A 35 year old male presented with headache and difficulty in walking of three months duration. Clinical examination revealed ataxic broad based gait, nystagmus in both the eyes, hypotonia of all the limbs and decreased tendon reflexes. An MRI of the brain was performed (Fig. 1). What is the diagnosis?
Solution to Radiology Quiz

Chiari 1 Malformation.

Figure 1 is a T1 weighted mid sagittal MRI of the brain showing enlarged cerebellar tonsils which extended below the foramen magnum for a distance of 8 mm up to the C2-3 level. The posterior fossa is normal and the rest of the brain and upper cervical cord is unremarkable.

Dr Hans Chiari in 1891 first described three hindbrain disorders associated with hydrocephalus. These disorders have neither an anatomical nor embryological correlation with each other, but they all involve the cerebellum and spinal cord and are thought to belong to the group of abnormalities that result from failure of normal dorsal induction (1). Five years later, in a further study on hindbrain deformities (2) he revised the second type of malformation as well as offered a new fourth type of malformation.

Chiari I malformation involved caudal displacement of the cerebellar tonsils, and sometimes the inferior vermis, through the foramen magnum into the rostral cervical spinal canal. Associated syringo-hydromyelia may be present. Chiari II malformation, also known as the Arnold-Chiari and Cleland-Chiari malformation, involves displacement of the brainstem and lower cerebellum into the cervical spinal canal. In such cases, the fourth ventricle is also caudally displaced and extends below the foramen magnum. Chiari II abnormalities are almost always associated with lumbar myelomeningoceles. Chiari III malformation involves downward displacement of the medulla with herniation of the cerebellum initially through the foramen magnum, then dorsally through a cervical spina bifida, resulting in a cervical encephalocele. Chiari IV malformation has severe cerebellar hypoplasia with displacement of brain.

Clinical presentation of Chiari malformations are varied and depend on the degree of tonsillar ectopia. Up to 30% of patients with Chiari I malformation are asymptomatic (3). Symptomatic patients present with cerebellar, long-tract signs and other symptoms which mimic demyelinating disease. They may also present with cranial neuropathies as the result of brainstem compression or with pain and dissociated anesthesia of the upper extremities resulting from associated syrinx-hydromyelia. Concurrent Chiari I malformations and syringo-hydromyelia have been estimated to occur in up to 25% of patients who are asymptomatic and 60-90% of those who are symptomatic (4). Imaging plays an important role in diagnosing and assessing the degree of tonsillar ectopia. MRI with T1 and T2 weighted sagittal and coronal scans is the modality of choice in evaluating Chiari malformations. It accurately evaluates tonsillar configuration and position as well as associated syringo-hydromyelia. Keeping the CSF dynamics in view, present day studies on patients with Chiari I malformations focus less on the static anatomic appearance of the tonsils in relation to the foramen magnum, and more on the dynamic motion (CSF). In the near future, MR imaging coupled with MR fluoroscopy of study CSF dynamics, will permit a comprehensive high resolution imaging of the craniocervical junction in asymptomatic patients with Chiari I malformations with the aim to identify which patient is likely to develop symptoms over a period of time so that corrective actions can be initiated before irreversible neurological damage occurs.

Asymptomatic Chiari malformations are difficult to detect on routine screening radiological investigations which are presently carried out for selection of aircrew. However if MRI of the spine is included in the initial medical examination for selection of aircrew this abnormality will be easily detected which may prevent loss of trained manpower in the future.

Treatment of Chiari I malformation is surgical and the options available seem to evolve and recycle with time. Among the choices that the neurosurgeon can offer and decompression of the foramen magnum with or
without duraplasty, Obex plug and a variety of shunts like ventriculoperitoneal and syringo-perritoneal. Best surgical results are seen with decompression of the foramen magnum along with duraplasty.

References: