

# Meningioma at the Craniocervical Junction.

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## Abstract

We report a case presenting progressive weakness of the limbs, paresthesia, disesthesias, and stiffness of the neck, radicular pain and vertigo. A multiplanar, multisequence MRI-spine showed a well defined dural based extra-medullary mass in the craniocervical junction anteriorly with intense homogenous enhancement post contrast administration. The proximal cord is effaced and displaced antero-laterally to the left by this lesion. The right vertebral artery is encased by the mass. There is pathological cord hyperintensity extending from the inferior medulla to C2 vertebral body in keeping with compressive myelopathy. A tumor was partially removed and pathological study concluded as meningioma. We review the medical literature to update our current knowledge about this topic.

## INTRODUCTION

Most of craniocervical junction (CCJ) disorders are congenital such as: atlas assimilation (fusion of the atlas and occipital bone that produces symptoms of cervical myelopathy when the anterior-posterior diameter of the foramen magnum behind the odontoid process decreases to less than 19 mm), atlas hypoplasia, Arnold-Chiari malformations (cerebellar tonsils or vermis descend into the cervical spinal canal), Klippel-Feil type I syndrome (fusion of cervical vertebrae usually asymptomatic except for a neck deformity with limited range of motion), achondroplasia Down syndrome, Platybasia (asymptomatic flattening of the skull base, and the angle formed by the intersection of the plane of the clivus and the plane of the anterior fossa should be less than 135° on lateral skull x-ray). Morquio's syndrome (mucopolysaccharidosis IV), or osteogenesis imperfecta causing atlantoaxial subluxation/dislocation (displacement of the atlas anteriorly in relation to the axis causing acute or chronic spinal cord compression). A craniocervical abnormality should be considered when fixed or progressive neurologic deficits refer to the lower brain stem, high cervical spinal cord, or cerebellum. Skull-base softening associated with some of these syndromes results in acquired basilar invagination or basilar impression (protrusion of the odontoid process into the foramen magnum leading to a short neck and combinations of cerebellar, brain stem, lower cranial nerve, and spinal cord signs).; other acquired abnormalities are: injury or disease that may involve occipital bone (foramen magnum/clivus), the first two

cervical vertebrae that decrease the space for the brain stem and cervical cord, ligaments, or both (<19 mm), and are usually caused by vehicle or bicycle accidents (50%) recreational activities, and falls particularly diving (35%) and other (15%). Such injuries may be osseous (fractures), ligamentous (luxations), or complex (subluxation of C-2, transaxial cervicomedullary junction injury, and osteoligamentous disruptions). Of course, minor neck injury can precipitate variably progressive symptoms and signs in patients with an underlying craniocervical junction abnormality mainly in patients with Klippel-Feil syndrome [1] Other causes of acquired abnormalities are: Rheumatoid arthritis, metastatic tumours, Paget's disease of the cervical spine, slowly growing CCJ tumours (neurofibroma, schwannomas, meningioma, and chordoma) that can impinge on the brain stem or spinal cord. A very important contributing factor for craniocervical syndrome is that neural tissue at this level is flexible and susceptible to slow and gradual displacement or compression. Rheumatoid arthritis and Paget's disease can cause basilar invagination, which can compress the spinal cord or brain stem. Regarding to investigations, CT after intrathecal administration of contrast dye provides anatomic detail of the neural structure abnormality and associated bony distortion but MRI is the investigation of choice to confirm the standard parameters of basilar impression.[2,3] and sagittal MRI also best identifies associated neural lesions (hindbrain herniation, syringomyelia, and vascular abnormalities) but can be preceded by plain x-rays (lateral view of the skull showing the cervical spine, anteroposterior view, and

