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References

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Cervical syrinx associated with an intramedullary metastasis: case report

SIR: Since its description by Oliver 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 been much discussed since the 1950s.

Metastasis to the spinal cord is unusual but well described. A syrinx associated with a spinal cord meningioma, secondarily 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 neurological disorder. 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 biceps, triceps, and wrist reflexes, 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 anal tone preserved.

The patient had admitted and a myelogram performed. (fig. a). A CT scan of the cord obtained the following day showed a take-up 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 tendon reflex 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 cystic cavity with a purple grey mass at the cord end. The mass was surrounded by oedematous spinal cord tissue and could not be excised completely.

After the operation the patient developed 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. Weitzen, 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 to 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 syringes were aspects of a common process. Syringomyelia was thought to be secondary to metastasis of the spinal cord, evidenced by glial cell proliferation in the cavity wall. Such gliosis is now felt to be secondary to syringomyelia, and discussion has for many years concentrated on two hypotheses: that syringomyelia results from ischaemic necrosis of the central cord, either as a primary event or secondary to adhesive arachnoiditis involving the meningeal vessels; or, that a disturbance of CSF dynamics allows the cord to be split by pressure waves. While the latter is a more attractive explanation of "communicating" syringomyelia and has experimental support, it cannot readily explain syringomyelia in association with spinal cord tumours. Williams in 1980 could not discount the old theory of metaplasia as an explanation of syringomyelia 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 and compression from extradural metastases. Jellinger considered that such softening was of vascular origin and was an early stage in the formation of post traumatic syringes. There have been several recent reports of such pencil-shaped softening (although without mention of a cyst) in association with intramedullary metasta-
sis.

Hashizume et al 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.

References

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It may be that "pencil-shaped softening" possibly of vascular aetiology, preceded cavi-
tation 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. Syringes associated with primary cord tumours may be formed in the same way, rather than by fluid exudation as has been suggested.

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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.1-2 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,3 but spinal haemangioblastomas presenting in this manner have only been described twice previously.4-5 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. 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 thyrocervical vessels showed a vascular intramedullary tumour at C2 (fig). Laminctomy 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 post-operative 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.2 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.5 Of two previously described lesions which presented as SAH, one was extramedullary at L2 and the other was an intramedullary cervical lesion.2 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.6-7 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 intracranial 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 intracranial 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.

Fig Vertebral angiogram, anteroposterior projection, showing vascular mass.

References