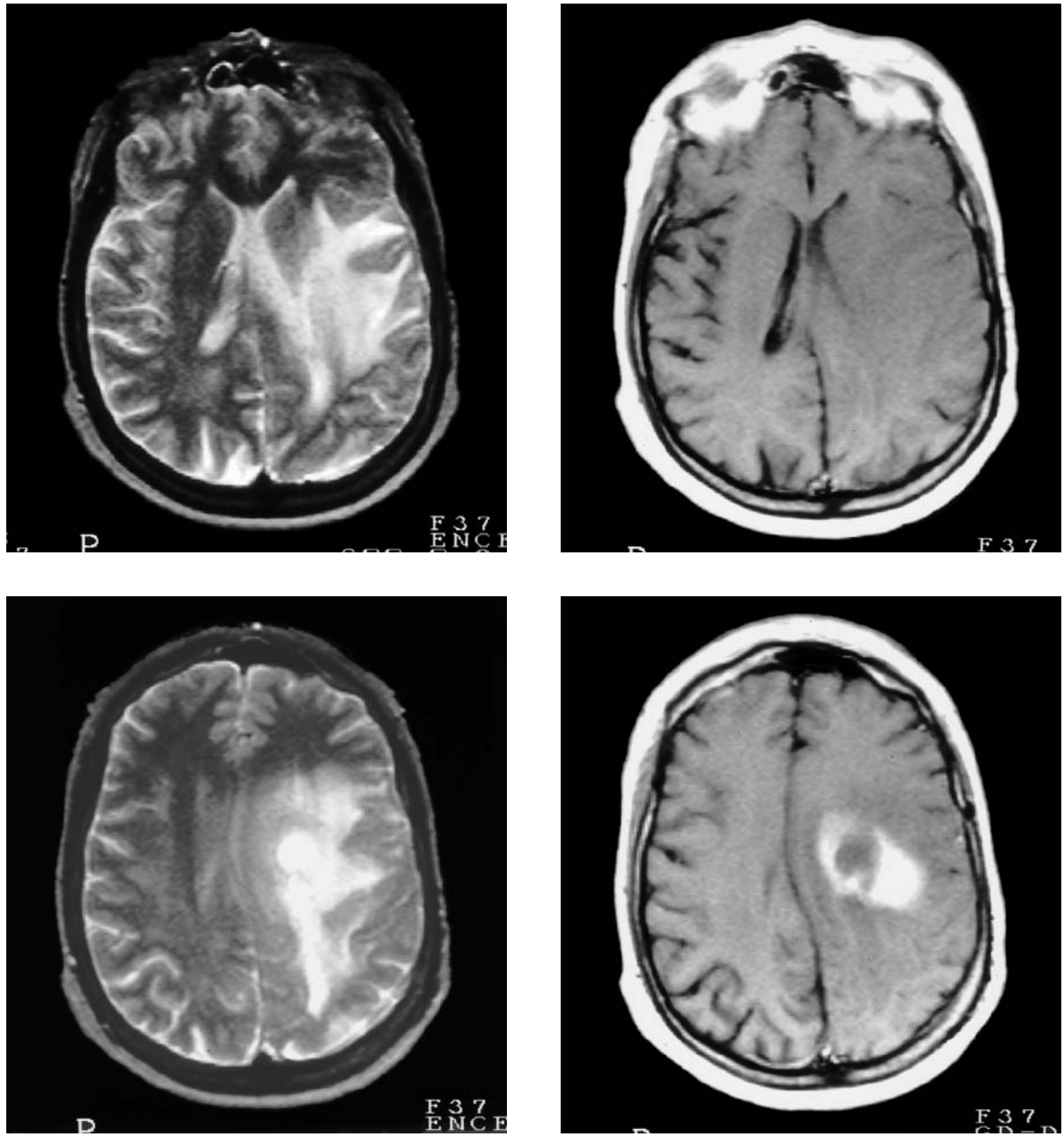


Fig. 3.2 - Vasogenic oedema caused by malignant cavitary glial neoplasm. The MRI study shows extensive oedema involving the white matter surrounding the neoplasm. Note the mass effect upon the lateral cerebral ventricles and the irregular mural contrast enhancement. [**a**), **b**) T2-weighted, **c**) unenhanced T1-weighted, **d**) and T1-weighted MRI following IV gadolinium (Gd) administration].

(Fig. 3.3). It causes the nerve cells to swell and an increase in the permeability of the blood-brain barrier. This type of oedema is both intra- and extracellular and consists of a plasma ultrafiltrate that includes proteins. On imaging these alterations demonstrate peripheral contrast enhancement and mass effect.

