been reported as a rare complication of brain tumour. However, symptoms of Parkinsonism do result, it is not uncommon to make an incorrect diagnosis of Parkinson's disease. However, symptoms of increased intracranial pressure or mental change and signs of cortico-spinal tract and sensory involvement eventually develop, suggesting the diagnosis of tumour. Any combination of Parkinsonian signs may be present, but most authors reported contralateral static tremor and rigidity.

The exact aetiology of the development of the Parkinsonian syndrome in our patient is not well understood and several mechanisms can be postulated: (a) mechanical pressure on basal ganglion nuclei could be caused directly by the tumour; (b) indirectly by torsion or compression of midbrain and tentorial herniation; (c) the deep situated glioma may directly involve the basal ganglia.

In this patient, the assumption of a causal relationship between Parkinsonism and the tumour is based on the lack of any associated precipitating factors (such as ingestion of drugs or poisoning) and the onset of tremor shortly after the contralateral hemiparesis had appeared. The mechanism by which Parkinsonian symptoms are produced contralaterally to cortico-spinal tract symptoms, may be explained by the medial localisation of the tumour and is due probably also to the direct involvement of the left internal capsule whose normal functioning would be essential in order for tremor and rigidity may appear.

Early recognition of an intracranial tumour as a cause for Parkinsonism is therefore very important if further neurological deficit is to be prevented. It would seem desirable to obtain a CT scan in any Parkinsonian patient with other associated neurologic manifestations, and may also be indicated in patients with hemiparkinsonian symptoms, essentially with tremor.

Huge epithelium-lined cyst: report of two cases

Sir: Epithelium-lined cysts of the central nervous system have been reported with such various names as “neuroepithelial cyst, ependymal cyst, parapressal cyst, choroid plexus cyst, and colloid cyst.” However, their exact origin is uncertain and the pathogenesis of these cysts is still controversial.

The following case reports describe two cases of huge epithelium-lined cysts in the posterior cranial fossa with extension to the middle cranial fossa. Although a review of the literature disclosed many reports concerning the location of the cysts, such cases as described here appear never to have been reported. We present details of these cases, with clinicopathological features of these cystic lesions.

Case 1 was a 2-month-old male infant with an increase in head circumference and horizontal nystagmus. He was admitted to our department for diagnostic workup and treatment. Neurological examination revealed only horizontal nystagmus. A computed tomography (CT) scan revealed a huge low density area in the posterior cranial fossa extending to the bilateral middle cranial fossa (fig 1). Metrizamide CT cisternography revealed no communication between the cyst and the ventricular system. Vertebral angiography demonstrated marked bowing and displacement of the basilar artery and an avascular area between clivus and pons. Suboccipital craniectomy was performed; during the operation, the cyst wall was exposed and was found to consist of tough membrane with many capillaries. Facial and acoustic nerves were stretched posteriorly over the cyst wall. The right cerebellar hemisphere was displaced to the left.

The cyst wall was widely opened and partially removed to establish a communication with the subarachnoid space. The cyst cavity was found to extend from the preoptic region to the bilateral middle cranial fossa beyond the incisura of the tent. No abnormalities of the cerebellum or brain stem were noticed, and there was no communication between the cyst and the ventricular system.

A ventriculo-peritoneal shunt was performed 8 weeks after the craniectomy, because hydrocephalus did not resolve following the partial removal of the cyst. The fluid obtained from the cyst contained a protein level of less than 0.1 g/l. Microscopic examination revealed that the cyst wall consisted of a single layer of ciliated cuboidal and columnar epithelial cells with an underlying basement membrane. The wall was supported in part by connective tissue (fig 2). The postoperative course was uneventful, and he was discharged 2 weeks after the second operation.

Case 2 was a 10-month-old female baby with arrested development. CT scan demonstrated a huge low density area in the posterior cranial fossa which extended to the right middle cranial fossa. CT cisternography did not show communication between the cyst and the ventricular system. Craniectomy was

References


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Fig 1 Plain computed tomography (CT) scan showing a huge low density area in the posterior cranial fossa extending to the squiggle middle cranial fossa.

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performed and the lucent cyst wall was exposed. The right cerebellar hemisphere was pressed to the left by the cyst, which was found to extend to the right middle cranial fossa and to the prepontine region. Subtotal resection of the cyst wall, in order to establish a communication with the subarachnoid space, was performed in addition to a ventriculo-peritoneal shunt. The protein level of the fluid obtained at the operation was 0.2 g/l. Histology revealed a single layer of ciliated columnar epithelial cells which were invested by a basement membrane. This appearance was consistent with a neuroepithelial cyst. Although the patient made an uneventful recovery, she unfortunately developed pneumonia 3 weeks after the operation.

