

TUMOURS IN THE REGION OF FORAMEN MAGNUM

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Abstract

A very unusual case of a neurofibroma extending from the level of the fifth cervical vertebra, through the foramen magnum into the posterior cranial fossa was operated on by the author in July 1978 and is reported here. The world literature on the subject of foramen magnum tumours is reviewed. Benign extramedullary tumours of the foramen magnum are rare and when they do occur they are likely to remain undiagnosed until tragic neurological sequelae occur, because their modes of presentation are generally not known and appreciated. And yet most of them are surgically removable if detected before severe neurological deficit occurs. The clinical presentation of the tumours in the region of foramen magnum frequently mimics multiple sclerosis, cervical spondylosis, intramedullary tumour, syringomyelia and even carpal tunnel syndrome.

Introduction

In 1929, Elsberg and Strauss reported six extramedullary tumours in the region of foramen magnum. The first comprehensive study of benign and malignant intramedullary and extramedullary tumours in this region by Love et al (1954), revealed that 40% were surgically treatable extramedullary tumours. This study also showed that benign extramedullary tumours with an excellent prognosis are far more frequent than the malignant extramedullary tumours.

The earliest contributions to the study of the clinical features in benign extramedullary tumours of this region are those of Abrahamson and Grossman (1921) and Elsberg and Strauss (1929). The most recent work on this subject is that of Yasuoka et al (1978) who studied 57 patients with benign extramedullary tumours of the foramen magnum (19-neurinomas, 37 meningiomas and one teratoma) who were operated on at the Mayo Clinic during 20 years period.

Foramen magnum tumours are of great interest to the neurologists and neurosurgeons for the following reasons:

1. Although surgically treatable with excellent prognosis, these tumours are likely to remain undiagnosed until serious and tragic neurological sequelae occur, because modes of presentation are not generally known or appreciated.
2. Even at a late stage when serious neurological deficit is present, they are misdiagnosed as another disease particularly cervical spondylosis and multiple sclerosis with the result that incorrect treatments are applied with tragic results.

To avoid these pitfalls in the diagnosis of the foramen magnum tumours the commonest modes of presentation and helpful investigations are summarized: Mayo Clinic series (Yasuoka et al., 1978) which is the largest and most authentic yet published on the subject is taken as a source of reference regarding the relative frequencies of various presenting signs and symptoms

Presenting Symptoms

The commonest presenting symptoms are paraesthesias of fingers and hands and pain in the neck and sub-occipital region. Other symptoms are gait disturbance, clumsiness of hands, weakness of extremities and bladder disturbances in descending order of frequencies. Numbness and tingling of the hands was complained by 95% and pain in neck and suboccipital region by 75% of cases in the Mayo Clinic series.

Neurological Signs

A-Motor Deficit

Weakness of extremities progresses through various patterns to ultimate dense and uniform tetraplegia-when it is probably already too late to diagnose. For early diagnosis it is important to appreciate earlier patterns of weakness extremities. Patients present with following patterns of limb weakness in descending order of frequency: Minimal to moderate weakness of all the four extremities, hemiparesis or hemiplegia, monoparesis of one upper limb and weakness of both upper limbs. In the series under consideration no patient presented with monoparesis or paraparesis of the lower limb. When patient presents with hemiparesis or tetraparesis upper extremity is always weaker than the lower.

B-Sensory Deficit

Sensory loss is usually more prominent in upper limbs than in lower limbs. Hypalgesia of the C-2 region when present may be diagnostic of the foramen magnum tumours. Patterns of sensory deficit in descending order of frequency are: Dissociated anaesthesia, sensory loss below C-5, and brown sequard syndrome.

C-Reflexes

Exaggerated deep tendon jerks in both upper and lower limbs are present in most of the cases and babinski's sign is positive in majority of cases.

D-Cranial Nerves

Palsy of 11th cranial nerve is often present. Other cranial nerve involvement is extremely rare.

