

LETTERS TO THE EDITOR

Misinterpretation of neuroradiological appearances of an epidermoid cyst

Epidermoid cysts have been described as the most frequent tumorous malformation of the CNS.^{1,2} The favourable outcome after total removal¹ make an early correct diagnosis highly necessary. We describe a 48 year old male technician who was healthy until April 1988, but who developed thoracic pain and sciatica-like backache on the right side which disappeared a few weeks later. In July neck stiffness and double vision occurred. He was admitted to our hospital in August 1988.

Nuchal rigidity, paralysis of both abducent nerves, weakness of the right superior oblique muscle and the left facial nerve, as well as papilloedema were found. There was a slight displacement of the palatopharyngeal arch to the left. He was afebrile. Routine laboratory data were normal. In September, episodes of confusion and a left-sided ptosis occurred. There was now plegia of both abducent nerves and a weakness of both recti superiores muscles. The cerebrospinal fluid (CSF) showed 40 cells per cubic millimetre. The protein content was slightly elevated to 52.5 mg%. A differential CSF cell count revealed 3% tumour cells, 60% monocytes, 4% macrophages, 4% granulocytes, 15% large lymphocytes, and 16% small lymphocytes. The tumour cells were three times as large as monocytes; the cytoplasm was broad and basophilic. Immunocytochemistry showed heavy labelling of tumour cell cytoplasm and membranes with the epithelial membrane antigen (EMA) indicating their epithelial origin.

Computerised tomography (CT) and magnetic resonance imaging (MRI) revealed an asymmetrically widened interpeduncular cistern and a cerebellopontine cistern on the left side without contrast media or Gd DPTA enhancement. No Hounsfield numbers were measured. MRI sequences were sagittal SE

700/15 T1 weighted and coronal SE 2500/15/90 T2 weighted. Using these "standard" sequences, no differences in the intensity of CSF and the widened cisternae were indicated. Radiographs of the skull did not show any destruction, nor calcifications. As a result of these findings and the typical localisation, an arachnoid cyst was suspected (fig). Finally, the patient developed an acute phase of hepatitis B. He died on 17 September after repeated cardiac arrests.

A complete necropsy showed multiple acute liver necroses, fibrinous endocarditis, subpleural petechiae, pulmonary oedema and the morphological signs of shock. The neuropathological investigation revealed a typical epidermoid cyst at the base of the brain extending from the interpeduncular fossa to the upper medulla oblongata with a mean diameter of 1 cm. The whole left aspect of the pons and the left cerebellopontine angle were covered by the tumour. Both oculomotor nerves, the left trigeminal, facial and the vestibulocochlear nerves as well as both abducent and glossopharyngeal nerves were covered with pearly masses. The tumour lacked calcifications. There was no bone destruction at the base of the skull. The brainstem was slightly displaced to the right.

We conclude that in cases with cranial nerve lesions and cerebellar signs or even meningeal signs an epidermoid cyst at the base of the brain should be excluded. As our observations show, routine performance of CT and MRI may fail to reveal the tumour.^{6,7} In most cases, epidermoid cysts have T1 MRI signals intermediate between brain and CSF whereas arachnoid cysts are identical with CSF.⁸ This characteristic MRI feature was not present in our case. However, the correct diagnosis might have been suggested by characteristic Hounsfield numbers on CT. Furthermore, a possible distinction between arachnoid and epidermoid cyst could have been achieved by additional MRI sequences. A multi-echo sequence with a TR of around 3000 and 6-9 echoes ranging from TE 26-TE 156 will nearly always show a difference between epidermoid cysts and CSF at least in some part of the series.⁹ Hadley and Patterson¹⁰ recommended a sequence sensitive to T1 changes, depending on field strength that is, at 0.35 Tesla an SE 300-500/15-30 or an inversion recovery 1600/400/40 to distin-

guish between both lesions. Using suitable sequences at 1.5 Tesla in our case, the epidermoid cyst might have been hyperintense to CSF. A displacement of basal brain structures suggesting a cystic process,^{6,7} may be missed in epidermoid cysts of small sizes. CSF examination may indicate aseptic meningitis or epithelial tumour cells after spontaneous rupture^{1,3-5} but gives negative results in cases with an intact cyst wall. Full use of modern neuroradiological techniques and CSF investigation is necessary to establish the diagnosis of an epidermoid cyst.

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Gingival hyperplasia due to sodium valproate

Gingival hyperplasia is a common disorder seen in patients with epilepsy treated with phenytoin.¹ The incidence is approximately 33% in all epileptic patients receiving phenytoin, and higher in institutionalised patients and those with poor oral hygiene.² Gingival hyperplasia is a rare side effect of sodium valproate therapy, and only a single case has been reported in a 15 month old child.³ A patient with epilepsy receiving sodium valproate is reported who developed gingival hyperplasia during therapy.