Benign intracranial cysts of childhood have come to the attention of neurosurgeons since CT scan became available. The surgical importance of these cysts including arachnoid cysts and epithelium-lined cysts has been stressed because of their benign nature, their tendency to behave as mass lesions, and their amenability to surgical cure. Among these cysts, epithelium-lined cysts are familiar to neurosurgeons as ependymal cyst; however, the origin and the pathogenesis of these cysts has not often been elucidated. Both cases described in this communication showed a huge low density area in the posterior cranial fossa which extended to the middle cranial fossa on CT examination. They were, therefore, thought to be a preptontine type of arachnoid cyst as described by Little et al. Histologically, however, the cyst wall of both cases was lined by a single layer of ciliated columnar or cuboidal epithelial cells. Such an appearance and absence of arachnoidal cells exclude the possibility of arachnoid cysts in these cases. The pathogenesis of epithelium-lined cyst is still obscure, and has been a matter of speculation. The origin of the cysts has been considered to be neuroectodermal. Shuangahoti et al. proposed a close relationship between epithelium-lined cyst and the choroid plexus. His hypothesis has been generally supported. Dandy reported a cyst in the cerebellar angle which communicated with the fourth ventricle. He considered the cyst to have been formed by an extra-ventricular protrusion of the ependyma of the fourth ventricle. An epithelium-lined cyst in the cerebellar vermis has been described by Hasegawa et al., which was also considered to be originated in embryonic neuroepithelium. Contrary to these reports, the hypothesis was advanced that the epithelium-lined cyst is derived from endodermal origin by Hirano et al. Based upon the ultrastructural studies of colloid cysts and an epithelial cyst, they reported that the cells which constitute the cyst wall are apparently different from those of choroid plexus or ependyma, and that their features are reminiscent of the epithelium of the upper respiratory tract. These cysts, therefore, may be derived from endoderm.

Recently, Hirai et al. presented microscopic evidence to support this hypothesis.

It is generally accepted that basement membrane is not commonly found in the ependyma, and that the cilia can be observed only in the ependyma in the central nervous system. On the other hand, the cells possessing both basement membrane and cilia are commonly found in the intestine and the respiratory tract. The fine structure of the upper respiratory tract is most reminiscent of that of the epithelial cyst. It has been believed that the cysts derived from choroid plexus arise in the midline, and that a paramedian location is evidence against an origin from the choroid plexus. In the present cases, the location of the cysts is unique in that they were located in the anterior aspect of the pons with extension to the middle cranial fossa. It might thus be possible that these cysts were derived from endodermal tissue rather than from neuroepithelium. The first patient had not shown any disturbances of cranial nerves, although the facial and the acoustic nerves were stretched by the cyst. This implies that these nerves had been elongated gradually in the early embryonic stage accompanied with the growth of the cyst.

Although asymptomatic cases may not require surgical treatment, three types of surgical treatment have been adopted for the symptomatic lesions as follows; repeated aspiration, excision of the cyst or a procedure causing the cyst cavity to communicate with the subarachnoid space or the ventricular system, and drainage of the cyst cavity into the peritoneal space. In some cases, cyst-peritoneal shunt alone is effective without craniotomy, which might be less risky. In the case of shunting, however, it is sometimes necessary to replace the shunt at a later time and there is a possibility of the shunt malfunctioning. According to our experience, therefore, we consider that the partial removal of the cyst might be useful and can be expected to cure permanently even though it is more risky than a shunting operation. In our two cases, unfortunately, ventriculo-peritoneal shunt was necessary along with the partial removal of the cyst. It also seems to be reasonable to add ventriculo-peritoneal shunt to the partial removal of the cyst even after establishing a communication with the subarachnoid space, because hydrocephalus often recurs in most of such cases presumably due to the malabsorption of CSF.

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Syncope and sudden unexpected death attributed to carbamazepine in a 20-year-old epileptic

Sir: Carbamazepine is known to cause atrioventricular conduction block but has not yet been implicated in sudden unexpected death in epilepsy. We report one such death which, we suggest, may have resulted from ventricular arrhythmia induced by carbamazepine.

A 20-year-old student teacher presented for routine review with a 7 year history of intractable petit-mal but with well controlled grand-mal epilepsy. She was taking valproate 400 mg tds and parathion 300 mg bd. Troxidone and ethosuximide had failed to control her petit-mal. A fresh history elicited features of temporal lobe epilepsy coinciding with her absences, namely deja-vu, jamais-vu, and amnesic spells. There were prodromal symptoms (strange epigastric sensations, extreme pallor) and postictal features (headache, unsteadiness). A review of her ECGs showed excess slow wave activity in the right frontal region in 1978 and in repeat ECGs in 1979, 1980 with a normal CT brain scan. The diagnosis was changed to temporal lobe epilepsy. Parathion was withdrawn and carbamazepine was started 200 mg bd. After 3 days, her mother reported that she began to suffer syncopal episodes within a few hours of starting treatment. These were different from any previous seizures, lasted a few minutes, were preceded by lightheadedness, and were followed by total recovery. Her family doctor visited and witnessed a typical attack. She became pulseless with no heart sounds. Resuscitation was unsuccessful. Necropsy revealed normal heart, brain and lungs, except for aspiration of stomach contents. There was no evidence of an overdose, anticonvulsant levels of the previous month were within therapeutic range and microscopic examination of the heart was normal.