Cytotoxic oedema

Cytotoxic oedema is most frequently caused by an ischaemic-hypoxic insult such as preceding cardiopulmonary arrest. Less frequently it may be related to water intoxication, the decompensation syndrome in dialysis patients, di-

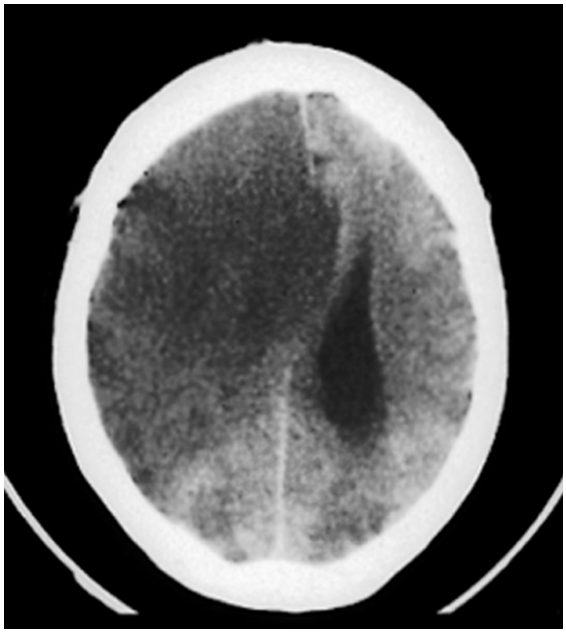


Fig. 3.3 - Hemispheric cerebral infarction. Unenhanced axial CT demonstrates a large hypodense area in the vascular distribution of the right anterior and middle cerebral arteries associated with mass effect.

abetic ketoacidosis, purulent meningitis, severe hypoglycaemia and methanol intoxication.

Brain swelling occurs within all cellular components of the cerebrum (e.g., neurons, glia, ependyma, endothelial cells), in both white and grey matter, with an increase in the total intracellular water content. Neuroradiologically, findings are most frequently observed in the cerebral and cerebellar cortex, the basal ganglia, the hippocampus and the vascular watersheds. The thalami and brainstem tend to be spared.

Mass effect when present results in ventricular compression and the effacement of the CSF spaces (e.g., sulci, basal cisterns). Due to reduced cerebral perfusion, there may be no enhancement after IV contrast medium administration.

Interstitial oedema related to hydrocephalus

This type of oedema is observed in obstructive hydrocephalus and is caused by the transependymal passage of fluid from the ventricles to the periventricular white matter, with consequent interstitial oedema. It is typically symmetric surrounding the anterolateral portion of the lateral ventricles (Fig. 3.4).

The grey matter is normal, and there is no abnormal enhancement after contrast medium administration. These periventricular alterations regress following proper ventricular shunting or spontaneous resolution of the hydrocephalus.

ICH RELATED TO ABNORMAL CSF PHYSIOLOGY

Pseudotumor cerebri

Pseudotumor cerebri is a condition (13) having an undefined pathogenesis that usually affects young, obese patients with or without hypercorticism and menstrual disorders. It is typically observed in females that are otherwise healthy. Electroencephalograms are normal, and the mental status is intact.

Signs and symptoms associated with pseudotumor cerebri include headache, nausea, vomiting and diplopia. Bilateral papilloedema is present, and visual loss is documented in one-third of cases, which becomes permanent in one of eight patients. The diagnosis is determined from lumbar punctures showing an increase in CSF pressure and from neuroradiological studies that exclude the presence of hydrocephalus, mass-forming processes and thrombosis of the dural venous sinuses.

In approximately 36% of cases, CT and MRI are negative; in the remainder, the following findings may be seen: a small ventricular system and a failure to visualize the basal cisterns; small ventricular system with normal visualization of the basal cisterns; empty sella turcica; and enlargement of the sheaths of the optic nerves. With normalization of fluid pressure, the ventricular and periencephalic fluid spaces return to normal.

HYDROCEPHALUS

CSF (3) is secreted by the choroid plexuses, especially those within the lateral cerebral ven-

