Ependymal lined paraventricular cerebral cysts; a report of three cases

D. C. BOUCH, I. MITCHELL, AND A. F. J. MALONEY

From the Neuropathology Laboratory, Department of Pathology, University of Edinburgh and Milesmark Hospital, Dunfermline

SUMMARY Three cases are reported, each with a benign ependymal-lined cyst which produced clinical signs and symptoms simulating cerebrovascular disease or cerebral neoplasm. The pathological features are described and their histogenesis discussed.

Intracranial cysts lined by ependyma which are large enough to give rise to symptoms are rare. Virtually all references to ependymal lined cysts in the literature relate to the colloid or para-physseal cyst of the third ventricle. This communication describes three cases in each of which a large cyst with watery content was found in the centrum semi-ovale. In no case could communication with the ventricular system be demonstrated. All presented with severe neurological signs and symptoms. None was diagnosed during life. Two were thought to have a neoplasm and the third was considered to suffer from cerebrovascular disease. As the pathological description will demonstrate, the cysts were non-neoplastic, probably developmental in origin, and should have been amenable to surgery.

CASE 1

(NP.871) (RJE.33/38) (Dr. A. C. P. Campbell and Dr. I. W. G. Hill) This 64 year old woman presented in January 1938, having experienced headaches and a feeling of pressure on the top of her head for one year. Over the last two months there was a gradual deterioration of memory and mental faculties with the onset of drowsiness and confusion. Terminally, her condition deteriorated rapidly necessitating her admission to hospital where she became comatose, dying two weeks later. Details of clinical investigations are lacking but she was considered to have sustained cerebral thrombosis.

Post mortem examination showed that the immediate cause of death was a massive pulmonary embolus with the main pathological features confined to the brain.

BRAIN Macro The brain showed swelling of the right frontal lobe, flattening of the overlying gyri, and shift of the midline structures to the left. Occupying most of the central white matter of the right frontal lobe was a unilocular cyst (Fig. 1) compressing, but not communicating with, the lateral ventricle and displacing the anterior corpus striatum downwards. The cyst, which measured 6.0 × 4.0 cm in

FIG. 1. Case 1. Coronal slice of cerebrum at level of optic chiasma showing a large, smooth-surfaced, unilocular cyst in the white matter of the right frontal lobe. Note compression and displacement of the ventricular system and rotation of the right basal ganglia.
maximum diameter in the coronal plane and contained clear watery fluid, was separated from the right lateral ventricle by white matter 0.1 cm thick and from the subarachnoid space over the frontal lobe convexity by white matter and cortex 0.8 cm thick. Its wall was smooth and thin.

The arteries were healthy.

Micro The cyst wall was composed of white matter lined on most of its inner aspect by a single layer of low columnar or cubical epithelial cells, some of which were ciliated (Fig. 2) and contained blepharo-plasts. In one part of the wall, beneath the epithelial lining, there was a small collection of tubules and canaliculi lined by similar, but flattened cells (Fig. 3). The white matter showed minimal gliosis particularly from those areas denuded of epithelium. There was no evidence of haemorrhage, haemosiderin, or neoplasm.

COMMENT It was considered that this cyst was simple, lined by ependyma, and probably of developmental origin.

CASE 2

(NPA.115/71) This 60 year old woman presented in April having noticed weakness of her left arm and dragging of her left leg for one year. This later became associated with minor left-sided sensory changes, slurring of speech, increased urinary fre-
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FIG. 4. Case 2. Depressed area of cortex (S) in the posterior part of the right frontal convexity which overlay the partially collapsed intracerebral cyst seen in Fig. 5. The bridging arachnoid mater has been removed. ×0·5.

quency, and occasional incontinence. Over the last three months intermittent frontal headaches had become a prominent feature along with some difficulty in recalling recent events. There was no history of head injury.

After hospital admission examination confirmed minimal left-sided weakness and also revealed increased tendon reflexes in the left upper limb, a slightly spastic gait, dysarthria, mental slowness, lethargy, and poor concentration. There was no abnormality of the cardiovascular system and the fundi were normal. It was originally thought that she had an intracerebral space-occupying lesion but plain skull radiographs, electroencephalogram, lumbar cerebrospinal fluid, and a cerebral isotope scan were normal. Other system investigations revealed equivocal thyroid function tests; the 48 hour uptake of $^{131}$iodine was consistent with, but not diagnostic of, hypothyroidism; the serum protein-bound iodine was normal. By now the only remaining clinical abnormalities were mental slowness and lethargy and treatment with thyroxin was started. There was marked improvement in her condition. She became alert and active and was allowed home after two weeks' further observation.

Three days after discharge from hospital, left-sided weakness and headaches recurred. Her condition rapidly deteriorated and she was readmitted to hospital 12 hours later in coma. The left limbs were flaccid, the right spastic, and both plantar responses were extensor. Both pupils were fixed, the right dilated and the left constricted. There was no papilloedema. Her condition continued to deteriorate and she died 12 hours after readmission.