The annual mortality rate in patients with epilepsy has been calculated as 7·8 per 100,000 population. This is three and half times that to be expected in all age groups up to the age of 50 years.1 Thirteen per cent of these are sudden and unexpected and sometimes, but not always, occur after a seizure. This incidence has proved remarkably constant over a 50 year period,2 3 despite advances in treatment, and affects 1:500 to 1000 epileptics.4 This population has been studied closely,5 and has been found to have a mean age of 33 years, and a seizure frequency of less than 1 a month. Their anticonvulsant levels are almost always below the therapeutic range and are often absent.4 5 They have no other medical history. Various reasons have been advanced for this phenomenon: autonomic collapse,8 disruption of the cardiac and respiratory centres in the brain stem,7 pulmonary oedema8 and cardiac arrhythmias.9

Autonomic disturbance is known to occur during a fit; tachycardia, a rise in blood pressure, ventricular tachycardia and both major and minor ischaemic changes on the ECG have been documented.10 Pulmonary oedema following a fit has been well recognised for many years11 and as early as 1910, this was described as a “typically present” post-mortem finding in epileptics, and as a frequent and dangerous condition following even single grand mal seizures.3 More recently,12 recurrent post-ictal pulmonary oedema has been reported in one patient, following three separate seizures, and a post-mortem study of eight young epileptics who died suddenly, revealed pulmonary oedema13 in all cases, with no myocardial lesion. By way of contrast, another post-mortem study of nine such patients14 showed myocardial changes reminiscent of the lesion of ischaemic cardiomyopathy, but no pulmonary oedema. In both studies, anticonvulsant levels were low or absent. Experimental evidence shows that lesions of the hypothalamus can produce pulmonary oedema13 14 and that this is mediated by the sympathetic outflow.15 Furthermore, electrical stimulation of the hypothalamus can produce ventricular tachycardia and can cause ischaemic damage to myocardium similar to that caused by the injection of catecholamines.16 17 It has, therefore, been postulated that hypothalamic injury initiates an adrenergic discharge, resulting in death by pulmonary oedema or ventricular arrhythmias. A change in permeability of the pulmonary vasculature is also thought to occur15 16 but no mechanism has yet been proposed.

Our patient, however, had no pulmonary or cardiac lesion, although a latent electrophysiological conduction abnormality cannot be excluded. Her anticonvulsant levels were within the therapeutic range one month before her death, and she had not been taken overdosage of carbamazepine, nor of any other drug. Abrupt withdrawal of parathion is not associated with serious adverse effects, such as autonomic epilepsy,19 20 although withdrawal of a related oxazolidine, trimethadione may precipitate fits.21 However, carbamazepine is known to affect the myocardium in pharmacological studies and to cause Stokes-Adams attacks and conduction block in clinical practice.

Carbamazepine is a group IA antiarrhythmic which blocks the fast inward sodium current during the depolarisation of the cardiac membrane.22 It increases atrioventricular conduction time in vivo and depresses ventricular automaticity in vitro by reducing the rate of phase 4 depolarisation.23 There are nine reported cases of carbamazepine causing atrioventricular conduction block or bradycardia when taken in therapeutic dosage.24-29 Stokes-Adams attacks occurred in two patients,24 25 complete heart block in five,24 26-28 and bradycardia in two other patients.29 30 In all cases, sinus rhythm was re-established after withdrawal of the drug. In the five cases which were specially monitored, the conduction block reappeared on restarting carbamazepine.24 25 28 29 Six of the nine cases had previous ECGs available for analysis; left anterior hemiblock was present in two cases and no abnormality in three cases. There appeared to be no relation between these pre-morbid traces, the severity of the later conduction block or the dose of carbamazepine. In fact, a daily dose of only 200 mg produced Stokes-Adams attack in one patient with a previously normal ECG.24 Nonetheless, it has reasonably been suggested that a latent defective conduction system might be a necessary pre-requisite for the induction of atrio-ventricular block.26 Some support for this comes from a small pilot study on patients with pre-existing block whose ventricular standstill time, or pre-automatic pause, was significantly prolonged when carbamazepine was administered. It should also be noted that heart block may intervene after many years of carbamazepine therapy24 and that it may last for three or four days.31 It has thus been