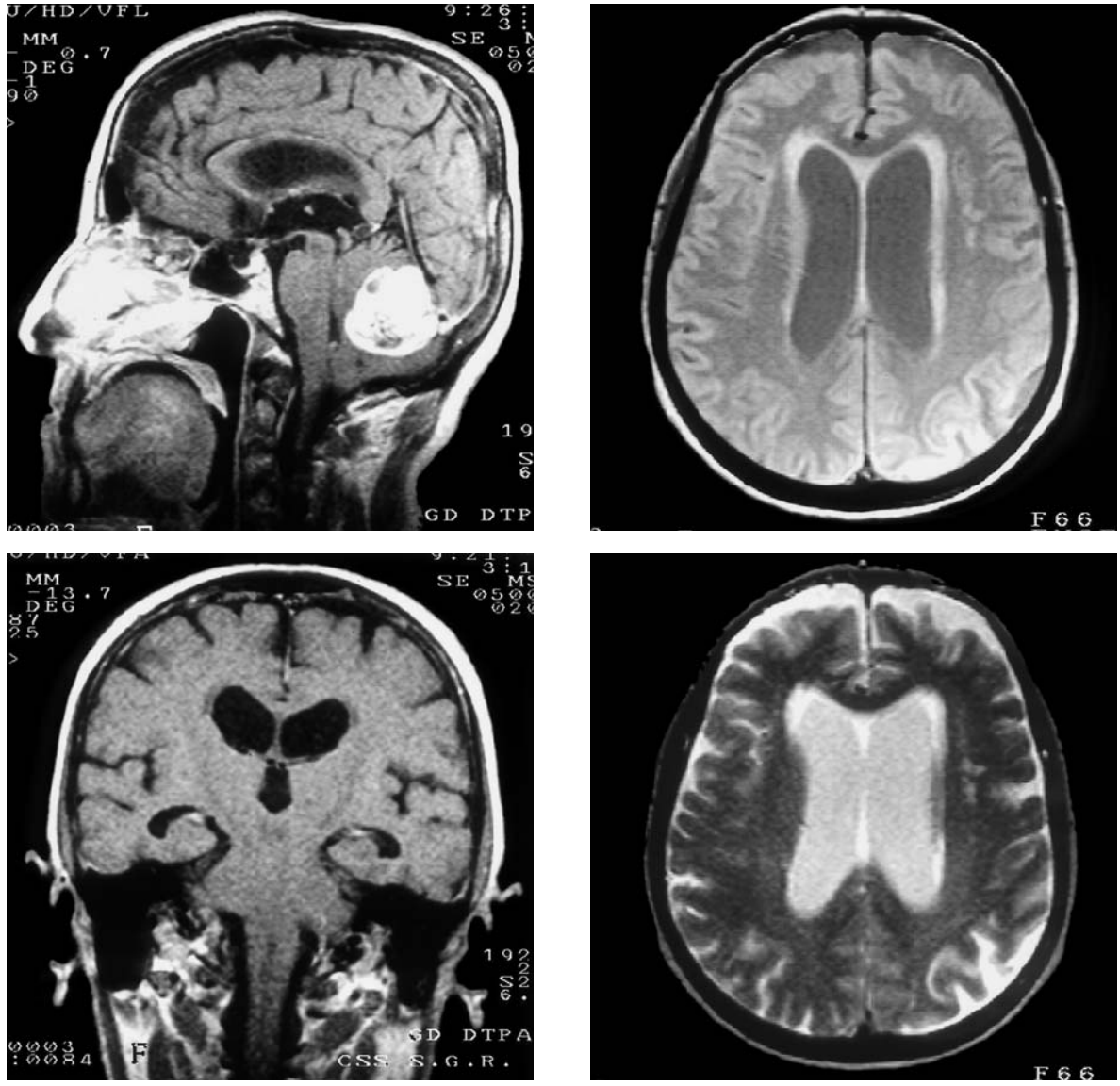


Fig. 3.4 - Neoplasm of the cerebellar vermis with obstructive hydrocephalus. The MRI examination shows a contrast enhancing mass (**a**) likely originating within the cerebellar vermis that compresses the 4th ventricle and has resulted in obstructive hydrocephalus (**b**). The T2-weighted images reveal transependymal extravasation of CSF into the periventricular white matter (**c**, **d**).

tricles. CSF is a clear, colourless fluid containing very few cells (approximately 2 per mm^3) and little protein (normal range: 25-40 mg/100 ml). It has a different ionic composition as compared to plasma, as active secretion mechanisms such as carriers and pumps contribute to its production.

Interposed between the capillary blood of the choroid plexuses and the intraventricular CSF is a blood-fluid barrier, which is permeable to water, oxygen, carbon dioxide and to a lesser degree to electrolytes, but is impermeable to cellular and protein components of the blood. Certain drugs (e.g., acetazolamide, furosemide) and metabolic and respiratory alkalosis reduce CSF production.

Once produced, CSF passes from the lateral ventricles, through the foramina of Monro into the 3rd ventricle and from there through the aqueduct of Sylvius into the 4th ventricle. From the 4th ventricle, the CSF passes through the foramina of Luschka and Magendie, to reach the subarachnoid spaces of the skull base.

Thereafter the CSF passes into the anterior pericerebellar cisterns and surrounds the brainstem. CSF therefore generally follows two pathways, one medial and one lateral.

Along the medial pathway, it reaches the prepontine, interpenducular, suprasellar, chiasmatic and terminal laminar cisterns, until arriving at the subfrontal regions. Then it reaches the superior sagittal sinus, through the interhemispheric spaces. Simultaneously, it flows upward through the pericerebellar, lamina quadrigemina and pericallosal cisterns until reaching the region surrounding the superior sagittal sinus. Along the lateral pathway, the fluid passes from the interpeduncular, prepontine, ambient and suprasellar cisterns into the Sylvian fissures, and from there through the subarachnoid spaces over the cranial convexity.

In these locations over the cranial convexity are the arachnoid villi of the Pacchionian granulations, which are essential for the proper reabsorption of CSF into the venous blood of the dural venous sinuses. This absorption partly depends upon the hydrostatic pressure gradient between the CSF and blood of the dural sinuses. When this gradient is sufficient, the microtubules of the granulations remain open and permit the passage of CSF towards the bloodstream. If, however, the difference in pressure is very high, the tubules close, thus preventing the reabsorption of fluid.

In addition to this reabsorption mechanism, limited absorption apparently takes place along the perineural sheaths of the cranial and spinal nerves, through the surfaces of the neuraxis bordering upon the subarachnoid space and through the ventricular ependyma.

The total volume of CSF within the subarachnoid spaces in normal adults is approximately 120-160 ml, and 30-40 ml is present within the cerebral ventricles. The intraventricular CSF pressure is 712 cm of water, and the lumbar pressure is 8-18 cm of water.

Hydrocephalus: diagnostic morphological aspects

The term hydrocephalus is used to describe any condition in which an abnormal increase in the volume of CSF occurs within the cranium. There are four possible types (1, 8, 11).

In *obstructive* hydrocephalus, there is a blockage to the passage of CSF through the ventricular cavities and outlets, with a dilation of the ventricular spaces proximal to the obstruction. Frequent causes are neoplastic and nonneoplastic mass-forming lesions; another relatively common aetiology is fibrotic adhesions secondary to inflammatory and haemorrhagic processes. The CSF obstruction may occur at the level of the foramina of Monro (e.g., neoplasia, inflammatory processes), the 3rd ventricle (usually neoplasia), the cerebral aqueduct of Sylvius (e.g., congenital atresia, inflammatory stenoses), posthaemorrhagic adhesions, or in the 4th ventricle (e.g., neoplasia, craniocervical malformation, infection or subarachnoid haemorrhage).

