

GIANT INTRACRANIAL EPIDERMOID CYST AND DEMENTIA

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ABSTRACT

Intracranial epidermoid cysts rarely present with a progressive dementia and ataxia. A 67 year old lady is reported who had been suffering from generalized tonic clonic seizures over a 60 year period. She presented with a 12 month history of progressive mental deterioration which was treated conservatively without benefit. Recent appearance of progressive ataxia prompted further investigations and a giant epidermoid cyst in the left middle and anterior cranial fossae was discovered. Subtotal excision has resulted in clinical improvement. The dangers of "waiting and watching" are highlighted.

INTRODUCTION

Mental deterioration in epileptics is a well documented yet poorly understood entity.¹⁻³ Drug toxicity may be responsible for mental deterioration in some patients rather than epilepsy itself.³ In long standing epileptics, development of delayed mental deterioration may not be due to epilepsy or drug toxicity but may be due to other underlying treatable causes such as subdural haematoma, normal pressure hydrocephalus or slow growing brain tumours. Unawareness and misdiagnosis may lead to a "wait and watch" approach and unwarranted increased morbidity and mortality in older patients, with long-standing epilepsy.

Such a scenario occurred in a 67 year old epileptic due to a giant epidermoid cyst which presented with a progressive mental deterioration over a 12 month period. The appearance of a progressive ataxia prompted further investigations and successful surgical management. Differential diagnosis in such cases is discussed.

CASE REPORT

A 67 year old lady presented with a 12 month history of progressive mental deterioration. A history of generalized tonic clonic seizures over a 60 year period was obtained. The seizures had been fairly well controlled on Diphenyl hydantoin (300 mgs daily), for the past 10 years. Apart from a reversible stroke involving the left sided limbs about 2 years before this presentation, she had no other medical history of note. Prior to admission she had been managed conservatively for about 8 months, a provisional diagnosis of dementia associated with a long-standing epilepsy having been made. On this "wait and watch" approach she further worsened. She developed a progressive difficulty in walking and became chair bound.

General physical examination was unremarkable. Examination of higher mental functions revealed severe

impairment of cognitive functions, emotions and both short and long term memory. She was confused and disorientated in time, place and person. She had no focal neurological signs except for an inability to stand or walk despite normal limb power. No evidence of papilloedema was noted.

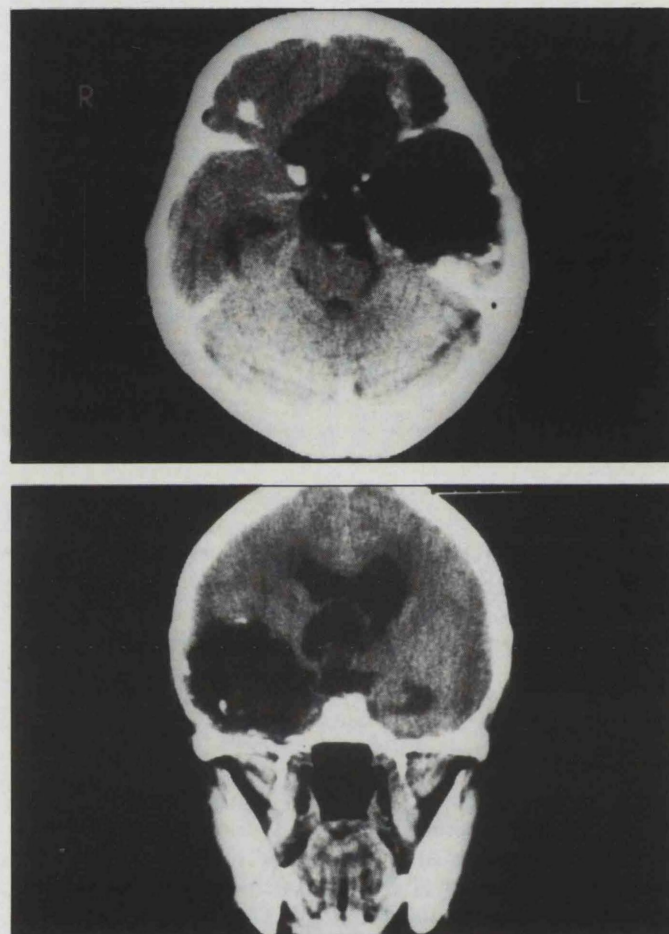


Fig 1A and 1B – Axial (1A) and coronal (1B) CT scans showing a large basal low density mass lesion in the middle and anterior fossa and suprasellar cistern with dilated cerebral ventricles.