Investigations

Myelography: is the investigation of choice. Incomplete myelographic studies leading to negative results and misdiagnosis are frequent. Complete myelographic study high up into the foramen magnum anteriorly and posteriorly is essential to avoid misdiagnosis.

Case Report

This 39 year old lady was admitted to the Neurosurgical Unit in July 1978 with almost complete tetraplegia and a loss of all modalities of sensation to a level just below the clavicles. Prior to admission here she was treated at various other hospitals as a case of carpal tunnel syndrome, cervical spondylosis and multiple sclerosis at different times. On admission her general condition was very poor with pressure sores at the sacrum.

History of her illness dated back to two years when she started getting pain in the neck and suboccipital region. Two months later she reported to a peripheral hospital with tingling and numbness of fingers of both hands. Progressive weakness and wasting of both the arms soon followed and weakness of both lower extremities was noticed six months later. Weakness, more in upper extremities than lower steadily progressed until she was unable to walk and bed-ridden one year prior to the admission here. At the time of admission she had been densely tetraplegic for the previous six months and had lost bladder control for the same period.

On Examination

There was severe weakness of all the four extremities. Sensory loss was complete and extended upto a level just below the clavicles. Deep tendon reflexes were increased in both upper and lower extremities. Plantars were up-going bilaterally. Palsy of 11th cranial nerve and hypalgesia in C-2 region was noted. Plane x-rays of the cervical spine showed mild osteoarthritic changes. Myelography showed a complete block at C-6 level with a crescentic margin to the myodil column at the level of the block (Fig. 1).