From an imaging point of view, obstructive hydrocephalus is characterized by a symmetric dilation of the ventricular system proximal to the point of obstruction. Characteristic is the rounded appearance of the dilation of the frontal horns, with a reduction of the angle between the medial walls of the frontal horns themselves (<100 degrees). If involved in the process, the 3rd and 4th ventricle are also dilated. The basal subarachnoid cisterns are either normal or mildly encroached upon, as are the superficial cerebral sulci, which are either absent or smaller than usual.

Unilateral obstruction of one of the foramina of Monro causes the distension of one lateral ventricle only. Such obstructions may be intermittent if the obstructive process is valvular, with clinical manifestations of headache, nausea and vomiting. Characteristically, these episodes resolve rapidly when and if CSF drainage is restored. On the other hand, if both foramina of Monro are involved, the obstructive hydrocephalus is limited to the lateral ventricles.

An obstruction of the aqueduct of Sylvius causes supratentorial triventricular dilation (i.e., the 3rd ventricle and lateral ventricles). Aqueductal stenosis is a frequent cause of hydrocephalus in infancy and the early stages of childhood. As at this age the cranial sutures are

not yet fused, macrocrania develops. If, however, hydrocephalus develops in late childhood after closure of the sutures, the enlargement of the skull is absent or at most modest. In adults the aqueduct is more frequently compressed by a tumour (e.g., periaqueductal astrocytoma) (Fig. 3.5).

If the 4th ventricle or its outlets are obstructed, all the other parts of the ventricular system are distended. Obstruction of the foramen of Magendie can be caused by a developmental malformation (e.g., Arnold-Chiari malformation [Fig. 3.6], platybasia and basilar invagination, atlantooccipital fusion, etc.) or alternatively a peri- or intraventricular tumour. The foramina of the 4th ventricle can also become non-patent due to a granulomatous ependymitis, caused for example, by the tubercle bacillus. An *entrapped* 4th ventricle refers to a simultaneous obstruction of the aqueduct of Sylvius and the foramina of Luschka and Magendie, with a consequent distension of its cavity (Fig. 3.7).

In decompensated obstructive hydrocephalus, the transependymal passage of fluid is more evident in the anterior-lateral portion of the frontal horns, and on CT has a reduction in periventricular density with regular margins (or an increase of signal on T2-weighted MR images, Fig. 3.4).

In *communicating hydrocephalus* there is a lack of fluid reabsorption due to the thrombosis of the venous sinuses, malfunction of the arachnoid granulations or poor circulation within intracranial subarachnoid spaces due to meningitis, meningeal carcinomatosis or following a subarachnoid haemorrhage. Blood, pus, meningeal metastases or adhesions may mechanically obstruct the fluid circulation path-

