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Benign Extramedullary Foramen Magnum Tumours

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Abstract

This report reviews the clinical features, operative data and outcome of 32 cases of benign extramedullary foramen magnum tumours. There were 17 meningiomas, 10 schwanomas and 5 neurofibromas. The most frequent initial complaints were neck and occipital pain (46.9%), dysesthesia of one upper limb (21.9%) and one upper limb weakness (12.5%).

The average time from initial symptom to diagnosis was 14 months. All the patients had motor weakness at admission and 18 (56.2%) cases were bedridden because of gross motor weakness. Contrast myelography was positive in all cases except one. Twenty five cases (78.1%) had good recovery and returned to normal work without significant disability. On follow-up tumour recurrence occurred in 4 cases (12.5%). The over-all mortality was 15.6%. The mortality rate appears to be higher in patients with pre-operative respiratory dysfunction and in those with tumour recurrence. The importance of early diagnosis and elective post-operative ventilation especially in patients with pre-operative respiratory dysfunction is emphasised.

Key words -

Foramen magnum tumour,
Meningioma,
Schwannoma,
Respiratory dysfunction,
Extramedullary tumour,
Benign

Extramedullary tumours at foramen magnum occur rarely. They are often misdiagnosed due to bizarre initial clinical symptoms and slow course of progression of symptoms. Elsberg and Strauss [1] found six (3.2 %) extramedullary tumours at foramen magnum region in his series of 185 tumours of the spinal cord. Love and Adson [2] reported that 40% of tumours of foramen magnum region were extramedullary and benign extramedullary tumours were more frequent than malignant tumours. Benign extramedullary tumours constituted 30 % of tumours in this region [3].

The earliest contribution to the clinical diagnosis of tumours in foramen magnum region are those of Abrahamson and Grossman [4] and Elsberg and Strauss [1]. Since then a number of reports have emphasised the bizarre initial clinical features leading to wrong localisation [5], [6], [7], a prolonged and relapsing course leading to erroneous diagnosis [3], [8], [9], [10], [11], [16] and incomplete myelographic studies leading to negative results and misdiagnosis [12], [13], [14], [15].

There are only a few published reports reviewing more than 20 cases of benign extramedullary tumours at foramen magnum [2], [3], [10], [11]. A significantly higher percentage of patients, presenting with gross neurological disability and respiratory dysfunction prompted us to review our case material, especially with respect to the outcome.

This report includes a detailed analysis of observations in 32 cases with tumours at foramen magnum.

Clinical Material

Between 1958 and 1983, 32 patients with benign extramedullary tumours at foramen magnum were treated at NIMHANS, Bangalore. Only those cases with tumours extending to both posterior fossa and intraspinal compartments have been included in this series. All patients were operated and the diagnosis confirmed. The pathological diagnosis was meningioma in 17, Schwanoma in 10 and neurofibroma in 5 cases.

Majority of patients were in 2nd and 4th decades of life at admission to hospital. The age group ranged from 11 - 70 years. There was no sex preponderance. The time lapse between the onset of initial symptom and diagnosis ranged from 8 days to 6 ½ years, with a mean duration of illness of 14 months.

Clinical Features

The most frequently encountered initial symptom (table I) was pain in the upper part of the neck and occipital pain (in 15 cases). Seven cases had dysesthesia of one upper limb as initial symptom. In 4 cases the initial symptom was unilateral upper limb weakness. Numbness in hands (1), dysesthesia involving both lower limbs (2), giddiness (1) and numbness in both feet (1) were other initial symptoms.

Table I - Initial Symptom

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Prolonged interval between initial symptom and progression of deficit often lead to erroneous diagnosis of cervical spondylosis with myelopathy, and even tension headaches in some of the patients.

The motor deficit (Table II) followed the "typical" pattern of progression in 18 cases involving one upper limb first, progressing to involve the ipsilateral lower limb, the contralateral lower limb and finally to involve the opposite upper limb. The progression was atypical in 14 cases.

