20 mg isosorbide developed an identical headache. On this occasion it was associated with a left ptosis and diplopia and he was referred for neurosurgical opinion.

On examination blood pressure was 120/80 mm Hg and temperature 37.4°C. There was no neck stiffness but a partial left third nerve palsy and left temporal hemianopia were present. Computed tomography showed a large mass of heterogeneous density arising from the pituitary fossa with 10 mm suprasellar extension. The serum prolactin concentration was normal (102 mU/L), normal <450) and ACTH deficiency was probable in view of the plasma cortisol of <60 nmol/l at the time of presentation. Three days after admission haemorrhagic pituitary tumour was resected via the transphenoidal route. Immunohistochemistry demonstrated a chromophobe adenoma negative for ACTH, GH, PRL, TSH, LH and FSH.

The third nerve palsy resolved within four days of tumour decompression. Six weeks after operation ACTH function had recovered, permitting the withdrawal of steroids, and the visual fields were normal. The patient remains well on no replacement therapy.

The onset of headache and neurological deficit ninety minutes after ingestion of 20 mg isosorbide mononitrate made it likely that the two events were connected, particularly as this is the time of peak plasma drug concentrations following oral administration of isosorbide.4 Lever et al demonstrated a marked pressor response to TRH in an acromegalic patient who had developed pituitary apoplexy following a TRH stimulation test,2 and it seems likely that changes in systemic blood pressure may produce apoplexy in some pituitary tumours. These tumours may be particularly susceptible to such changes because of the local vascular anatomy; some tumour regions are probably solely dependent on a tenuous blood supply from the hypophysial-portal capillary network.1

The patient described here was naturally unwilling to be rechallenged with isosorbide so blood pressure changes following the drug were not known. Although there are no previous reports in the literature connecting isosorbide and tumour haemorrhage there are a few papers describing transient neurological deficit associated with the vasodilator.5 6 We conclude that vasoactive drugs may induce pituitary apoplexy in some patients with pituitary tumours.

Letters

20 mg isosorbide developed an identical headache. On this occasion it was associated with a left ptosis and diplopia and he was referred for neurosurgical opinion.

On examination blood pressure was 120/80 mm Hg and temperature 37.4°C. There was no neck stiffness but a partial left third nerve palsy and left temporal hemianopia were present. Computed tomography showed a large mass of heterogeneous density arising from the pituitary fossa with 10 mm suprasellar extension. The serum prolactin concentration was normal (102 mU/L), normal <450) and ACTH deficiency was probable in view of the plasma cortisol of <60 nmol/l at the time of presentation. Three days after admission haemorrhagic pituitary tumour was resected via the transphenoidal route. Immunohistochemistry demonstrated a chromophobe adenoma negative for ACTH, GH, PRL, TSH, LH and FSH.

The third nerve palsy resolved within four days of tumour decompression. Six weeks after operation ACTH function had recovered, permitting the withdrawal of steroids, and the visual fields were normal. The patient remains well on no replacement therapy.

The onset of headache and neurological deficit ninety minutes after ingestion of 20 mg isosorbide mononitrate made it likely that the two events were connected, particularly as this is the time of peak plasma drug concentrations following oral administration of isosorbide.4 Lever et al demonstrated a marked pressor response to TRH in an acromegalic patient who had developed pituitary apoplexy following a TRH stimulation test,2 and it seems likely that changes in systemic blood pressure may produce apoplexy in some pituitary tumours. These tumours may be particularly susceptible to such changes because of the local vascular anatomy; some tumour regions are probably solely dependent on a tenuous blood supply from the hypophysial-portal capillary network.1

The patient described here was naturally unwilling to be rechallenged with isosorbide so blood pressure changes following the drug were not known. Although there are no previous reports in the literature connecting isosorbide and tumour haemorrhage there are a few papers describing transient neurological deficit associated with the vasodilator.5 6 We conclude that vasoactive drugs may induce pituitary apoplexy in some patients with pituitary tumours.

Cervical syrinx associated with an intramedullary metastasis: case report

Sir: Since its description by Olliver in 1827, syringomyelia and its pathogenesis have continued to be the subject of debate. Syrinxes associated with neoplasms of spinal cord origin have long been recognised, and both the cyst cavity and the associated tumour can now be readily demonstrated with high resolution metrizamide CT scanning and magnetic resonance imaging.1–3 However, the pathogenesis of such cysts has not yet been much discussed since the 1950s.

Metastasis to the spinal cord is unusual but well described. A syrinx associated with a spinal cord secondary, found in a post-mortem specimen, has previously been described.4 We report a case of syrinx associated with an intramedullary spinal cord metastasis diagnosed during life. Its pathogenesis is discussed.

