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Short report

Multiple intracranial enterogenous cysts

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SUMMARY The case of a 40-year-old woman with increasing ataxia is described. Although the clinical presentation and evoked response studies raised the possibility of multiple sclerosis, further investigation revealed multiple cystic intracranial lesions. Surgical excision of one of the lesions relieved the patient's symptoms. Histological examination revealed that this was an enterogenous cyst. Although single cysts of this type have rarely been reported occurring in the posterior cranial fossa, the occurrence of multiple lesions, some in the supratentorial compartment, appears to be unique.

Case report

A 40-year-old-lady presented with a four month history of progressive difficulty in walking. She had become progressively more unsteady on her feet and had also noticed increasing stiffness in her legs with a tendency to spontaneous jerking of her legs when resting. For two months prior to presentation she had been aware of weakness in her arms when carrying heavy objects. She had had some hesitancy of micturition but no urgency or incontinence. There were no ocular or sensory symptoms and apart from occasional episodes of migraine, there were no headaches suggesting raised intracranial pressure and no systemic symptoms. Prior to her presenting illness, in childhood, a small "cyst" had been excised from her scalp in the occipital region. Regrettably no further information was available regarding the nature of this lesion. There was a history of Huntington's disease in her maternal family but her mother had died aged 62 years without manifesting this condition. Our patient had two older and two younger siblings and three children, all of whom were perfectly well. Examination revealed no systemic abnormalities. Examination of the scalp revealed no evidence of a cutaneous sinus. There were no involuntary movements and higher cerebral function was intact. The visual fields, fundi and acuity were normal. Sustained first degree horizontal nystagmus was present on lateral gaze to both sides, the maximum amplitude was on gaze to the right. The rest of the cranial nerves were intact and speech was normal; she walked with a spastic and ataxic gait. A mild spastic tetraparesis was present with symmetrically exaggerated reflexes, bilateral extensor plantar responses and absent abdominal reflexes. The jaw jerk was exaggerated. Past pointing and intention tremor were elicited in the right arm only, but this was not accompanied by impairment of repetitive or alternating movements and there were no other cerebellar signs.

Radiographs of the chest, skull, dorsal and lumbar spine were normal. Radiographs of the cervical spine revealed failure of fusion of the posterior arch of the atlas but no other abnormality. Pattern reversal visual evoked responses (VERs) were abnormal with symmetrical delay and dispersion of the major components. Somatosensory evoked responses from median nerve stimulation at the wrist were normal. Brainstem auditory evoked responses (BAERs), however, were abnormal, being significantly asymmetrical (right V–I 3·7 ms, left V–I 4·6 ms), this delay occurring between waves III and IV. An electroencephalogram (EEG) revealed a significant generalised excess of theta and delta activity, which was sharply contoured in the left temporal region and of highest amplitude in both frontal regions. Cranial computed tomographic (CT) scans (fig 1) revealed a large, low attenuation lesion (+2 to −139 Hounsefield units) in the region of the fourth ventricle which could not be separately visualised. There was evidence of acute obstructive hydrocephalus with periventricular lucency and on higher CT plane cuts, six other similar separate small rounded lesions with similar attenuation to the posterior fossa lesion were visualised (fig 1). The majority of the lesions appeared to be associated with the ventricles and basal cisterns, although some were subependymal in position. The higher, more dorsally situated, supratentorial lesions appeared to be separate from the ventricular system and within the brain parenchyma (fig 1). There was no enhance-

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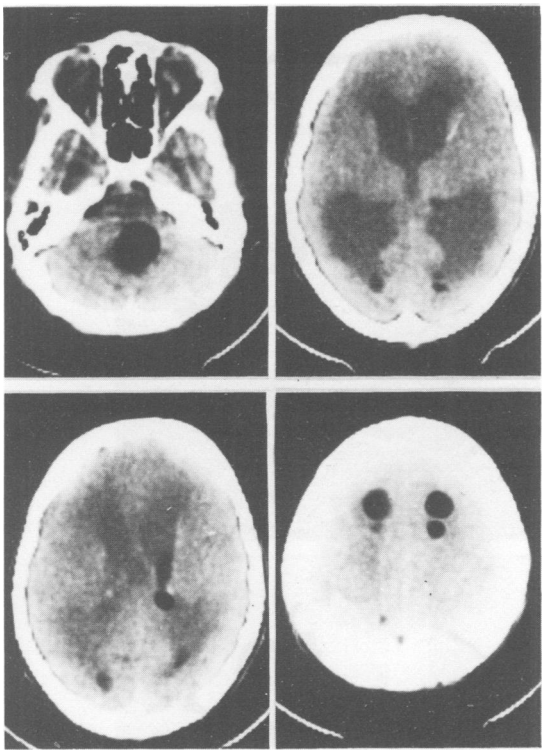

Fig 1 *Composite photograph of four CT images in different planes showing multiple low attenuation lesions in: the posterior fossa, body and posterior horns of the lateral ventricles and, at the highest plane, within the brain parenchyma. Attenuation coefficients within the lesions +2 to −139 Hounsefield units.*

ment in any of these lesions after intravenous contrast administration.