Haematological and biochemical tests showed normal results. Blood levels of Diphenylhydantoin were within the therapeutic range. The computed tomographic scan of the head (CT Scan) demonstrated an extensive low attenuation lesion in the left middle cranial fossa extending anteriorly in the anterior cranial fossa and medially in the suprasellar cistern. This extrinsic lesion was deforming the anterior horns of the lateral ventricles bilaterally and causing ventriculomegaly with periventricular lucencies suggesting raised intraventricular pressure. Scattered foci of calcification were noted. The appearances were suggestive of an extrinsic cystic basal lesion. (Figs 1A & B). Bilateral internal carotid angiography confirmed the findings on the CT scan and ruled out a vascular lesion. There was displacement of anterior

cerebral branches across the midline to the right with displacement and splaying of the left middle cerebral artery branches (Fig 2A & B).

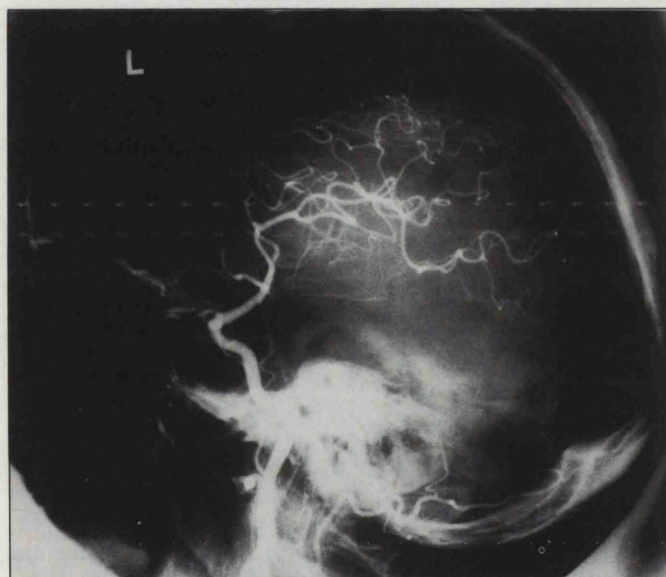
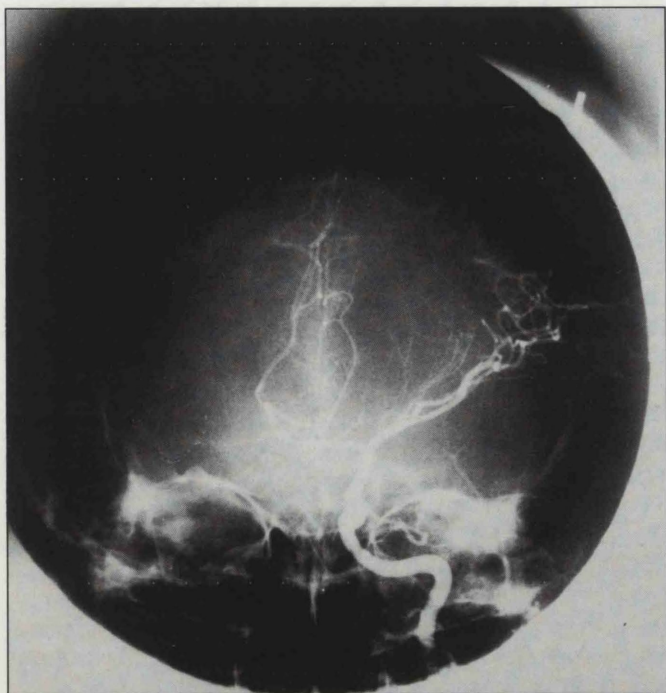


Fig 2A and 2B – Antero-posterior (2A) and lateral (2B) angiographic views showing upward displacement and splaying of the left middle cerebral branches with shift of the anterior cerebral vessels across the midline suggestive of a large extrinsic avascular mass lesion.

A left fronto-temporal craniotomy was performed and a giant basal epidermoid cyst was encountered. The contents of the cyst were evacuated and using a microvascular technique the cyst wall was excised partially due to its extensive and adherent nature to the neurovascular structures. The lesion appeared to be encasing the middle cerebral branches completely. The post operative recovery was uneventful. The histopathological examination (Fig 3) confirmed the nature of the lesion. Gradual improvement in her mobility and mental state occurred. When seen in the outpatient clinic at 5 months following surgery, she was fully orientated in time, place and person. She was cheerful and had improved in both long and short term memory. Her cognitive functions also showed improvement. She could walk without support but mild ataxia was still evident. She continues under out-patient review.

