NEUROSURGERY CASE STUDY: Chiari Malformation (Congenital Cerebellar Tonsillar Herniation)

Asher Taban, MD
Medical Director,
Neuroscience
Institute

A 32-year-old white female patient presented with sudden onset of severe left peri-auricular pain with extension to the left side of the cranium, neck, and shoulder. The patient's discomfort was constant and did not respond to conservative measures. Additional complaints, such as left heel pain and sense of continuous tightness of her entire left upper extremity, including her hand and fingers, were reported. MRI scan of the brain and cervical and thoracic spine was performed. This disclosed very extensive cervical syrinx, extending from the odontoid to C7 spinal canal. This cyst occupied over 90% of the spinal canal in this region. There were also several septations within the cyst cavity. Also, low cerebellar tonsils and tight posterior fossa were noted without evidence of hydrocephalus. Her tentorium was found to be very steep. The cerebellar tonsils were at least 15 mm below the level of the foramen magnum.

Her past medical history was consistent with systemic hypertension, asthma, gall bladder and kidney stones.

Her examination revealed decreased sense of hot and cold temperature in the right side of her face, neck, lateral trapezious and chest wall. Subjectively, patient complained of a decreased sense of heat in her left cranium, neck and shoulder area. There were no sensory deficits in her lower limbs. No motor deficits or other abnormalities were discovered.

Patient underwent sub occipital craniectomy at the level of the foramen magnum and C1 laminectomy with partial resection of the right cerebellar tonsil and duraplasty with use of autologous pericranium and Gortex membrane. Patient's weight was 295 pounds. Her post-op course was uneventful. Shortly after surgery, the patient showed gradual improvement of her symptoms. Upon her last examination in December of 2010, her neurological examination was unremarkable. Her repeat MRI scan of the cervical spine revealed over 90% reduction of the cervical syrinx and increased spinal cord diameter with no more evidence of tight posterior fossa or foramen magnum being impacted.

Chiari malformation represents a hind brain abnormality, which consists of four different types. The majority are type I and II. Type I Chiari malformation consists of caudal dislocation of the cerebellar tonsils into the cervical spinal canal, which usually becomes symptomatic in young adults who mostly complain of **upper cervical pain**. This pathology may also present with hydrocephalus and spina bifida – myelomeningocele. In a patient with Chiari malformation, the cerebellar tonsils usually extend at least 5 mm. caudal to the foramen magnum without caudal migration of the medulla. Usually, there is no involvement of the brainstem, and lower cranial nerves are not elongated, nor are the upper cervical nerve roots coursed cephalad. Usually, arachnoid fibrosis and even pial adhesions are noted around the brainstem and cerebellar tonsils. Syringomelia of the cervical spinal cord may be present, but not frequently. There is a slight female preponderance, and the average duration of the symptoms is three years. However, if headache, which is more nonspecific, exists, then this group of patients could be considered symptomatic for over seven years prior to the discovery of the pathology. **Headaches, most of the time, are at the craniocervical junction.**

Overall, symptoms are due to either:

1) Compression of the brainstem at the level of the foramen magnum
2) Syringomyelia
3) Hydrocephalus

About 70% of the patients present with headache, which is mostly located in the posterior fossa – upper cervical region. Other symptoms, such as L'Hermitte's sign or unilateral or bilateral upper and lower extremity neurological dysfunction, such as decreased hand grip or spasticity or weakness of the lower limbs, could be the presenting symptoms. In a smaller percentage of patients, atypical pain of the upper or lower limbs is reported without discovery of cervical nerve root compression or extremity symptoms. **Over 50% of the patients present with weakness or numbness of one or more extremities. Unsteadiness of gait is seen in at least 40% of this patient's population.** However, other less frequent symptoms, such as diplopia, dysphagia, tinnitus, vomiting and dysarthria, could be the only presentation of this pathology or in combination with other symptoms as the initial presentation of this congenital abnormality. Less than 3% of the patients may present with dizziness, deafness, hiccups, facial hyperhyrdrosis and even facial numbness. About 10% of these patients will have a normal neurological examination. The patient may remain stable for many years with intermittent deterioration and spontaneous recovery.

The best diagnostic method is an MRI, which easily could detect the position of the cerebellar tonsils and their relationship with the proximal spinal cord, as well

as evaluation of the brainstem, hydrocephalus and syringomyelia.

The location of the cerebellar tonsils from the foramen magnum in normal individuals may range from 8 mm above the foramen magnum to 5 mm below with mean position of 1 mm above the foramen magnum. In Chiari type I malformation, cerebellar tonsils are, on average, 13 mm below the foramen magnum, which ranges from 3 to 30 mm below the foramen magnum. Syringomyelia may be present in 20-30% of the cases on the basis of the MRI scan study. Also, ventral brainstem compression with restriction of CSF flow at the craniocervical junction is the other MRI criteria for diagnosing Chiari type I malformation. In general, 5 mm cerebellar herniation below the foramen magnum is considered to be low enough to qualify. CT myelography could be an additional diagnostic tool, which is normally not necessary.