Fig. 1(a) : X-ray Cervical Spine

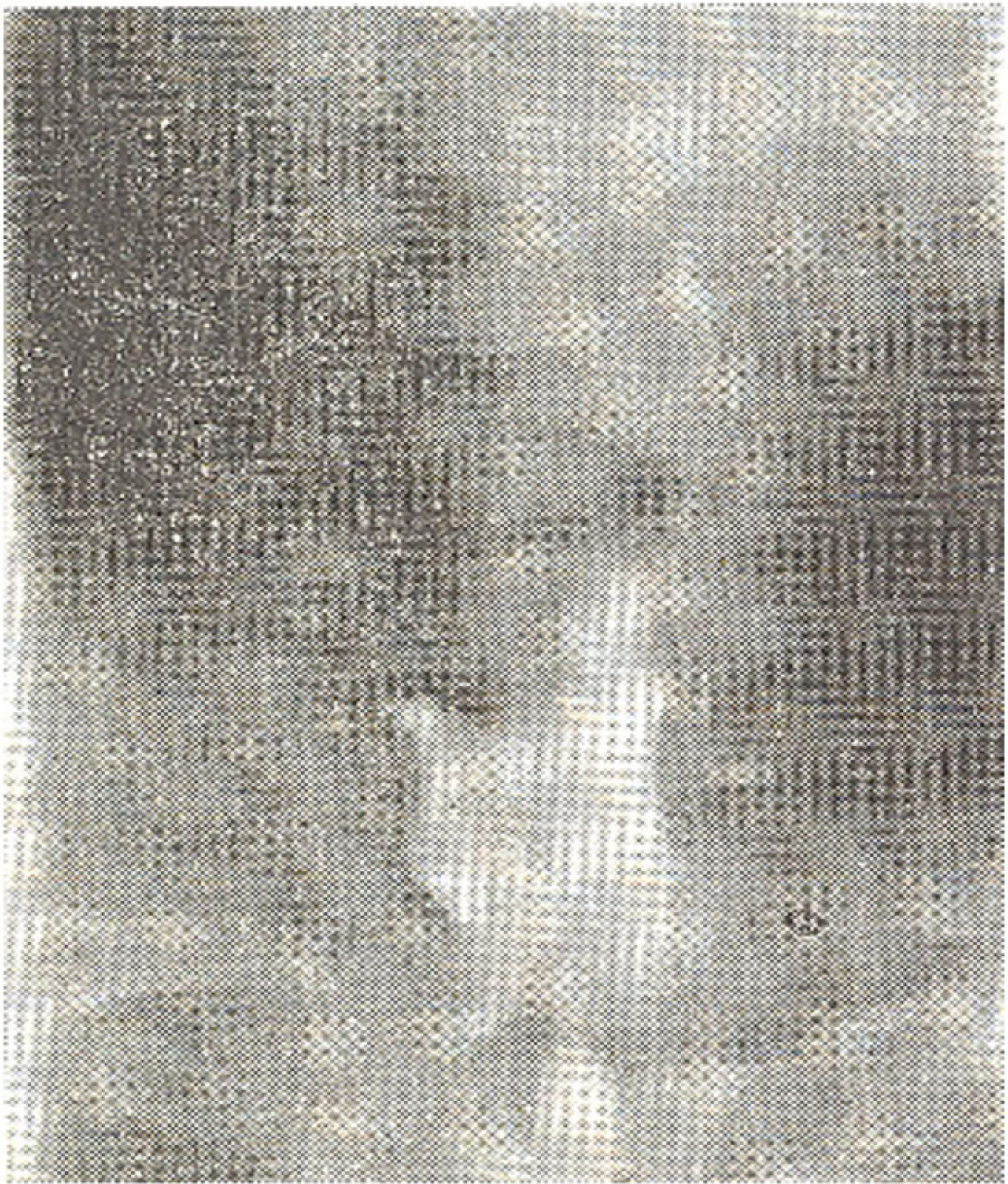


Fig. 1(b) : Myelograms

A pre-operative diagnosis of cervical tumour was made and a cervical laminectomy extending from C-7 to C-3 was carried out. On opening the dura a small arachnoid cyst was found to be covering the lower pole of a postro-laterally situated tumour, the upper limit of which could not be reached through this

exposure. The incision was consequently extended upwards to the level of external occipital protuberance and the spine and laminae of axis and posterior arch of the atlas were removed and dural incision extended to the level of the foramen magnum. Upper limit of the tumour could still not be reached as it was found to be entering the posterior cranial fossa through the foramen magnum. Partial suboccipital craniectomy was carried out to deliver the intracranial portion of the tumour. Origin of the tumour could be traced to the 3rd cervical root on the right side.



Fig. 2 : Operative Findings

Fig. 2 shows the operative view after incision of the dura. Lower pole of the tumour is clearly seen as well as the dilated vessels on the surface of the cord.