At the age of 12 years the patient developed myoclonic epilepsy, attacks being frequent in the early morning. She did not receive any anticonvulsant drugs. A year later she developed generalised tonic-clonic seizures. When first seen at the age of 14 years, she was alert, intelligent without any dysmorphic features. General and neurological examination was normal. She was advised to take sodium valproate 600 mg daily. Blood count, liver and renal function tests were normal.

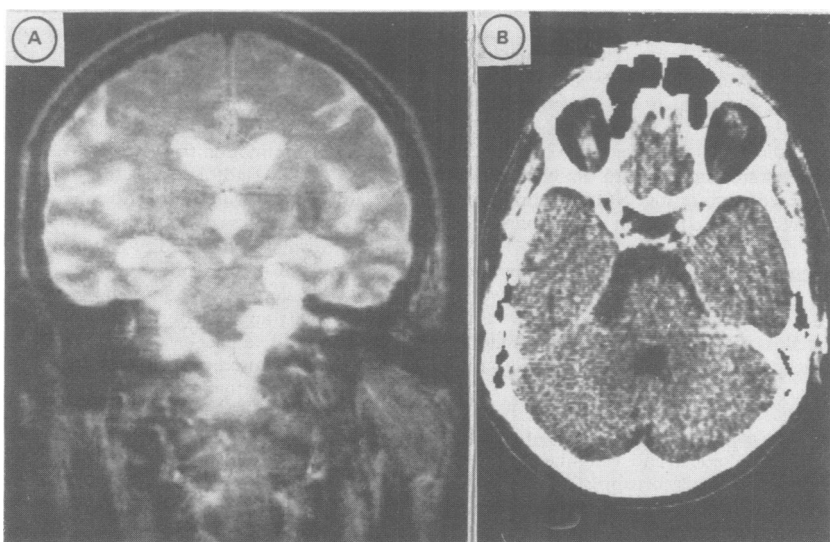


Figure a) MRI (coronal plane, SE 2500/15/90 T2 weighted); b) CT (transverse plane). Neuroradiological findings only disclosed an asymmetrical widened interpeduncular cistern and a cerebellopontine cistern on the left side without contrast media or Gd DPTA enhancement.

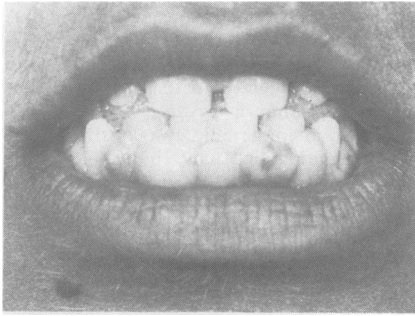


Figure Photograph showing gingival hyperplasia.

Electroencephalography showed frequent generalised bursts of polyspikes interposed on normal background activity. Eighteen months after starting sodium valproate therapy, gingival hyperplasia was noted which increased progressively. The gums were swollen, shiny, pale pink, firm and were covering approximately one half of the teeth. They did not bleed when touched and there were no engorged vessels (fig). The gingival hyperplasia was most prominent on the labial surface of the anterior teeth of the lower jaw. Investigations including haemogram, liver and renal function tests were again normal. Serum levels of valproate were not carried out. The drug regime was changed to a combination of carbamazepine (600 mg daily) and nitrazepam (15 mg daily) which resulted in poor control of seizures but a complete regression of gingival hyperplasia in three months.

This case appears to be only the second one of sodium valproate induced gingival hyperplasia. This patient differs from the earlier report of a 15 month old child with infantile spasms.³

The patient belonged to an upper middle socio-economic group and was of average nutritional status without any evidence of nutritional deficiency, and she had never received diphenylhydantoin. She did not have any hormonal disturbances, was neither using oral contraceptive drugs nor was she pregnant and the fact that it regressed within three months of stopping sodium-valproate suggests that it is unrelated to puberty. That her gingival hyperplasia was not of an idiopathic hereditary variety is confirmed by the fact that there was no family history of gingival hyperplasia and that it regressed on stopping treatment with sodium valproate. Her blood counts were normal on two occasions and the gums were neither friable nor bled when touched which excluded the possibility of gum hyperplasia associated with leukaemia.