Mr JT had undergone an anterior resection for a Duke’s stage ‘C’ colonic adenocarcinoma in 1980 and had no evidence of systemic recurrence. He presented in November 1984 aged 66 years, with a 2 month history of mild weakness of the left arm and leg of gradual onset. He felt well, and there was no history of neck injury or neurologic disorders. Examination revealed slight weakness of the left leg and a left extensor plantar response. He was followed in out-patients and his condition remained stable for several months. His condition deteriorated in April 1985 and he presented with a one week history of rapidly progressive weakness in the right leg and severe pain and hyperaesthesia in the right arm. Examination revealed bilaterally brisk arm flexes, with marked hyperaesthesia in a C5/T1 distribution on the right. There was bilateral pyramidal weakness in the legs. Bladder and bowel function were normal and an ileal tone preserved.

The patient was admitted and a myelogram performed. (fig. a). A CT scan of the cord obtained the following day showed uptake of metrizamide into a cystic lesion extending over the same segments. (fig. b). Five days later the patient developed painful urinary retention and faecal incontinence. Examination revealed increasing motor deficit in his right arm and legs, an enlarged tender bladder and loss of anal tone.

At operation no extradural or intradural extra-medullary pathology was noted. The spinal cord exhibited a smooth swelling extending over four centimetres. A fine needle was passed into this and 1 ml of clear fluid aspirated; it was indistinguishable from CSF previously obtained at myelography. Exploration confirmed a cyst cavity with a purple grey mass at the candel end. The mass was surrounded by oedematous spinal cord tissue and could not be excised completely. After the operation the patient developed a worsening tetraparesis, and he died in early June. Histology of the biopsy specimen showed a poorly differentiated adenocarcinoma similar in appearance to that of the patient’s bowel tumour. Post-mortem examination revealed widespread metastatic disease affecting the ribs and chest wall, the abdominal lymph nodes and the body of the seventh thoracic vertebra. No intracranial deposits were found. Sections of the spinal cord showed cavitation in the upper cervical region with intramedullary tumour. Imaging, operative and post-mortem findings confirm the presence of a syrinx in this patient. Weitlander, in 1969, made the only previous report of such a finding, in a post-mortem specimen from a neurologically asymptomatic patient.4 He concluded that the syrinx could have predated the metastasis, since there were no clues from the history. In our case, the history suggests that syrinx formation was of recent origin, presumably secondary to the metastasis. This finding may give a clue to the pathogenesis of some tumour-associated "non-communicating" syrinxes.
In the first half of this century, many believed that spinal cord tumours and all syrinxes were aspects of a common process.\textsuperscript{5–7} Syrinx formation was thought to be secondary to metaplasia of the spinal cord, evidenced by glial cell proliferation in the cavity wall. Such gliosis is now felt to be secondary to syrinx formation, and discussion has for many years concentrated on two hypotheses: that syrinx formation results from ischaemic necrosis of the central cord, either as a primary event or secondary to adhesive arachnoiditis involving the meningeal vessels;\textsuperscript{6–8} or, that a disturbance of CSF dynamics allows the cord to be split by pressure waves.\textsuperscript{10,11} While the latter is a most attractive explanation of "communicating" syringomyelia and has experimental support,\textsuperscript{11} it can not readily explain syrinx formation in association with spinal cord tumours. Williams in 1980\textsuperscript{12} could not discount the old theory of metaplasia as an explanation of syrinx formation related to primary tumours, but the association of such cavities with metastatic cord tumours cannot be accounted for in this way.

Pencil-shaped softening of the central cord, often with a cystic component, is well described in association with spinal cord trauma\textsuperscript{13} and compression from extradural metastases.\textsuperscript{14} Jellinger considered that such softening was of vascular origin and was an early stage in the formation of post traumatic syrinxes. There have been several recent reports of such pencil-shaped softening (although without mention of a cyst) in association with intramedullary metastasis.\textsuperscript{15–17}

Hashizume et al\textsuperscript{14,17} reported such softening in three out of five cases of intramedullary metastasis. The changes extended over several segments of either side of the tumour deposit, in the ventral part of the posterior columns and the central grey matter, the same area as is affected in post traumatic cases. Hashizume et al's three cases were all associated with bronchial carcinoma, and the average interval between symptom onset and death was seven weeks.

In our case the primary tumour was a colonic carcinoma, which tends to run a slower course than bronchial carcinoma. The interval between the onset of symptoms in our patient and his death was nine months, far longer than in Hashizume's cases.