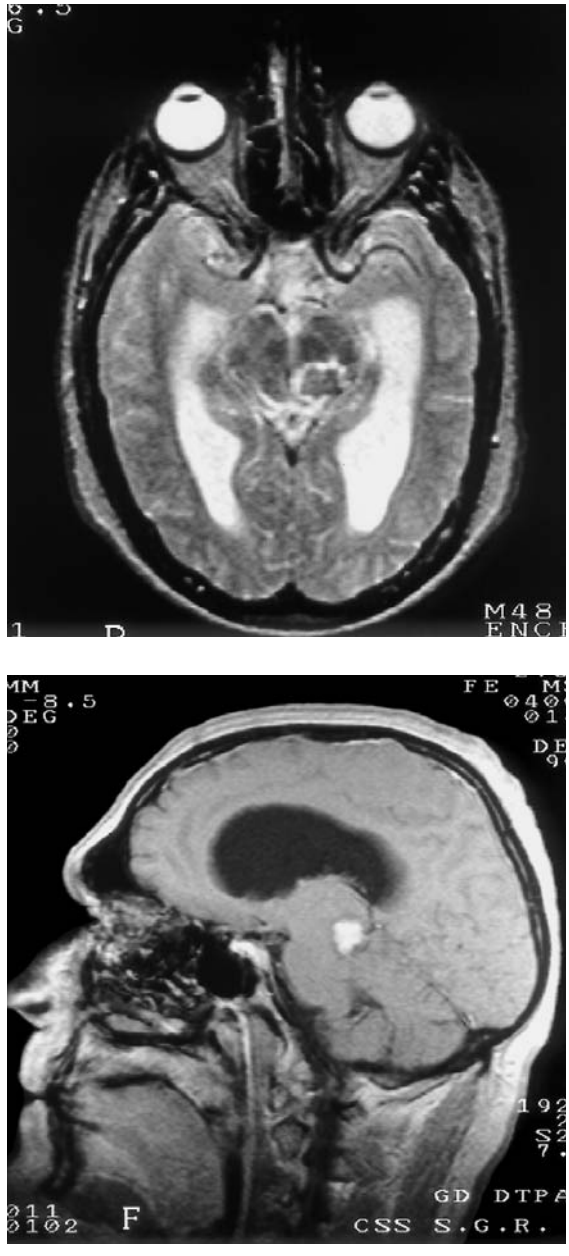
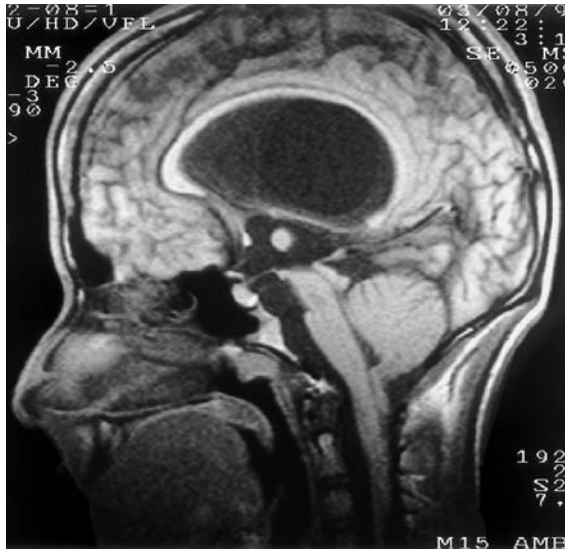
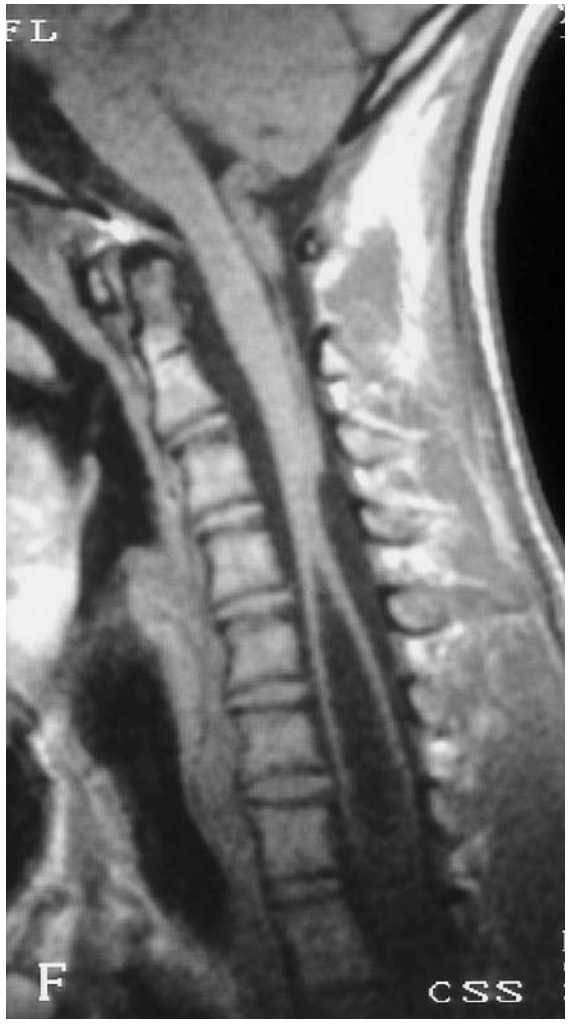


Fig. 3.5 - Mesencephalic tuberculoma with obstructive hydrocephalus. The MRI shows a contrast enhancing mass lesion of the posterolateral midbrain resulting in stenosis of the aqueduct of Sylvius and obstructive hydrocephalus. [**a**) axial T2-weighted and **b**) sagittal T2-weighted MRI; **c**) coronal T1-weighted MRI following IV Gd].



a

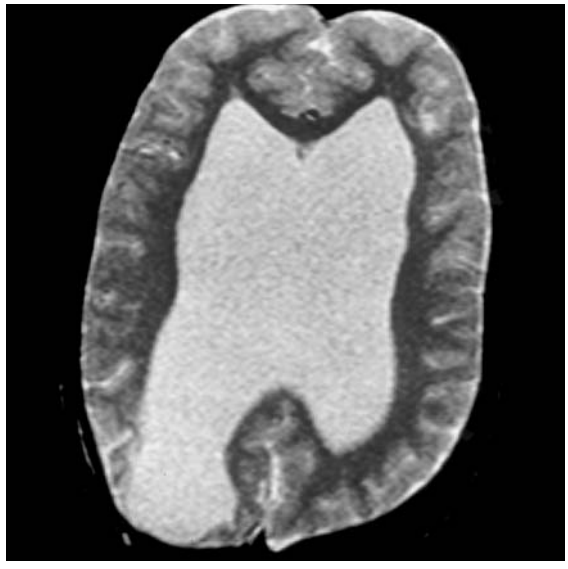


b

ways in the basal subarachnoid cisterns and/or involve the arachnoid granulations. The connection of the cranial subarachnoid space to the spinal subarachnoid space may remain patent.

After a certain time, chronic inadequate fluid reabsorption can produce an enlargement of the ventricles without an attendant increase in fluid pressure (normotensive hydrocephalus). In acute hydrocephalus, the CSF is reabsorbed vicariously by the minor resorption systems, firstly by the transependymal pathway, with the appearance of typical findings of hypodensity on CT and hyperintensity on T2-weighted MRI in the periventricular white matter (Fig. 3.8). This absorption pathway is more permeable than the normal one and permits the passage through the periventricular white matter of even large protein molecules.