oblique views of the cervical spine) which are used to identify factors that influence treatment including reducibility of the abnormality, bony erosion, the mechanics of compression, and the presence of abnormal ossification centers and epiphyseal growth plates with anomalous development. Looking for McRae's line and Ranawat's line in the initial screening for basilar impression is also recommended and Chamberlain's line and McGregor's line are viable alternatives, but clinicians must be aware of the potential for false positives. [2] Other cause of CCJ's disorder is Dubowitz syndrome that is a disturbance involving craniofacial abnormalities, growth retardation and mental retardation. Approximately 142 cases have been reported, with various associated other anomalies. These include cardiovascular, urogenital and endocrine abnormalities, as well as a predisposition to infections and haematological malignancies. Scoliosis has been described in association with this syndrome, as have isolated vertebral abnormalities. There has, however, been no description of craniocervical abnormalities. Swartz et al reported three Dubowitz patients with significant craniocervical abnormalities. [4] Among infectious disease affecting the craniocervical junction tuberculosis is rare but may cause significant complications with severe sequelae due to the nearby presence of important neurovascular structures [5]. Severe degeneration in the atlantooccipital joints appears to be a rare condition, with no degeneration found in the occipital condyles and no changes in the superior articular cartilage of the atlas. However degeneration of the inferior articular cartilage of C-1 and the superior articular cartilage of C-2 indicates that the atlantoaxial joint faces more intense mechanical exposure, which is increased at the upper joint surfaces [6, 7].

In 2008, Menezes [8] reviewed all the symptoms presenting in children with CCJ abnormalities and found that children with craniovertebral abnormalities present with failure to thrive, weakness, basilar migraine, dysphagia, sleep apnea, scoliosis, and the cervical central cord syndrome. Torticollis was seen in the very young. Unexplained neurological symptoms and signs associated with craniovertebral abnormalities in children require angiography. The initial treatment of bony lesions at the craniocervical junction consists of posterior decompression with enlargement of the foramen and removal of the posterior arch of the atlas and axis vertebra. This is regardless of the site of compression. These abnormalities can result in neckache; hydro-syringomyelia; cerebellar manifestations, lower cranial nerve signs, and spinal cord deficits; and cerebro-vascular

manifestations on the vertebrobasilar territory. Vertebral angiography or MRI is used selectively to identify fixed or dynamic vascular compromise. Treatment often involves reduction, followed by stabilization with surgery or an external device.

Non-missile penetrating spinal cord injuries are uncommon, and involvement of the CCJ is even less frequent and its prognosis is not usually good, however Elgamal [9] reported a case of 42-year-old male who presented with quadriplegia immediately following stab injury inflicted with a kitchen knife to the back of his neck. The knife was retained in the patient's neck but after retrieval of the knife, his quadriplegia recovered.

Several types of cranio-cervical abnormalities can cause or contribute to cervical spinal cord or brain stem compression. Fusion of the atlas (C1) and occipital bone causes spinal cord compression if the anteroposterior diameter of the foramen magnum behind the odontoid process is narrowed. Basilar invagination (upward bulging of the occipital condyles) results in a short neck and compression that can affect the cerebellum, brain stem and cranial nerve deficits include sleep apnea, internuclear ophthalmoplegia (ipsilateral eye adduction weakness and horizontal nystagmus in the contralateral abducting eye on lateral gaze), and spinal cord. Atlantoaxial subluxation or dislocation (displacement of the atlas anteriorly in relation to the axis) causes acute or chronic spinal cord compression. The Klippel-Feil malformation (fusion of cervical vertebrae) deforms and limits motion of the neck but usually does not have neurological consequences. Platybasia (flattening of the skull base so that the angle formed by the intersection of the clival and anterior fossa planes is  $> 135^\circ$ ), seen on lateral skull x-ray, may be asymptomatic or cause cerebellar or spinal cord deficits or normal-pressure hydrocephalus.

Various types of bone tumours may be found at the CCJ. Confusion between benign and malignant tumours or pseudo tumours must be avoided, sometimes requiring a biopsy. Surgery using a lateral approach usually permits the surgeon to achieve a complete resection either preserving the stability of the CCJ whenever intact or associated with a stabilization procedure. Bone tumours located at the cranio-cervical junction (CCJ) are rare. Tumoral involvement of the neighbouring structures including bone, nerves and vertebral artery and the dynamic aspects of the bone structures raise technical difficulties in the surgical approach. The surgical management includes tumoral resection and stabilization of the CCJ [10]. Because the spinal cord is flexible and

therefore susceptible to intermittent compression, several types of lesions at this level can cause symptoms that vary from patient to patient and that can be intermittent. Because meningioma growth slowly despite its anatomopathological presentations at this region then no clinical differences can be easily identified except if additional vascular disturbance coexist. In this situation other considerations should be taken, being this aspect the main goal of this report.

