Missed Diagnosis

Cervical spondylotic myelopathy with occult foramen magnum meningioma – a case for MRI?

K. Chakravarty, D.G.I. Scott, ¹D. Dick and ²J. Visick

Department of Rheumatology, ¹Department of Neuromedicine, ²Department of Radiology, Norfolk & Norwich Hospital, Norwich, UK

Summary: A fatal case of occult foramen magnum meningioma is described which presented with features of cervical myelopathy confirmed by cervical myelogram. Surgical decompression led to initial improvement followed by progressive deterioration and death. Postmortem examination revealed a 4 cm diameter foramen magnum meningioma. The dangers of using only standard radiographic investigations of the cervical cord are discussed.

Introduction

Tumours of the foramen magnum often present with ill-defined symptoms mimicking multiple sclerosis, syringomyelia, normal pressure hydrocephalus and degenerative diseases of the spinal cord.¹ We describe a patient with a benign tumour of the foramen magnum who presented with features of cervical myelopathy with myelographic evidence of cord compression especially at C5 and C6.

Case report

A 76 years old, previously fit woman presented with a 10 week history of coldness and numbness of her hands. She also admitted to progressive lower limb stiffness and an unsteady gait. She had no headaches, sickness or vomiting and no visual, bladder or bowel disturbances. Physical examination revealed distal upper limb weakness and impaired joint position sense in her hands with mild symmetrical paraparesis. Cranial nerve and fundal examination was normal.

Investigations revealed a haemoglobin of 12.1 g/dl, white cell count $4.6 \times 10^9/l$ with normal platelet count and erythrocyte sedimentation rate. The following investigations were normal or

negative: urea and electrolytes, liver and thyroid function tests, bone chemistry, syphilis serology, serum B₁₂ and red cell folate. The antinuclear factor was negative but anti-DNA antibody was raised at 94 IU/l (NR <50 IU/l). Rheumatoid factor, complement levels, creatine kinase and antibody to extractable nuclear antigens were within the normal range.

A preoperative cervical spine X-ray showed severe degenerative changes at C5/6 and C6/7 (Figure 1). The chest radiograph and the electrocardiogram were normal. A cervical myelogram (Figure 2 and 3) revealed marked indentation of the cord between C5/6 and C6/7 due to large osteophytes suggesting considerable cord compression. Cerebrospinal fluid analysis was normal except for a slightly raised protein of 0.75 g/l (NR 0.10–0.40).

She underwent cervical laminectomy and decompression with good response apart from some initial postoperative dysphagia, thought to be due to delay in the lift of the larynx as a result of possible damage to C3, 4 and 5 nerve fibres. Her dysphagia later improved allowing her to swallow food normally.

However, 6 weeks postoperatively, her dysphagia returned and she became progressively weak and immobile with episodes of choking. Her clinical course was of rapid and progressive deterioration and she died within 10 days.

A postmortem examination revealed a 4 cm firm tumour at the foramen magnum in front of the

Correspondence: K. Chakravarty, M.B., M.R.C.P., Rheumatology Research Unit, E6, Addenbrooke's Hospital, Cambridge CB2 2QQ, UK.

Accepted: 20 January 1993

Figure 1 Preoperative X-ray of cervical spine showing severe degenerative changes. (a) Anterior-posterior, (b) lateral.

medulla oblongata and the histology confirmed a meningioma. The spinal cord was atrophic with a distal area of infarction in the upper thoracic region. The cervical spinal cord appeared unremarkable.

Discussion

Our patient illustrates the problems associated with the diagnosis of benign tumours at the foramen magnum, particularly in the presence of cervical spondylosis. Cervical myelography gave no specific information above the lower cervical cord and in retrospect was potentially dangerous because of the risk of coning.

The clinical presentation of foramen magnum meningioma is varied and neurological examination rarely demonstrates signs indicating the exact localization of the pathology. In one series 66% of patients presented with only neck pain, often in the C2 nerve root distribution, 49% with gait disturbances, 22% with bladder dysfunction and only 12% with dysphagia.¹ Our patient developed pronounced dysphagia in the immediate postoperative period followed by gradual improvement, only to

recur as a preterminal event. It is possible that the severe post-surgical dysphagia was related to tissue oedema adjacent to the primary lesion associated with increase in cerebrospinal fluid pressure which improved with wound healing. The late relapse probably reflected progressive herniation of the tumour into the foramen magnum resulting in coning and death.

Our case highlights the risks of standard investigative procedures such as cervical myelography in elderly patients with clinical features of cervical myelopathy. Cervical myelography is essentially an invasive in-patient procedure and requires considerable time and experience of the radiologist with its attendant risks, as evident in our case. In one study computerized tomography (CT) scanning with intravenous contrast was diagnostic in 75% of cases. Another study demonstrated that metrizamide myelography and CT scanning was more informative than myelography alone.² However, myelographic demonstration of the tumour requires complete contrast filling of all portions of the subarachnoid space, requiring more time than a 'routine' procedure. CT scanning is marginally superior to the myelography but the medulla and the cervical spinal cord are virtually never shown

Figure 2 X-ray (lateral-cervical myelogram) showing severe compression at C5/6.

well in absence of bony changes, calcification or alteration with contrast enhancement.³ Previous studies have recommended magnetic resonance imaging (MRI) as the most accurate noninvasive and cost-effective way of evaluating abnormalities of the cranio-cervical junction.⁴⁻¹²

A recent survey from a regional neurological centre has shown a significant change in diagnosis (20%) and management (27%) following MRI scan.¹³ It was also shown to be cost effective if other radiographic procedures could be reduced in the run up to MRI with a slightly higher risk of inappropriate request for MRI scans. A similar

Figure 3 Anterior posterior view of the cervical myelogram.

study¹⁴ from Cambridge has shown that MRI scan of the neck is less expensive than a CT myelogram which includes in-patient stay for at least 24 hours in uncomplicated cases.

It is clear from our case that MRI scan would have changed the diagnosis and saved the patient from unnecessary surgery. Perhaps a correct diagnosis would have helped her to undergo the appropriate surgery which could have saved her life. It is important to recognize that foramen magnum meningiomas are totally resectable and a good quality of life can be expected postoperatively. MRI scan is clearly the investigation of first choice in patients with cervical myelopathy and should replace other older investigations.

Acknowledgement

We are grateful to Mr Waters for his help with the surgical management of the patient.

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