Incidental discovery of asymptomatic low cerebellar tonsils does not qualify the patient for any kind of surgical treatment. The patient who is treated within two years of the onset of the symptoms has a better outcome after surgical corrections are made. The most successful surgical outcome is achieved with posterior fossa decompression at the level of the foramen magnum and C1 and possibly C2 or C3 laminectomy, combined with partial cerebellar tonsillar reduction, usually unilaterally, supplemented with dural patch grafting. In over 60% of the cases, the cerebellar tonsils descend to C1 level and 25% to C2 and in only 3% this may extend to C3. In over 40% of the time, adhesions are noted between both the cerebellum and proximal cervical spinal cord and at the level of the outflow of the 4th ventricle. Cervical syringomyelia is seen about 30% of the time and, if not treated in a timely manner, may extend into the thoracic cord. The more extensive the syrinx formation, the less chance of satisfactory recovery. Drainage of the syrinx may be necessary if the posterior fossa decompression fails. This includes fenestration of the spinal cord at the level of the dorsal root entry zone with or without shunting. Rarely, 4th ventricular shunting or opening of the foramena magendie becomes necessary. Some neurosurgeons consider shrinkage of the cerebellar tonsils with bipolar cautery. In case of ventral brainstem compression, trans-oral odontoid resection has been performed in cases where the patient shows deterioration and progression of the basilar impression on serial MRI scanning, which is done only after posterior fossa decompression is attempted and fails.

The main benefit of surgery is to arrest the progression of this destructive lesion. The most favorable results occur when the patient has cerebellar symptoms, whereas the worse outcomes are noted when the patient has muscle atrophy, scoliosis, or symptoms lasting more than two years. Patients with pre-operative complaint of pain respond well to surgery. Usually, sensory symptoms, due to sphenothalamic involvement, respond much better than those with posterior column deficits.

Long term follow-up, mean of four years, reveals early improvement of pre-op symptoms over 80% and improvement of pre-op signs about 70%. Close to 15% show no change, compared to pre-op.

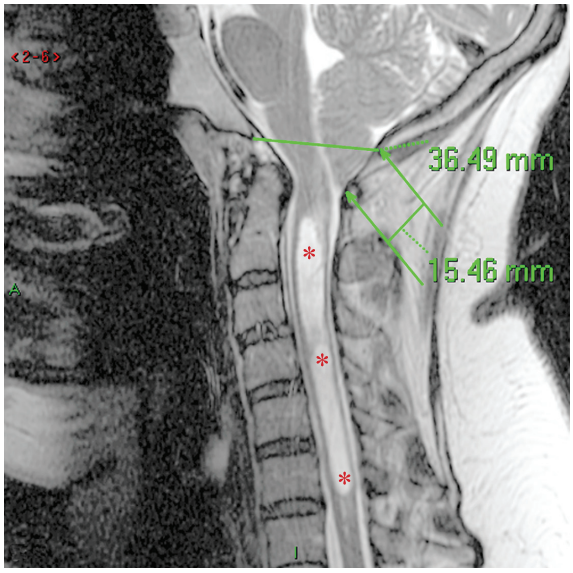

Fig. 1 – Pre-op MRI scan of the cervical spine discloses 15.4 mm caudal herniation of the cerebellar tonsils below the foramen magnum and very extensive cervical syrinx, seen by 3 red asterisks, showing the extent of the cystic degeneration of the spinal cord, extending from mid-C2 to upper C7 level.

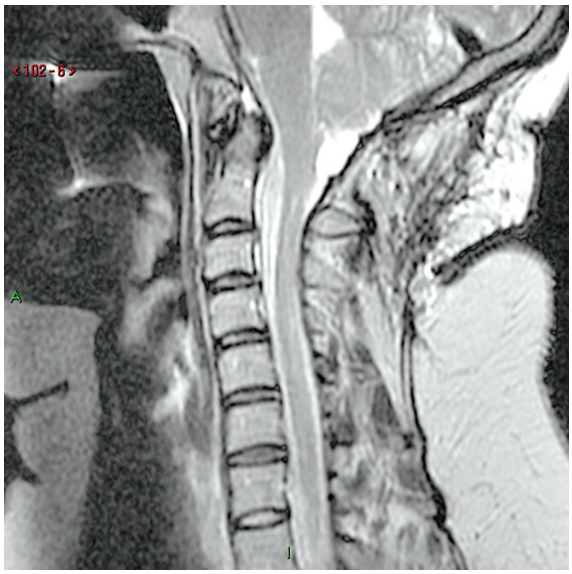

Fig. 2 – 2 ½ years post-op scan, shows dramatic regeneration of the cervical spinal cord and clearance of the syringomyelia and spacious foramen magnum, due to expansion of the subarachnoid space by bony decompression and duraplasty.