

Answer to Medical Quiz: Images

The first picture is of upper limbs of the patient showing- left hand and forearm is wider than the right. The second image is of MRI of cervical spine showing multiple T1 hypointense and T2 hyperintense areas in lower cervical and upper dorsal spinal cord, suggesting large syrinx in the spinal cord. The final image is of sagittal section of MRI of brain showing cerebellar tonsil herniation through foramen magnum, suggesting Chiari type 1 hind brain malformation.

Diagnosis:

Chiari type-I hind brain malformation with a syringomyelia of cervical and dorsal cord

Review: Arnold–Chiari Malformation & syringomyelia

Arnold Chiari malformation was first described by Austrian pathologist Hans Chiari.

It is the downward displacement of the cerebellar tonsils through foramen magnum. It is of four types. In type 1 syringomyelia of cervical/cervicothoracic spinal cord can be seen as well as medullary kink/ brainstem elongation. Patient is usually asymptomatic during

childhood. Later on develop head ache, neck pain, unsteady gait. In type 2 both cerebellum and brain stem extend into foramen magnum, causing paralysis below the spinal defect. Type 3 is associated with an occipital encephalocele and hydrocephalus. In type 4 defect there is lack of cerebellar development and cerebellum & brain stem lie in posterior fossa with no relation to foramen magnum. This type is not compatible with life.

Syringomyelia is fluid filled cyst in spinal cord surrounded by glial tissue, mostly at the level of C8 and T1 segment. Expanding cavity may disrupt anterior horn cells of spinal cord, lateral spinothalamic tract. It may involve the brainstem called syringobulbia. In 25 % cases it is associated with of Chiari malformation. Patients present with the triad of –Dissociated sensory loss in neck, shoulder, arms, LMN signs in upper limb and UMN signs in lower limbs.

The aims of treatment are to relieve pressure on brain and spinal cord and re-establish normal fluid circulation through and around affected area. Treatment options include posterior fossa decompression, electrocautery and spinal laminectomy.