Several complications of sodium valproate are known⁴ but gingival hyperplasia is described only as a single case report in a 15 month old child with infantile spasm whose gingival biopsy showed oedematous connective tissue, dilated capillaries and dense infiltration by inflammatory cells especially mast cells and the authors implicated the role of mast cells in the pathogenesis of gingival hyperplasia due to sodium valproate.³ The effect of sodium valproate on the periodontal and oral health of epileptic patients (adults⁵ and children⁶) has been carried out in prospective studies which showed no unwanted effects on oral and dental health. Gingival hyperplasia is a common side effect of phenytoin therapy, but has also been observed with

mephenytoin,⁷ and primidone and sulthiam⁸ therapy. The pathogenesis of gingival hyperplasia during phenytoin therapy may be due to stimulation of fibroblasts⁹ or due to folic acid deficiency.¹⁰ There is no indication of the cause of the gingival hyperplasia in this patient.

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A case of angiographically occult spinal AVM

Intramedullary haemorrhage, or haematomyelia is not an unusual clinical symptom of a spinal arteriovenous malformation (AVM). Recent developments in neuroradiological methods usually identify the aetiology of haematomyelia pre-operatively, but we report a case in which the AVM that was not identified by angiography.

A 40 year old woman was in good health until two days before admission, when while asleep she had an attack of sudden back pain on the right side radiating to the chest, and accompanied by vomiting. Next morning, she noticed weakness in her right leg and bilateral sensory loss below the thoracic level. Neurological examination on admission showed weakness in the right lower-extremity, increased bilateral patellar and ankle reflex with positive Babinski reflex, hypesthesia below left T-7, hypalgesia and thermohypesthesia below right T-7, bilaterally diminished vibration sense below the thoracic cage and mild urinary disturbance. Haematological and coagulation tests were normal. The cerebrospinal fluid (CSF) was clear. Computed Tomography (CT) scan showed a haematomyelia as a high density area from the level of T-2 to T-7 with a "string-like" enhanced lesion at the T-4 level (fig 1). Myelography followed by a CT scan

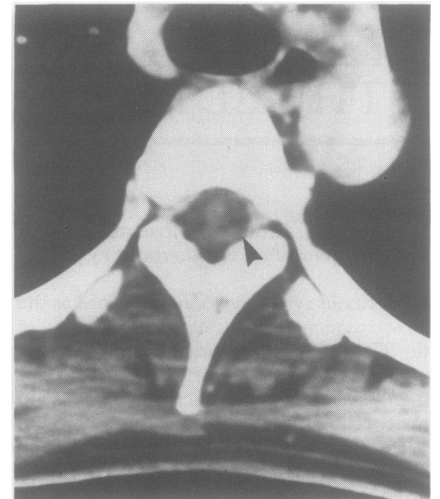


Figure 1 CT scan with intravenous contrast medium showing the haematomyelia and an intramedullary "string-like" enhanced lesion (arrow) at T-4 level.

showed cord swelling from the level of T-2 to T-4 without any abnormal vascular images. Magnetic Resonance Imaging (MRI) demonstrated a mixed intensity lesion at T-3 and T-4 level in both T-1 and T-2 weighted images. Complete spinal angiography was performed one week after admission. The right thyrocervical and costo-cervical trunk angiograms showed the anterior spinal artery, from the ascending cervical artery, to the T-4 level without any intradural vascular malformation (fig 2). Other angiograms did not show abnormal findings. Her Adam-Kiewics artery branched from the left L-1 lumbar artery.

Four weeks after admission, we performed a laminectomy from T-2 to T-5 and made a small midline myelotomy at the T-3/4 level to evacuate the intramedullary haematoma, but did not examine the tissue surrounding the haematoma. Transient neurological improvement followed but then there was further neurological deterioration due to recurrent intramedullary bleeding, shown by CT scan two weeks after the first operation.

A second operation was performed about three weeks after the first operation. The midline myelotomy was extended and the intramedullary haematoma, surrounded by the fibrous tissue, was evacuated. Total removal of the haematoma and fibrous tissue revealed a tiny abnormal vascular lesion; it was completely removed and was shown by histological examination to be an AVM. The weakness of the right leg and the sensory disturbance had worsened and she was troubled by numbness of the right leg but she was able to walk with an aid of a stick three months after the second operation.

Although cavernous angiomas of the spinal cord are usually angiographically occult,⁴ we could not find a report of a case of haematomyelia caused by a spinal AVM that could not be identified by adequate spinal angiography. Only Veerpan's case⁵ of haematomyelia with a cryptic spinal AVM resembled our case, although in this case spinal angiography was not performed. In our case, it was difficult to determine the aetiology due to conflicting results of the neuro-radiological examination, and the presence of AVM was not confirmed until the second operation.