### **CASE REPORT**

A 33-years-old female complaining of generalized body weakness of two months duration came to Neurology Unit at Nelson Mandela Academic Hospital in Mthatha, (South Africa). She was well until four months ago when she felt cramps on the right upper limb, and mild pain in the nape of the neck followed by weakness on the same side and inability to use her hand; few days later she felt cramps in the right lower limb and it progressed until she was unable to move that limb. This was followed by stiffness of the neck, sensation of "heaviness of the head" and mild radicular pain from the head to the upper back. She went for consultation several times but she was not getting better. Two months later she started to complain of cramps and tingling sensation on the left lower limb followed by weakness and inability to raise her leg and three weeks later same symptoms came to her left upper limb and she became unable to use her four limbs. From time to time those symptoms were associated with neck pain spreading to the arms and suboccipital headache radiating to the skull, subjective vertigo, dysarthria, incoordination, shortness of breath, and neckache. All symptoms worsened with head movement and can be precipitated by coughing or bending forward.

PMH: Arterial hypertension since April 2009. FH: Hypertension and tuberculosis. SH: non-remarkable information is obtained.

On physical examination: Mild pallor on conjunctiva, distended abdomen, soft, non-tender, no shifting dullness, no visceromegaly were found Nervous system: generalized hypotonia and areflexia, power 0/5 proximal and distally on four limbs, sensory level for light touch and pin-prick at C-1 and C-2. Lhermitte's sign is found (tingling down the back, and often into the legs, on neck flexion). Downbeat nystagmus (fast component downward) No other remarkable signs were found

All blood tests were normal and multiplanar, multisequence MRI-spine (Figure 1) showed a well defined dural based

extra-medullary mass in the CCJ anteriorly with intense homogenous enhancement post contrast administration. The proximal cord is effaced and displaced antero-laterally to the left by this lesion. The right vertebral artery is encased by the mass. There is pathological cord hyperintensity extending from the inferior medulla to C2 vertebral body in keeping with compressive myelopathy. Conus medullary and cauda equine nerve roots demonstrate no abnormality. Tumor was partially removed and pathological study concluded as meningioma

### **Figure 1**

Figure 1: Preoperative MRI enhanced with gadolinium revealed ventro-lateral intradural extramedullary mass lesion on the craniocervical junction. Pathological report was a meningioma.



### **COMMENTS**

Patients presenting progressive weakness of one upper limb followed weakness of the ipsilateral lower limbs and progression to contralateral lower one with an associated pain on the neck (attributed to compression of the C-2 root and greater occipital nerve and to local musculoskeletal dysfunction) should be considered as a candidate for a diagnosis of CCJ's tumour until proven otherwise. Slowly growing tumors at this region have different clinical manifestation according to different factors basically: vascular involvement. Meningioma (from meninges) and chordoma (arising from the remnants of the notochord) at the CCJ are the two most common tumour at this level and they produce symptoms by impinging on the brain stem and the spinal cord. Because of their deep location of chordoma,

its local infiltrative nature, and involvement of surrounding bone, treatment of chordoma is a challenge. [11] Bhadra and Casey [12] treated 175 patients with a chordoma over a ten-year period and they found that two had a family history of the condition. They undertaken a literature review of familial chordoma and found chromosomal abnormalities associated with the condition but in our opinion it is an anecdotal finding that will not modify its management and prognosis. Colli and Al-Mefty [11] analyze the data and prognostic factors obtained during the follow-up period (range 1-150 months, median 38 months) in 53 patients with craniocervical junction chordoma and found a better prognosis in patients with chondrosarcoma compared with those harbouring chordoma. Histological pattern and the patient's age at symptom onset were not factors that influenced prognosis in cases of chordoma. They also confirmed that extensive resection and possibly adjuvant proton-beam radiotherapy provided better prognosis for these patients.