A right sided ventriculoperitoneal shunt was uneventfully inserted which relieved the hydrocephalus. Following this procedure there was no improvement in her gait and her legs became progressively weaker over the next two days. Water soluble contrast myelography, combined with CT scanning of the cervical region was performed. This demonstrated that the space occupying lesion within the fourth ventricle extended caudally, compressing the medulla, through the foramen magnum to the level of C1. The spinal cord was displaced forward and at the foramen magnum, compressed to a thin ribbon.

An occipital craniectomy was performed with removal of the posterior arch of the foramen magnum, the arch of the atlas and the spine and laminae of C2. The dura was opened in this procedure and found to be adherent to a tumour passing through the foramen of Magendie into the fourth ventricle, extending caudally to compress the first and second cervical segments. The cerebellar hemispheres were normal. The tumour was dissected from the dura and incised to reveal a large quantity of yellowish fluid, with the consistency of thick cream. In addition there was firmer tissue with a waxy consistency but no foreign material such as teeth or hair was identified within the cyst cavity. After the contents had been evacuated, as much as possible of the cyst wall was removed leaving portions which were densely adherent to the floor of the fourth ventricle, the cerebellar tonsils and the dorsum of the cord to the level of C2. After operation, the patient made an uneventful recovery. At the time of writing, nine months after surgery, she has made a complete clinical recovery and has no neurological deficit. Follow up CT scans show no evidence of any recurrence of the posterior fossa lesion and no alteration in the size of the other lesions. Further EEG recordings reveal resolution of the delta activity and significant generalised improvement with minor residual theta abnormalities remaining in the left temporal region only. Repeat evoked potential studies are within normal limits.

Pathology

Two fragments of the cyst wall (2·0 × 1·5 cm and 0·8 × 0·8 cm) each approximately 0·2 cm thick were examined. The tissue was soft, fibrous and nearly white in colour. Portions of the cyst wall were fixed and embedded by standard paraffin techniques. Histological examination was performed on sections stained with haematoxylin and eosin and by silver impregnation for reticulin. Mucin was identified with mucicarmine, alcian blue and the periodic acid Schiff (PAS) reaction with and without diastase digestion. Stains for enterochromaffin granules (alkaline diazo, lead haematoxylin, Masson-Fontana and Grimelius silver methods) were performed. Other sections were examined immunohistochemically for the presence of epithelial membrane antigen, milk fat globule antigen, cytokeratin and glial fibrillary acid protein (GFAP). A small portion of the cyst wall was processed separately for electron microscopy. A sample of the cyst contents which had a thick creamy amorphous consistency and pale yellow colour, was examined separately.

Histological examination showed the cyst wall to be composed of connective tissue lined with cuboidal and flattened non-ciliated epithelial cells containing pale, eosinophilic, slightly vacuolated cytoplasm (fig 2). The nuclei were basally orientated, particularly in the larger cells and contained stippled chromatin. Mucin stains demonstrated intracytoplasmic neutral mucin which was not digestible by diastase. No enterochromaffin granules or GFAP were identified. Immunohistochemical studies demonstrated epithelial membrane antigen, milk fat globule antigen and cytokeratin within the lining cells (fig 2). Electron microscopy showed that the epithelial cells of the cyst wall lay on a basement membrane which separated them from bundles of collagen comprising the wall itself. In some areas, the epithelial lining cells of this cyst appeared to assume a double layer with the inner cells showing dense chromatin and a well defined nucleolus, but over most of the cyst wall, a single epithelial layer was present. Microvilli were present, but appeared stunted and indistinct. No intracellular lumina were seen within the cell cytoplasm, organelles were sparse and the nuclei were convoluted with margins of condensed chromatin. Cilia and blepharoplasts were not seen. Histochemical examination of the solid portion of the cyst content showed this to consist entirely of neutral lipid, the fluid portion of the cyst contents was not available for study.