Post mortem examination revealed pathological changes in the brain only.

BRAIN Macro The leptomeninges appeared healthy except for slight brownish discolouration in relation to a sunken area of cortex bridged over by arachnoid mater and measuring $6·0 \times 5·5$ cm across and $2·0$ cm deep located in the right frontoparietal region close to the falx (Fig. 4). It appeared to have...
resulted from collapse of an underlying intracerebral cyst, a tiny opening being seen in the floor through which clear fluid issued. Section of the brain showed a large, partially collapsed unilocular cyst situated in the white matter of the supramedial quadrant of the posterior aspect of the right frontal lobe (Fig. 5). This cyst communicated with the subarachnoid space via the small opening already noted in the cortex but nowhere did it communicate with the ventricular system. The cyst measured 6.0 × 4.0 cm in the coronal plane at the level of the mamillary bodies and had a thin wall with a smooth glistening lining. The ventricular system was compressed and displaced from right to left with the right basal ganglia rotated downwards and laterally. The cyst had extended into the convoluted white matter and, bounded by cortex, had crossed the midline under the faix. The arteries were healthy.

**Micro** The wall of the cyst was lined internally by an incomplete single layer of non-ciliated, flattened, or cubical epithelial cells lying upon slightly gliotic white matter. Blepharoplasts were not identified with certainty. In one area an additional thin layer of acellular collagen was interposed (Fig. 6). There was no evidence of haemorrhage, haemosiderin, or tumour.

**COMMENT** This cyst resembles that described in case 1 but has partially collapsed after rupture into the subarachnoid space through attenuated cortex. It is uncertain when the wall ruptured.

**CASE 3**

(NPA.23/72) This 52 year old woman presented in January 1972 having been essentially well until seven months before when she began to feel lost and confused. After an initial slight improvement her condition deteriorated with the development of headaches and a right hemiparesis. Later, increasing drowsiness and a Parkinsonian type of tremor of the right limbs became apparent.

On admission to hospital, examination showed that her hemiparesis was accompanied by minor sensory deficits and there was a right homonymous hemianopia. Lumbar cerebrospinal fluid and plain radiographs of the skull and chest were normal. An electroencephalogram indicated a left cerebral space-occupying mass which appeared to be lying in the posterior frontal region.

Craniotomy and careful needling of the left posterior frontal region failed to identify any lesion. She failed to recover consciousness after operation and gradually became comatose, remaining so until her death two weeks later.

Post mortem examination showed that death was the result of brain swelling and pulmonary oedema. The major pathological lesions were in the brain.

**BRAIN Macro** Operative needle tracks were present in the left frontal lobe. There was generalized swelling of both cerebral hemispheres, particularly the left, with associated uncal herniation and shift of midline structures to the right. Occupying most of the white matter of the posterior half of the left cerebral hemisphere was a large unilocular cyst measuring 4.0 × 3.0 cm in maximum diameter in the coronal plane (Fig. 7). This cyst had compressed the left lateral ventricle and was separated from it by a layer of translucent white matter which was stretched.
to 0.1 cm in thickness and from the subarachnoid space by white matter and cortex thinned to 0.5 cm in the parasagittal area of the left parietal lobe. The arteries were free from disease.

Micro The cyst wall was formed of white matter lined internally by a single layer of epithelial cells some of which were columnar and ciliated (Fig. 8) and contained blepharoplasts, while others were low, cubical, or flattened. In areas the epithelial lining was lost and there was minimal gliosis. There was no evidence of tumour, haemorrhage or haemosiderin.

COMMENT This case is virtually identical with case 1.

DISCUSSION

In each of the three cases described a large unilocular cyst was located entirely within the white matter of one cerebral hemisphere close to, but not communicating with, the adjacent lateral ventricle.

In cases 1 and 3 the cysts were lined in some places by tall columnar, ciliated epithelium con-
taining blepharoplasts indistinguishable from ependyma. In case 2 the epithelium was essentially similar, with the exception that cilia were not identified. Nevertheless, we consider on morphological grounds that all three cysts were lined by ependyma.

There are few references to ependymal-lined cysts in the literature and the majority of these are concerned with the colloid cyst of the third ventricle (Kolin, 1970; Sakata Yamada, Hoshino, Imamura, Matsui, and Takahashi, 1970). Only Jakubiak, Dunsmore, and Beckett (1968) and Argyopoulos and Heppner (1970) have described histologically proven, ependymal-lined cysts morphologically similar to those found in our cases. A recent paper by Harrison (1971) describes 14 cases of hydrocephalus in children, which were associated with arachnoid cysts; five of these were lined by ependyma. Jakubiak et al. (1968) reported four cases of supratentorial brain cysts: two were lined by ependyma; one of these lay within the white matter of the left frontal lobe and did not communicate with the ventricular system or the subarachnoid space, the other was situated in the subarachnoid space over the left parietal lobe and did not communicate with the ventricular system; the remaining two cases were described as arachnoid cysts and were lined by fibrous tissue. Argyopoulos and Heppner (1970) described a single case of an ependymal-lined cyst within the white matter of the right temporoparietal region. It did not communicate with the ventricular system or the subarachnoid space; there were also several similar but smaller associated cysts.