In communicating hydrocephalus, the dilation starts from the anterior and temporal horns, followed by the occipital horns and the 3rd ventricle. The 4th ventricle is not necessarily dilated. Occasionally, dilated sulci can also be observed in hydrocephalus associated with a block of the subarachnoid spaces over the cranial convexity.



c

Fig. 3.6 - Type II Arnold Chiari malformation with obstructive hydrocephalus The MRI examination demonstrates cerebellar tonsillar ectopia, fourth ventricular outlet obstruction and hydrocephalus. In addition, there is right occipital encephalomalacic porencephalic cyst and cervicothoracic syringohydromyelia. [**a**) sagittal T1- weighted cranial MRI; **b**) sagittal T1-weighted cervical MRI; **c**) axial T2-weighted cranial MRI].

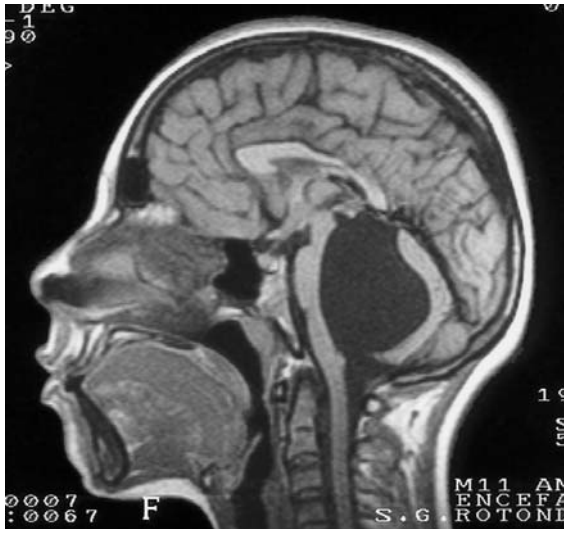


Fig. 3.7 - Trapped 4th ventricle with hydrocephalus. T1-weighted MRI showing a trapped 4th ventricle associated with obstructive hydrocephalus in a patient with tuberculous meningitis.

In *hypersecretory hydrocephalus,* there is an increase in the production of CSF (beyond the normal 0.3-0.4 ml/minute) in the choroid plexuses due to infection or the presence of a choroid plexus papilloma.

Lastly, in *ex-vacuo ventricular enlargement* there is a passive increase in the ventricular and extraventricular CSF spaces without an increase in the intraventricular fluid pressure. This type of ventricular enlargement is due to a generalized atrophy of the brain parenchyma (Fig. 3.9). Generalized atrophy results in a symmetric dilatation of the ventricular system, which may principally involve the lateral ventricles although the 3rd ventricle may also be affected. If there is atrophy of the structures of the posterior fossa, the 4th ventricle will also be passively enlarged. The angle between the frontal horns is always greater than 110 degrees. The basal subarachnoid cisterns and the superficial cerebral sulci can be normal, however they more frequently are widened in synchrony with the overall atrophy.

Functional diagnosis

CT and MRI are excellent techniques for illustrating the morphological characteristics of hydrocephalus as well as the presence of the underlying pathology when this pathological change is a mass. However, both techniques have limitations in their ability to directly demonstrate the obstructing element to CSF circulation resulting from adhesions of inflammatory or haemorrhagic origin.

Radionuclide myelocisternography (12) or myelocisternal-CT with water-soluble contrast medium can be used to study CSF circulation, subject to the intrathecal spinal introduction of the tracer or contrast agent (e.g., 111-In DTPA and Iopamiro 300, respectively) and subsequent serial imaging in order to follow the progress of the agent through the subarachnoid spaces of the cranium.

In normal subjects, the basal subarachnoid cisterns show presence of the tracer or contrast by the first to third hour, and the Sylvian fissures at the fourth to sixth hour. The subarachnoid spaces over the cranial convexity are normally opacified by the 12th hour, and more completely at the 24th hour. In normal subjects, the cerebral ventricles are never opacified. The imaging appearance of the cerebral ventricles in normal subjects depends in part upon the age of the patient. CSF opacification is more rapid in children, typically disappearing by the 24th hour, than in the elderly, in whom opacification over the convexity can persist beyond the 48th hour.

The radioisotopic technique can also be used to study CSF/plasma clearance of the tracer by means of a series of blood samples. In normal subjects, this will give the following haematological activity values as a percentage of the dose injected: 2nd hour: 1.6%; 6th hour: 10%; 24th hour: 33%; 48th hour: 41%.

The study of cerebrospinal fluid circulation and CSF/plasma clearance in normotensive hydrocephalus enables some prognostic indication for the efficacy of the treatment with surgically placed shunts. This shunt operation proves to be effective in clinical practice in patients with normotensive hydrocephalus, in whom myelocisternal scintigraphy shows a constant, early and persistent opacification of the cerebral ventricles after the 48th hour.