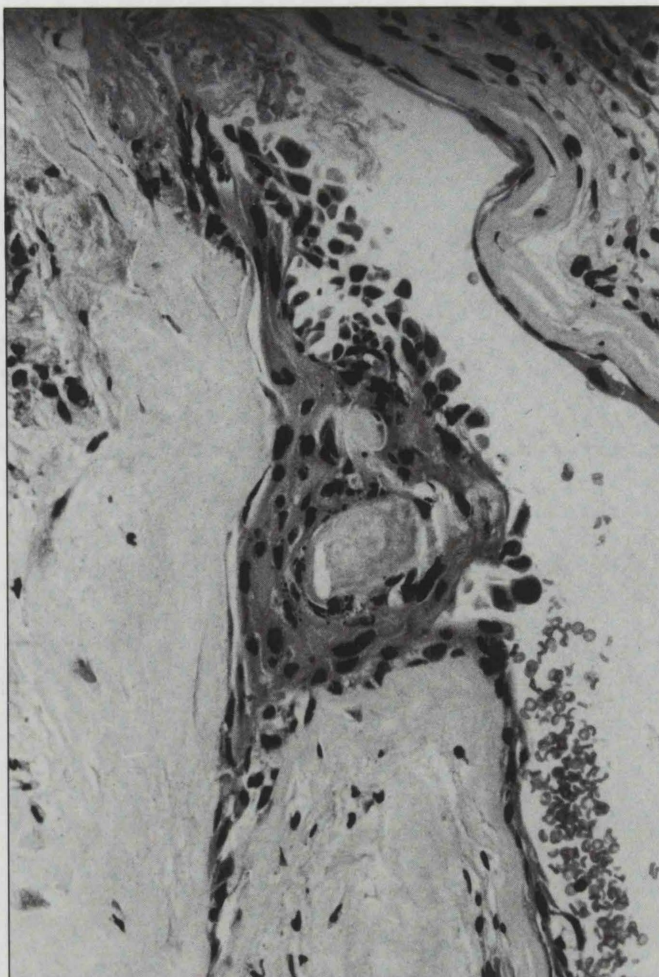


Fig 3 – Photomicrograph of wall of intracranial epidermoid cyst (H and E X 330) showing squamous epithelial lining.

DISCUSSION

It has been established that mental changes can occur in patients with long standing epilepsy,¹ for which a wide range of causes exists. Lesser et al¹ noted that in a survey of 1905 patients, 36 per cent revealed an impaired mental state. Neuropathological studies have shown that progressive cortical cellular changes are present in many longstanding epileptics.² These may well account for cognitive and emotional deterioration. However, a significant proportion of patients have brain pathology prior to the onset of symptoms,¹ such as intracranial tumours, vascular disorders, perinatal brain injury, congenital anomalies, degenerative disorders, or inborn errors of metabolism.

Drug toxicity may also be responsible for mental deterioration in some instances, rather than epilepsy itself. The diagnosis of anticonvulsant toxicity should be made on clinical criteria, supported by biochemical tests.³ The patient under consideration was on Diphenylhydantoin (300 mgs daily) for a period of 10 years and had tolerated the drug well. She had no nystagmus, slurring of speech or tremors and blood levels of Diphenyl hydantoin were within the therapeutic range.

Mental deterioration in the elderly may occur due to presenile or senile dementia. The term dementia describes a progressive disintegration of intellect, memory and abstract powers of thought. Patients present with errors of judgement, loss of memory, emotional lability and personal neglect. In 43 to 87 per cent of cases of dementia, the aetiology can be determined.⁴

However in the rest of the cases it cannot be predicted accurately during life.⁴ Progressive mental changes in this age group are known to occur due to a variety of conditions which include intracranial tumours, normal pressure hydrocephalus, and chronic subdural haematomas. Neurological signs are often absent especially during the early stages and a treatable cause may be missed as happened in this patient.

In the differential diagnosis, normal pressure hydrocephalus (NPH) should also be considered in patients such as the one reported here. NPH is a type of communicating hydrocephalus of unknown aetiology. Usually there is no history of antecedent intracranial disease although it has been reported following head injury and leptomeningeal inflammation.⁵ A triad of symptoms is known to occur in NPH: Progressive dementia, ataxia and incontinence of urine. This triad may also occur with other lesions, e.g. brain tumours, subdural haematomas. In this patient, there was no incontinence of urine but on clinical grounds alone NPH could not be ruled out.

Intracranial epidermoid cysts are rare, comprising less than 1% of brain tumours.⁶⁻¹³ They are benign, slow growing lesions derived from epithelial cells destined to form skin during the third to fifth weeks of life.

The absence of hair follicles and sweat glands distinguishes them from dermoids. The capsule of epidermoid cysts is usually semi-transparent and encloses laminated strands of keratin and epithelial debris. Despite their developmental aetiology, the majority of epidermoid cysts present later in life with maximum incidence occurring between thirty and sixty years. Symptoms vary with location, the most common site being the cerebello-pontine angle, less commonly the para pituitary – chiasmal region and least commonly in an intracerebral – intraventricular location.⁷ Epidermoids within the cerebral hemisphere (about 15% cases) or near temporal lobes can give rise to seizures and this may be the only symptom for many years.⁵ In our patient, the seizures were present over 60 years. To our knowledge, a maximum duration of 30 years only has previously been recorded.