Benign intracerebral cysts have been described in previous papers; the majority of the cysts were considered to be examples of porencephaly (Pendergrass and Perryman, 1946; Drew and Grant, 1948) with a much smaller number either not thought to be porencephalic (Handa and Bucy, 1956) or not classified (Miller, 1952). These reports were mainly concerned with diagnosis and treatment; little attention was paid to the actual structure of the cysts and the histological appearances were not described.

From a study of the macroscopical reports the single case described by Miller and two of the three cases described by Handa and Bucy seem to resemble our cases closely, but in the absence of histological details, no final decision can be reached.

Porencephaly was originally described by Heschl (1859) as a defect in the cerebrum allowing communication between the ventricular system and the subarachnoid space. The meaning of the term porencephaly has been modified to include almost any type of cerebral cyst, not obviously secondarily acquired whether or not it communicates with the ventricular system, the subarachnoid space, or both. The application of such terms as pseudoporencephaly and closed porencephaly have merely added to the confusion. We think that Heschl's original description should be retained and therefore do not consider any of the cysts in our cases to be examples of true porencephaly. Although in case 2 the cyst did communicate with the subarachnoid space, we believe this followed rupture of its attenuated wall.

Jakubiak et al. (1968) considered that the ependymal-lined cysts in their two cases arose during embryological development of the brain, being 'developmental defects in which there is a disorder in the formation of the mantle layer leading to displacement of the ependyma into the subarachnoid space. Isolated and pinched-off immature lining cells give origin to these cysts'. In support of this view they emphasized that: (1) the cysts were lined in part by ciliated ependyma and they contended that ciliated ependyma is only found in the ventricles of normal, newborn, or foetal brains; (2) the indentations, inclusion cysts, and diverticula lined by similar, but sometimes flattened, epithelium found close to the cyst walls, may represent immature choroid plexus; (3) groups of ependymal cells and small ependymal-lined canals are often found close to the occipital horns of normal lateral ventricles. Russell (1950), however, states that cilia may persist on the ventricular ependyma into adult life, an observation that we have confirmed, so that the mere presence of ciliated epithelium in the cyst wall does not alone support a developmental origin. Of greater import is the fact that ependyma does not regenerate after being damaged or lost from a previously normal ventricular wall as a result of hydrocephalus (Russell, 1949). It would therefore seem improbable that a secondarily acquired cystic lesion in white matter could acquire de novo an ependy-
The subarachnoid space.

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Ependymal lining, ciliated or non-ciliated, especially if the cyst did not communicate with the ventricle in any way. For this reason, and in view of the absence of tumour, haemorrhage, or significant gliosis from the cyst walls, we consider that in each of the three cases now presented the cyst was of developmental origin.

Ependymal-lined tubules, similar to those described by Jakubiak et al. were found in case 1 close to the cyst wall but these we consider to be extensions. It is of interest that clinical features did not develop until late in life. It is not possible to state how large the cysts were during the latent period or what caused them to increase in size. Ependyma was so readily found in sections taken from widely separated areas of the wall in each case that the cysts, while still symptomless, must presumably have been of an appreciable size. No trace of choroid plexus was found in any of the cysts. Unfortunately, the fluid was not examined biochemically. Jakubiak et al. (1968) reported that the protein content of the cyst fluid in one of their cases was 14 times greater than the protein content of the cerebrospinal fluid and suggested that such a level of protein would tend to draw in fluid from the surrounding tissues and cause enlargement of the cysts. They postulated a direct secretory activity for the lining of ependymal cells but this does not explain why the protein secretion should apparently have been delayed for many years. The finding of an incomplete ependymal lining upon slightly gliotic white matter in each of our cases is reminiscent of the histological appearance of the ventricular wall in hydrocephalus. The average clinical history was about one year and if the intermittent nature of the symptoms was due to the cysts periodically enlarging and causing rupture of parts of their ependymal lining, thus exciting a mild gliosis, the ultimate appearance of the cysts is readily explained. A slightly higher intracystic pressure in case 2 would account for the flattening of the ependymal lining cells and the rupture of the attenuated wall into the subarachnoid space.

The clinical history and examination and results of other investigations are not diagnostic of the presence of a benign intracerebral cyst. The intermittent nature of the symptoms and the lengthy history may indeed suggest the presence of cerebrovascular disease in the middle-aged subject. When evidence of raised intracranial pressure is found and a space-occupying lesion detected by special techniques, a diagnosis of neoplasm will almost certainly be made unless the cyst is entered accidentally during ventriculography (Handa and Bucy 1956). A firm diagnosis can be made only at operation with the help of histological examination of the cyst wall.

Treatment by establishing drainage of the cyst into the subarachnoid space (Jakubiak et al., 1968), or into the ventricular system (Argyopoulos and Heppner, 1970) appears to lead to complete cure.

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