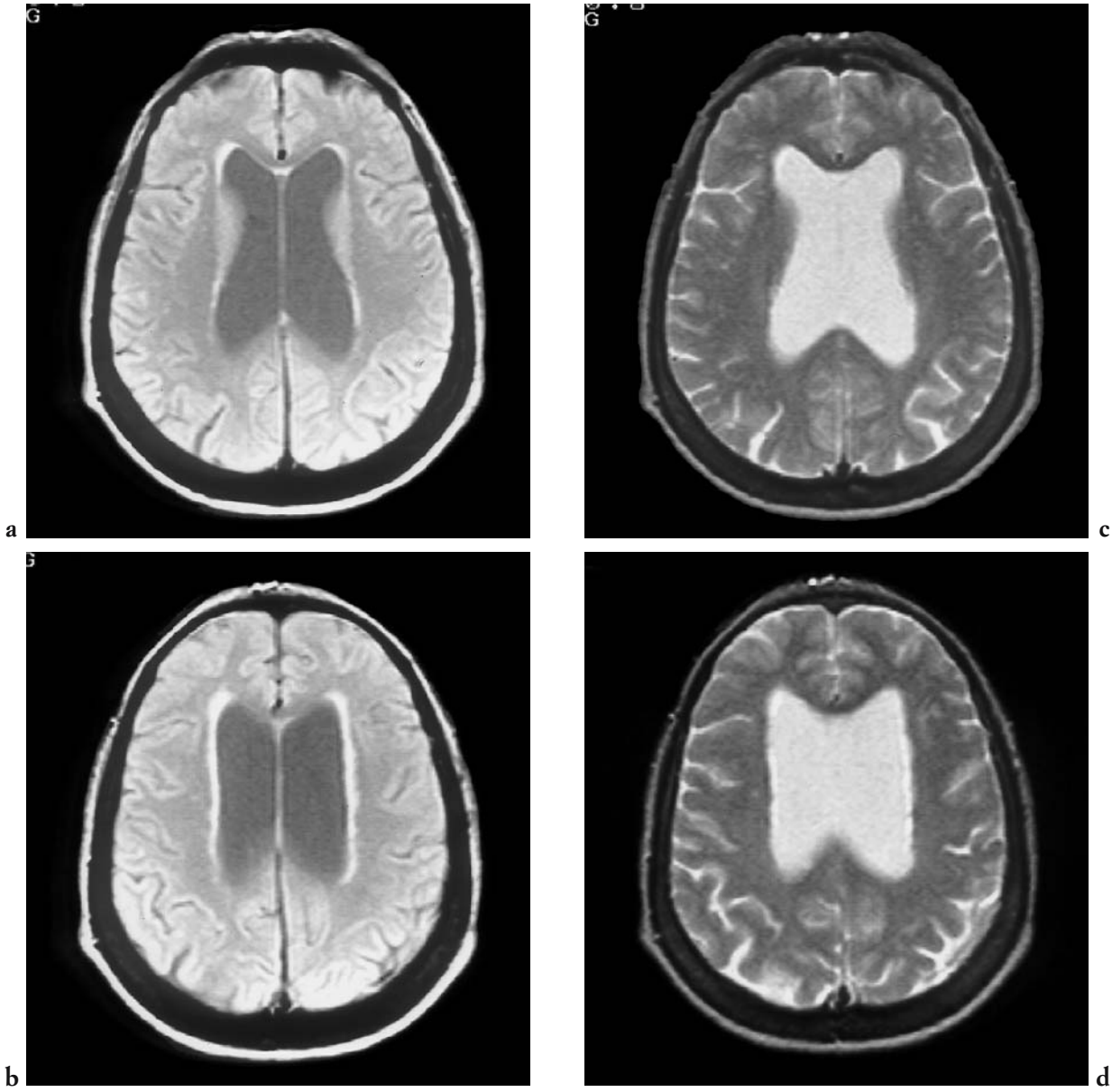


Fig. 3.8 - Normotensive communicating hydrocephalus. The MRI images show a thin margin of hyperintensity representing chronic gliosis surrounding the lateral ventricles in a patient with clinically diagnosed chronic normotensive communicating hydrocephalus, not transependymal extravasation of CSF. [**a**, **b**) proton density-weighted MRI; **c**, **d**) T2-weighted MRI].

Potentially important diagnostic information can also be supplied by prolonged and continuous pressure monitoring of ventricular fluid using a catheter or by Katzman's lumbar infusion test. The latter technique involves the continuous infusion of the lumbar subarachnoid space with a physiological solution at a speed of 0.8 ml/minute; in normal subjects there is an increase in fluid pressure up to a plateau of approximately 20 mm Hg after about 30 minutes. In CSF reabsorption disorders such as hydrocephalus there will be a marked and early increase in CSF pressure values.

If the subarachnoid spaces, typically at a spinal level, are isolated from one another due to a blockage of CSF circulation at some point, the CSF below the blockage will undergo an

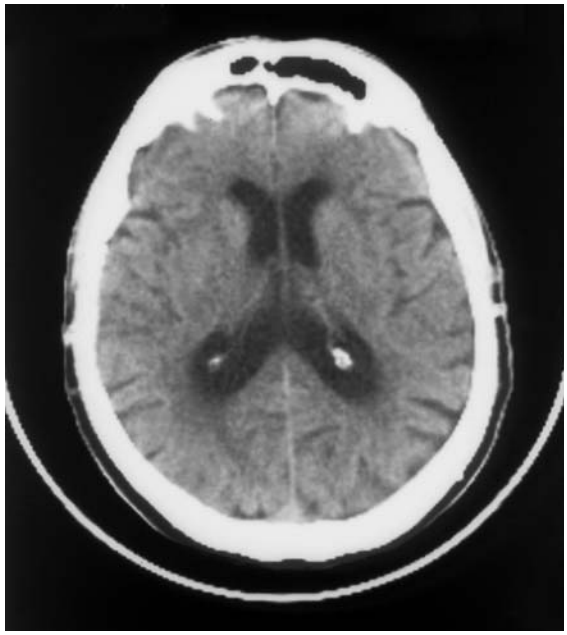


Fig. 3.9 - Ventriculomegaly with passive cerebral atrophy. Axial CT shows passive cerebral atrophy associated with ventriculomegaly.

abnormal increase in cells (i.e., Froin's syndrome), while that above the obstruction will have a normal cell and protein content.