Table II - Clinical Features

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All the 32 cases had motor deficits at the time of admission. Asymmetric quadriparesis (62.5%), symmetric quadriparesis (31.3%) and hemiparesis was seen in 2 cases. Hemiparesis in the absence of sensory deficit can lead to problems in clinical localisation. However the absence of facial involvement lead to localisation of a spinal cord compression. Hyper-reflexia was found in all the patients. Wasting of small muscles of hand bilaterally was seen in 6 (18.8%) cases. In one patient there was wasting of muscles on one side of the whole upper limb.

Sensory loss was detected in 23 cases only. There was graded sensory loss from C2 downwards in 14. Graded sensory loss below C5 or C7 dermatome level was seen in 7 cases and cape like sensory loss was seen in 2 cases.

Spinothalamic sensory loss was detected in 18 cases, and loss of joint and vibration sense in 19 cases. No dissociated sensory loss was seen in any of the patients in this group. The classical steroanaesthesia with "piano playing fingers" was seen in one case only.

Two patients had papilloedema and other features of raised intracranial pressure. There was no obstruction to the 4th ventricular outlet in these patients. CSF protein levels were elevated in both the patients. However minimal ventricular dilation was present in one patient on investigations. Accessory nerve palsy was present in 4 patients, ipsilateral sluggish corneal reflex was seen in 8 cases. In one patient the gag reflex was sluggish and in another there was ipsilateral 12th nerve involvement. One patient had episodes of unconsciousness associated with neck flexion.

Bladder dysfunction in the form of hesitancy, precipitancy or reflex neurogenic bladder was seen in 14 cases and 9 patients were in respiratory distress at the time of admission to hospital. In all 18 cases were bed-ridden at admission to hospital due to gross neurological deficits. Evidence of neurofibromatosis was there in 4 cases.

Diagnostic tests

Plain x-rays of the cervical spine were normal in 29 cases. In 3 cases there was widening of intervertebral foramen between C1-2. Myodil myelography was positive in 31 cases. It was however normal in one patient. This patient had multiple neurofibromatosis along with widening of intervertebral foramen between C1-2 in plain x-rays.

CSF protein levels were elevated in 24 cases, the highest level being 480 mg%. Only one patient among these had minimal ventricular dilation. C.T. scan of the foramen magnum and upper cervical spine was done in one case.

Surgical techniques

A wide high cervical laminectomy and a small suboccipital craniectomy has been the standard operative approach in all cases. Except in the last 4 cases, all the patients were operated in sitting position. It is our practice at present to operate on cervical spine and posterior fossa lesions in recumbent position.

At surgery, 6 tumours were extradural and 26 intradural extramedullary in location. Among these, tumour location was anterior to the cervico medullary junction in 5 cases, anterolateral in 10, posterolateral in 9 and posterior to the cord in 2. To achieve better exposure of tumour, dentate ligaments were sectioned, in anteriorly or anterolaterally located tumours. Dorsal upper cervical nerve roots were sectioned for exposure of tumour whenever felt necessary. Tumours were removed piecemeal. Total tumour removal was achieved in 30 cases. An operating microscope was used in tumour removal in most cases operated after 1979. It has been particularly useful in disecting blood vessels free of tumour tissue.

Operative mortality was 15.6% (5 cases), out of which 2 cases had surgery for tumour recurrence. Three patients died of respiratory failure. All of them had pre-operative respiratory insufficiency. One patient died suddenly on the 5th post-operative day wherein no obvious cause of death could be ascertained and autopsy was not available. One patient died of CSF leak and meningitis.

Four patients underwent surgery for recurrence. Tumour removal was partial at first surgery in two of these cases. In one case of meningioma there was malignant change at recurrence.

The survivors were followed-up for a period ranging from 1-24 years with a mean follow-up of 29 months. Among the survivors 15 (46.9%) had no deficits and 10 had minimal deficits but were gainfully employed. Only 2 patients had persistent gross disability necessitating assistance in activities of daily life.

Discussion

There are a number of reports on foramen magnum region tumours, wherein both intra and extramedullary neoplasm are included [2], [8], [18].

Cushing and Eisenhardt [19] advocated that the term foramen magnum tumour be restricted to neoplasms extending into both the posterior fossa and cervical canal. Since there are few reports where the above criteria were adhered to in patient selection [3], [9], [10], [16], [20].