Letters

It may be that "pencil-shaped softening", possibly of vascular aetiology, preceded cavitation in the genesis of the syrinx we report. One would expect such cavities to be rare because death usually occurs shortly after the development of intramedullary metasta-
sis.\textsuperscript{15–17} Syrinxes associated with primary cord tumours may be formed in the same way, rather than by fluid exudation as has been suggested.\textsuperscript{12,18}

OJF FOSTER
HA CROCKARD
The Departments of Neurology and Neurosurgery, and the Rita Lila Weston Institute of Neurology, Jules Thorn Building, The Middlesex Hospital Medical School, Mortimer Street, London W1N 8AA, UK

The authors are grateful to Mrs G Stylianou for preparation of the manuscript.

References

Recurrent spontaneous subarachnoid haemorrhage due to spinal haemangioblastoma

Sir: Spontaneous subarachnoid haemorrhage (SAH) of spinal origin is uncommon and accounts for less than 1% of all cases of non-traumatic SAH. When haemorrhage occurs from a lesion in the high cervical region the clinical features may be difficult to distinguish from SAH due to an intracranial lesion. Spinal tumours are a recognised cause of spinal SAH, but spinal haemangioblastomas presenting in this manner have only been described twice previously. Recurrent SAH due to cervical haemangioblastoma has not been reported before.

A 37 year old man presented with sudden onset of headache, vomiting, photophobia and neck stiffness. For one month he had been aware of mild numbness and clumsiness of his left arm. Examination he had meningism plus minimal weakness and proprioceptive deficit in the left arm. SAH was confirmed by lumbar puncture, and the clinical signs in the arm were attributed to a right parietal lesion, possibly an arteriovenous malformation. Cranial computed tomography (CT) showed blood in the 4th ventricle, but no structural lesion. Bilateral carotid and left vertebral angiography showed no source of haemorrhage. A second SAH occurred 4 weeks later which produced no permanent neurological deficit, and 2 weeks after this repeat angiography was again normal. He was discharged home, but 14 weeks after his initial SAH he collapsed with severe headache and neck stiffness during sexual intercourse. On examination there was again neck stiffness and mild weakness of the left arm. CT showed intraventricular blood, but, as previously, none in the subarachnoid space. On myelography the cervical cord was expanded, and selective angiography of the vertebral and left throracic vessels showed a vascular intramedullary tumour at C.2 (fig).

Laminectomy was performed 6 weeks after the third SAH and revealed a haemangioblastoma. At this operation five arterial feeding vessels were obliterated, and at a second procedure the lesion was totally excised. The patient made a good postoperative recovery, but is left with some residual weakness of the left arm. Haemangioblastomas account for 1.6% to 2.1% of all spinal cord tumours and 3.3% of intramedullary tumours. Forty percent of the spinal tumours occur in the cervical region of which 60% are intramedullary, and they most commonly present with features of spinal cord compression. Of two previously described lesions which presented as SAH, one was extramedullary at L. and the other was an intramedullary cervical lesion. When the haemorrhage arises within the cord in the cervical region, bleeding may extend intracranially, and the clinical differentiation from subarachnoid bleeding from an intracranial source may be extremely difficult. Recurrent haemorrhages at short intervals are characteristic of aneurysmal subarachnoid bleeding, and the natural history of the condition in our patient increased the diagnostic difficulties.

In retrospect, the weakness in the left arm should have been a pointer to the true site of the lesion, but this symptom was mistakenly attributed to an intracerebral lesion, initially a possible arterio-venous malformation, and latterly cerebral ischaemia. Furthermore, the absence of blood in the subarachnoid space following the ictus on two occasions was an indication that it had tracked to the ventricular system via the central spinal canal. If cerebral angiography is negative in cases of recurrent haemorrhage a lateral cervical series should be included to exclude a cervical lesion. This would not prolong the procedure or put the patient at increased risk. Spinal haemangioblastoma is a rare cause of spontaneous SAH and may lead to diagnostic difficulties when its presentation mimics intracerebral SAH. This is the first report of recurrent SAH from such a lesion, and it should be considered when other, more common sources of haemorrhage have been excluded.

References


Fig. Vertebral angiogram, anteroposterior projection, showing vascular mass.
Cervical syrinx associated with an intramedullary metastasis: case report.
O J Foster and H A Crockard

*J Neurol Neurosurg Psychiatry* 1987 50: 637-639
doi: 10.1136/jnnp.50.5.637

Updated information and services can be found at:
http://jnnp.bmj.com/content/50/5/637.citation

**Email alerting service**
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/