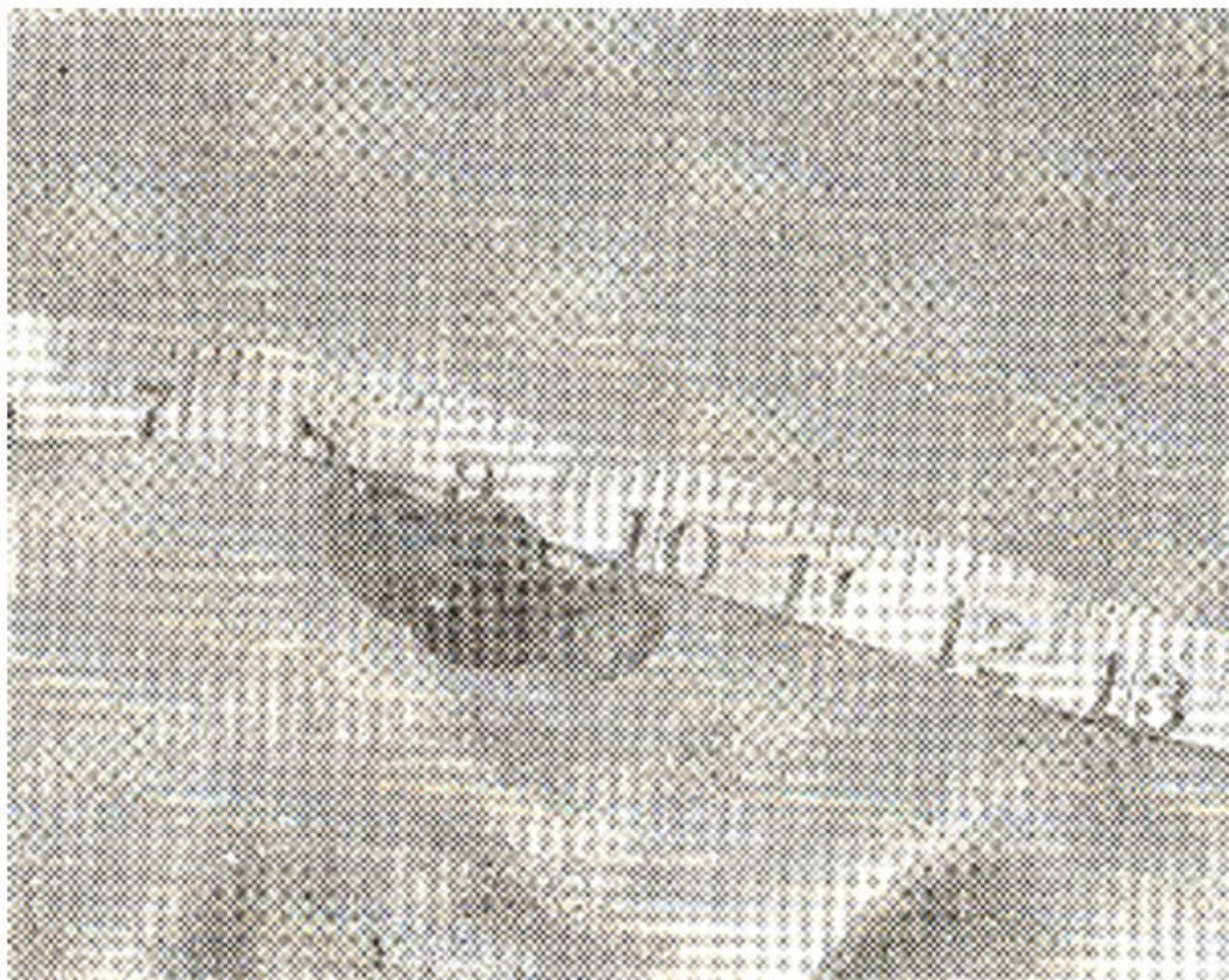


Fig. 3 : Specimen of tumour

Fig. 3 shows the operative specimen of the tumour completely removed and folded on itself. Slight constriction marks the site of the foramen magnum-the smaller portion above the constriction was intracranial and the larger bulk below the constriction was the intraspinal portion. The lower pole of the tumour extended to the level of C-5 and the capping arachnoid cyst explains the level of myelographic block at C-6.

Biopsy showed the tumour to be a neurofibroma. Post-operative course was uneventful. There was a gradual and remarkable return of motor and sensory functions. Three months after the operation sensation was restored almost to normal and she could walk with help. A follow-up one year after the operation showed that she was carrying out her normal daily activities.

Discussion

All the 19 neurinomas reported from the Mayo Clinic were spino-cranial:-18 arising from C-2 nerve root and only one from C-1 root. The case reported here is unique as none reported yet arose from C-3 root and extended as low as C-5 level and upwards through the foramen magnum into the posterior cranial fossa. Although early diagnosis before severe neurological deficit is desirable to prevent tragedies; this case shows that results of surgical removal even at this late stage (with complete

tetraplegia, complete loss of sensation and bladder control) can be quite dramatic and surgery always be offered even in very late cases.

The clinical appearance of foramen magnum tumour is protean leading to misdiagnoses. Search of the world literature reveals that the commonest and typical course of foramen magnum tumour is as follows: In early stages-pain in neck and suboccipital region and paresthesias of upper extremities develop followed by clumsiness of hands, disturbance of gait, progressive weakness of all the extremities and bladder disturbance follows. Sensory loss and weakness is more pronounced in the upper extremities than the lower. Hypalgesia of the C-2 region and the 11th cranial nerve palsy when present are the most reliable localizing signs.

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