Benign tumours of the foramen magnum are mostly meningiomas or schwanomas [3], [7], [8], [9], [19], [21], [22]. Rarely other tumours such as dermoids [20], teratoma, lipoma, menigeal melanocytoma [8], [9], [18] etc., have been reported. In our series there were 17 meningiomas, 10 schwanomas and 5 neurofibromas.

There was no sex preponderance in our series unlike in other series [3], [11], where a female sex preponderance was found. The initial clinical symptom is characterised by suboccipital and neck pain, unilateral upper limb weakness or paresthesia in most cases. The 'typical' progression of motor deficits involving one upper limb followed by ipsilateral lower limb, and subsequent contralateral lower limb and upper limb involvement, was seen in 56.2% of cases.

Other reported findings are horizontal nystagmus, lower cranial nerve palsy especially the 11th cranial nerve, and rarely other lower cranial nerves and exceptionally the motor branch of 5th nerve [3], [7], [10], [11], [19], [22].

Atrophy of small muscles of hands was present in 18.8 % of cases. Though the pathogenesis of this finding is not clear, various explanations [3], [9], [10], [11] have been offered. They are -arterial ischemia in lower cervical cord, venous congestion, edema and hypoxia, a secondary syringomyelia as a result of blockage to flow of CSF and mechanical stresses because of anchoring of cord by dentate ligaments affecting lateral columns and anterior horn neurones either directly or by compromising blood supply.

Sensory examination in C2 root distribution is important [3], [10], [11] for clinical localisation. Hypoesthesia was seen in C2 dermatome distribution in 43.8% of our cases. Presence of stereoanaesthesia (1) [9], [11], [21], cerebellar signs (1) papilloedema (2) have been uncommon in our series.

Yasuoka et al. [11] have divided the clinical features of extramedullary foramen magnum tumours into

two stages. In the early stage the clinical findings are suggestive of of cervical spondylosis and in the advanced stage the neurological examination is suggestive of an intramedullary lesion. Though most of our patients came to the hospital in an advanced stage of the disease, there was clinical suspicion of intramedullary tumour in only 2 cases.

Respiratory dysfunction and sphincter disturbances appear late in the clinical picture [3]. Symptoms of neurogenic bladder dysfunction was seen in 43.8% cases and respiratory distress in 27.5% cases at admission. Similarly a large number of patients had severe disability and were bed-ridden at admission.

Plain radiographs were normal in 90.6% of cases and CSF protein was elevated in 75%. Myelography can give false negative results if not examined carefully. This is due to the larger anterior subarachnoid space and cisterna magna. The examination must be done in prone, supine and in lateral inclination maneuvre positions in cases with suspicion of foramen magnum tumours [13], [15], [23]. Vertebral angiography [14], [24] is a useful adjunct prior to surgery to delineate the relationship between the tumour and the vascular structures in the region.

Recently C. T. scan along with metrizamide myelography is being increasingly used to demonstrate the lesion. However our experience is limited to one case. Current reports suggest that MRI [10], will become the method of choice for investigating foramen magnum lesions, as anatomical display and ease of examination make it superior to current imaging modalities.

Despite the advanced stage of the disease in majority of our patients results were excellent in 78% of cases. The mortality rate of 15.6% (Table III) was because of severe pre-operative disability and respiratory insufficiency pre-operatively, in a higher percentage of patients. Routine post-operative ventilatory assistance is recommended in patients with pre-operative respiratory dysfunction.

Table III - Results

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Mullan et al [25] advocated transoral approach to intradural tumours at foramen magnum. Though we advocate this approach for extradural foramen magnum lesions, we have not used it for intradural tumours. We have found posterior approach combined with microsurgical techniques adequate to excise all tumours in this region. It should be emphasised that attempt should be made to excise the tumour totally at first surgery, as mortality for reoperation is high in our series.

Despite the advanced stage of the disease a majority of cases made total recovery, provided good pre and post-operative care is offered. However the mortality and morbidity could be reduced further if the disease is diagnosed early. The use of ultrasonic tumour aspirator will be a useful adjunct to the magnification of microscope in the total excision of these tumours.

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