Clinically, the Queckenstedt test (i.e., jugular compression test) (2) is usually sufficient to establish whether there is a partial or total CSF obstruction. With the patient in a lateral decubitus position, a lumbar puncture is performed and fluid pressure is measured. In normal conditions this pressure alters in synchrony with pulse and breathing. In normal subjects CSF pressure alters in a very marked and rapidly reversible manner with abdominal compression. The mechanism for this is via an increase in intraabdominal pressure originating from a temporary blockage of the spinal veins, in turn resulting in an increase in intraspinal fluid pressure; this demonstrates that there is no obstruction in the subarachnoid space at the spinal level.

If this abdominal compression test confirms that the subarachnoid space is not obstructed, and in the absence of intracranial hypertension and an intracranial mass, one can perform the Queckenstedt manoeuvre by compressing both internal jugular veins simultaneously. This will cause an increase in intracranial venous pressure with a resulting increase in intracranial fluid pressure, which in normal conditions will be transmitted along the spinal subarachnoid spaces to the lumbar level of CSF pressure measurement. However, if there is a blockage in the spinal canal or at the cranio-cervical junction, the expected pressure increase will not reach the pressure gauge. If the increase in CSF pressure at the level of the pressure gauge is slow and incomplete in returning to normal once the pressure on the jugular veins has been removed, an incomplete blockage can be assumed.

MRI is also capable of supplying important information on spinal and intracranial fluid spaces and its circulation. In sectors of the subarachnoid space with high pulsatile CSF speeds, the MR signal normally disappears due to the flow void phenomenon. This can typically be observed in the aqueduct of Sylvius, the foramina of Monro, the 3rd ventricle and in the region of the foramen of Magendie.

A circulation blockage with CSF stasis will bring about a disappearance of this phenomenon. Using gradient-echo sequences combined with ECG gating, these phenomena can also be studied dynamically with CSF flow measurements (6).

ICH RELATED TO VASCULAR CAUSES

A third cause of ICH is a relative increase in the amount of blood contained in the cranial cavity. This form of ICH may involve the venous or the arterial system. ICH may have a venous cause when the circulation of the returning blood is obstructed, which can occur in cases of venous thrombosis associated with thrombophlebitis of the dural venous sinuses, or in instances of mediastinal compressive pathology. In this type of ICH, the venous congestion results in a partial inhibition of normal CSF drainage.

Increases in intracranial blood volume may also affect the arterial capillary sector of cerebral circulation. In active vasodilatation, the effect is usually due a local increase in CO_2,

which, irrespective of its origin ultimately results in a vicious circle arising once intracranial hypertension has begun. In passive vasodilatation there is a loss of cerebrovascular autoregulation as a consequence of a systemic acidosis. In this situation, the vessels become passively distended by the systemic blood pressure.

AETIOLOGICAL CAUSES OF INTRACRANIAL HYPERTENSION

Intracerebral tumours represent the most frequent cause of intracranial hypertension. Beyond the size of the mass itself, the principal pathophysiological mechanism involved in ICH production in many neoplasms is oedema. However, not all tumours are equally productive of oedema, the most oedema-inducing being glioblastomas and neoplastic metastases. Low degree gliomas by comparison cause little or no oedema, and when present it remains localized to the immediate area around the lesion. Extracerebral tumours such as meningiomas, on the other hand, may become quite large before causing ICH.

Expanding lesions localized to the subtentorial compartment can lead to intracranial hypertension in part by CSF obstruction. This tends to occur earlier in intraventricular tumours and those of the midline (e.g., medulloblastomas and ependymomas) than in laterally positioned tumours (e.g., neoplasia of the cerebellar hemispheres and the cerebellopontine angle), which typically cause a delayed increase in intracranial pressure.

Intracranial hypertension of a vascular cause is observed in a number of conditions. For example, in arterial hypertension, oedema can also occur as a result of paroxysmal hypertension. Subarachnoid haemorrhages are always accompanied by an initial episode of intracranial hypertension due to a blockage of the CSF reabsorption pathways by the haemorrhage. The subarachnoid blood usually reabsorbs without sequelae; however, if the haemorrhage has been widespread or is a rebleed, subarachnoid adhesions can form with the possible development of a secondary form of hydrocephalus. Intraparenchymal haematomas with perilesional oedema behave as a mass formation and can occasionally result in the onset of ICH. Intracranial pressure usually spontaneously returns to normal with resolution of the haematoma.

Among the types of intracranial hypertension of infectious origin, acute meningitis is typically associated with ICH. Pyogenic cerebral infections are almost always oedema-inducing, and ICH is rarely absent in the acute phase. Viral encephalitis can also be accompanied by considerable cerebral oedema and resultant ICH.

Serious cranial trauma is often associated with varying degrees of ICH, due both to the presence of an intraparenchymal haematoma in parenchymal contusion foci, as well as to related oedema and haemodynamic alterations. In fact, in the initial phase that follows trauma, an increase in cerebral blood volume plays an important role in the pathological increase in intracranial pressure.

ICH can also be observed in cases of intoxication from carbon dioxide, lead, arsenic or following allergic reactions.

In summary, while there are many potential causes of ICH, the majority are due to disorders of fluid dynamics that can be primarily attributed to mass-forming processes as well as to obstructions to CSF circulation due to haemorrhagic, infectious or neoplastic involvement.

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