As mentioned before, CCJ's tumors have different pathological natures, growth patterns and neurological involvement; appropriate surgical approaches facilitate tumor removal with less surgical morbidities; nevertheless recurrent tumors occasionally occur, so long term follow-up is mandatory. [13] Presentation varies because bony and soft tissue abnormalities in various combinations can compress the cervical spinal cord, brain stem, cranial nerves, cervical nerve roots, or their vascular supply. Vascular symptoms include syncope, vertigo, drop attacks, intermittent periods of confusion and altered consciousness, episodic weakness, and transient visual disturbance that may be provoked by moving or by changing head position. An abnormal head posture is common, and in some patients, the neck is short or webbed but we like to highline again that there is no single symptom or neurological finding pathognomonic for a lesion in this location [14] and because the generous size of the subarachnoid spaces at the cervicomedullary junction as well as the possibility of expansion of tumors into the high nasopharynx that makes symptoms arise only after the lesions have achieved large volume [15]. Stabilization with rigid screw/rod fixation is the treatment of choice for craniocervical disorders requiring operative stabilization [16]

The venous circulation of the CCJ, anchored by the suboccipital cavernous sinus, is well seen on contrast-enhanced, fat-suppressed T1-weighted MR images. The axial and coronal planes provide the best view. All the

venous structures studied were easily identifiable in all patients, except for the posterior condylar vein, which was highly variable in terms of size, symmetry, and presence. The sub occipital cavernous sinus, a vertebral venous plexus surrounding the horizontal portion of the vertebral artery at the skull base, provides an alternative pathway of cranial venous drainage by virtue of its connections to the cranial dural sinuses, the vertebral venous plexus, and the jugular venous system. Knowledge of the anatomy of this system facilitates interpretation of images and might reduce the number of false-positive diagnoses of lesions, such as adenopathy or schwannoma. [17]

### **MENINGIOMAS**

Occasionally, meningiomas may develop without dural attachment. Clear cell meningioma is a histological distinctive uncommon variant of meningioma that may behave aggressively with local recurrence and progression as well as cerebrospinal fluid-borne metastasis. Meningiomas near the craniocervical junction represent challenging tumours, requiring special considerations because of the vicinity of the medulla oblongata, the lower cranial nerves and the vertebral artery.[18-20]

Samii M et al [21] undertaken a retrospective analysis of 38 patients who were operated on for 40 meningiomas of the craniocervical junction between September 1977 and August 1995 to determine which factors influenced resectability, complications, and postoperative outcomes. They concluded that the relationship of the tumor to neighboring structures, i.e., the vertebral artery in particular, determines its resectability and recommend using extreme caution with recurrent or en plaque meningiomas and tumors associated with extensive arachnoids scarring although other problems have been reported. Primary meningioma occurring within the jugular foramen are exceedingly rare lesions

The experience of Roberti F et al [22] suggests that although posterior fossa meningiomas represent a continuing challenge for contemporary neurosurgeons, such tumors may be completely or subtotally removed with low rate of mortality and acceptable morbidity, allowing most of these patients to achieve a good outcome in a long-term follow-up.

Foramen magnum meningiomas are challenging tumors, requiring special considerations because of the vicinity of the medulla oblongata, the lower cranial nerves, and the vertebral artery.

Bruneau M and George B [23] classify meningiomas based

on the compartment of development, the dural insertion, and the relation to the vertebral artery. The compartment of development is most of the time intradural and less frequently extradural or both intraextradural. Intradurally, foramen magnum meningiomas are classified posterior, lateral, and anterior if their insertion is, respectively, posterior to the dentate ligament, anterior to the dentate ligament, and anterior to the dentate ligament with extension over the midline. This classification system helps to define the best surgical approach and the lateral extent of drilling needed and anticipate the relation with the lower cranial nerves.

The extent of the surgical approach to foramen magnum meningiomas must be based on the main point of dural attachment and tailored individually case-by-case. The differentiation between the clivospinal and spinoclivial types, as well as anterior and anterolateral types, is crucial for the neurosurgical planning of foramen magnum meningiomas.[24]

Removal of foramen magnum meningiomas can be performed safely today with the use of microsurgical techniques and intraoperative monitoring. The posterior suboccipital approach is suitable for the majority of these tumours [25]

In our limited personal experience, patients with CCJ's meningioma have good prognosis if the tumour can be removed completely that is usually possible if there is not an associate vascular deficiency, therefore early diagnosis is mandatory.

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