A high incidence of minor mental symptoms¹⁰ and an absence of papilloedema in half the patients¹² are features which make clinical diagnosis of intracranial epidermoid cyst difficult⁶ as happened in our patient. Epidermoid cysts typically appear as low density mass lesions on the CT scan but a few hyperdense ones have been described.¹³ A focal bone erosion is noted in the calvarial or extradural cases. The typical angiographic vessel displacement or encasement is seen in extrinsic – subarachnoid epidermoid cysts as in our patient. Despite their benign nature, the epidermoid cysts involving the subarachnoid spaces are extremely difficult to remove totally on account of their extensive and adherent nature.^{8,9}

However results of subtotal excision are still good. Post operative meningitis due to cholesterol spillage may occasionally occur. Yamakawa et al⁷ have reported a 93% twenty year survival rate with recurrence of symptoms occurring only after many years.

The manifestation of progressive dementia and ataxia in our patient with a giant epidermoid cyst may be due to two different phenomena, which overlap. The first phenomenon is the space occupying nature of the tumour and the second related to ventriculomegaly secondary to an extensive obliteration of the basal subarachnoid cisterns by the tumour. It is well known that the epidermoid cyst slowly increases its

volume and it simply 'flows' into any available subarachnoid space.⁸ It conforms to the shape of the subarachnoid space which it enters and does not displace normal neural and vascular elements until all available subarachnoid space is occupied. However at a later stage a giant epidermoid cyst not only displaces the neuro-vascular elements but compromises CSF flow in the basal subarachnoid spaces, as happened in the present case. However partial release of subarachnoid cisterns had obviated the need for shunt surgery in this case.

CONCLUSIONS

Delayed progressive dementia and ataxia may occur in elderly patients with a longstanding epilepsy secondary to a treatable cause such as an intracranial epidermoid cyst. A high index of suspicion, prompt neuro-radio-diagnostic investigations (CT scan) and aggressive surgical intervention are necessary. The dangers of "waiting and watching" can not be over-emphasized.

ACKNOWLEDGEMENTS

We thank Miss Louise Young for the preparation of this manuscript and the Medical Illustration Department, Royal Preston Hospital for the photographs.

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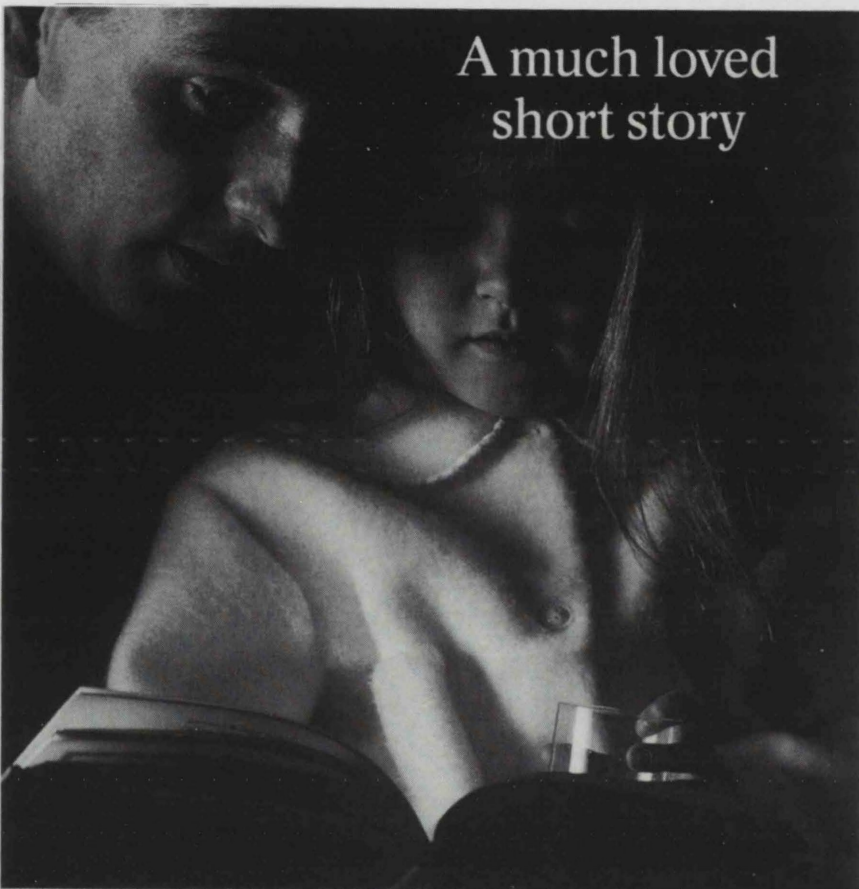
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