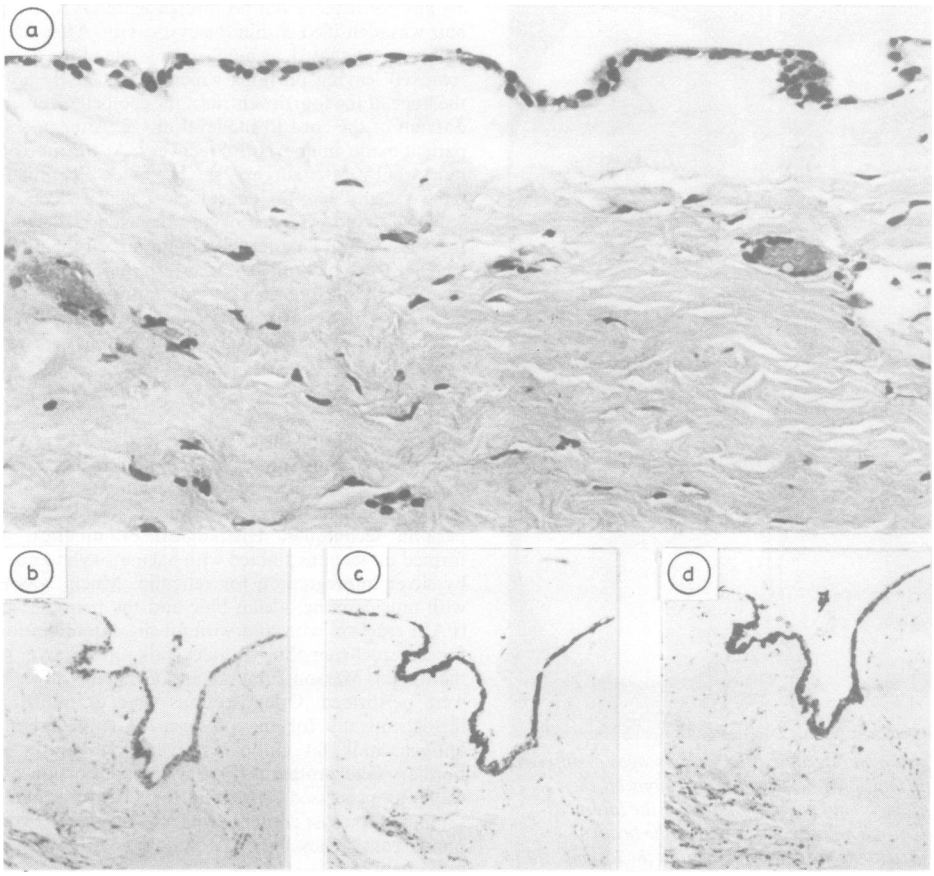

Fig 2 *Composite photograph of sections from the cyst: (a) histological appearance showing the epithelial lining of the cyst lying on connective tissue forming the cyst wall (haematoxylin and eosin, × 260). Immunohistochemical preparations for: (b) Epithelial membrane antigen (× 75), (c) Milk fat globule antigen (× 75), (d) Cytokeratin (× 75), showing positive reactions for each of these within the epithelial cyst lining.*

Discussion

Enterogenous cysts of the neuraxis are unusual cystic lesions which have an epithelial lining resembling gastrointestinal mucosa.[1 2] They most frequently occur in the spinal canal, when they are a well recognised[3 4] but uncommon cause of spinal cord compression and can occasionally pursue a fluctuating clinical course which may mimic mutiple sclerosis.[5] Anatomically they occur most frequently near the junction of the cervical and thoracic spinal cord, particularly in young patients[1] and are often associated, as in our patient, with developmental vertebral anomalies. We have been able to locate only two previous case reports[6 7] in the literature describing intracranial enterogenous cysts, although other reports describe such lesions in the high cervical region[8] and around the foramen magnum.[9] We have found no previous reports of patients with either multiple enterogenous cysts, or with such cysts above the tentorium. The developmental origin of such cysts has been discussed previously[1 10] and a number of theories have been proposed about their aetiology although this issue is still unresolved.

In the present case the lesion removed from the posterior cranial fossa has the histological features of an enterogenous cyst. The immunohistochemical studies lend further support to this diagnosis. The absence of GFAP in the cyst lining cells and the cyst wall are features against an ependymal origin of the

cyst. The presence of epithelial membrane antigen, cytokeratin and milk fat globule antigen suggest that this cystic lesion was embryologically derived from the primitive foregut. Ultrastructural studies are compatible with this diagnosis.

In the absence of a biopsy, the nature of the supratentorial lesions visible on the CT scans remains uncertain. Their small size and asymptomatic presence do not indicate the need for surgical removal. Radiologically however they have attenuation coefficients identical to the posterior fossa mass. It is probable therefore that they have a similar pathological basis and are multiple enterogenous cysts although if so they are in a hitherto undescribed site for such lesions and their multiplicity appears to be unique.

The initial abnormalities in the evoked potential studies are also noteworthy. The delay in the BAERs was presumably due to distortion of the conducting pathways within the brainstem by direct pressure from the cyst. The delayed and dispersed visual evoked responses, however, do not appear to have been due to a pathological lesion in the optic pathway. The resolution of these abnormalities following surgery suggests that they were due simply to the increased intracranial pressure and obstructive hydrocephalus as has been previously described.[11]

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