Adult chiari malformation presenting as bilateral vocal cord paralysis

S. Chawla · D. T. H. Wee · V. Arora

Abstract In the Chiari malformations, herniation of the cerebellar tonsils through the foramen magnum into the cervical spinal canal can often result in headaches and spinal cord deficits. Bilateral vocal cord paralysis has been reported as a result of Chiari malformation but is rare and the vast majority of reports have been in the paediatric population. We report a case in a 68-year-old man with adult Chiari malformation where aspiration pneumonia from bilateral vocal cord palsy was the only manifestation of this malformation.

Keywords Adult Chiari malformation · Arnold Chiari malformation · Vocal cord palsy

Introduction

The Chiari malformations are a rare group of congenital abnormalities involving the hindbrain and spinal cord. Herniation of the cerebellar tonsils through the foramen magnum into the cervical spinal canal places the spinal cord under pressure. Anaesthesia and weakness are present in about 50% of cases. This is typically in a “cape like” distribution involving the upper limbs and thorax. Pain in the upper limbs and neck (up to 70%) however is the most common complaint. Occipital headaches are estimated to occur in up to 35% of patients [1]. Other symptoms include ataxia, trigeminal neuralgia and vocal cord palsy [1, 2].

We report a case of adult Chiari malformation in which bilateral cord palsy was the only isolated presenting complaint.

Case report

A 68-year-old man was admitted to Hospital under the care of the physicians after having a series of conscious collapses. One-week history of flu like symptoms and shortness of breath was obtained. Patient also reported experiencing difficulty swallowing for the previous 4 weeks. Although softly spoken, the patient denied history of any change in voice. Physical examination findings and chest X-Ray was consistent with aspiration pneumonia. Further history revealed that the patient had never suffered any episodes of significant headaches, neck pain, altered sensation or any motor weakness in the past.

The Otolaryngology team was consulted and a functional endoscopic assessment of swallow (FEES) was performed in conjunction with a speech pathologist. The patient had a weak voice. The gag reflex was absent and there was widespread pooling of saliva in the pyriform sinuses. Bilateral vocal cords palsies in a paramedian position were present. There was absence of abduction of the vocal cords with evident aspiration.

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Further imaging in the form of magnetic resonance imaging (MRI) of the brain and computed tomography (CT) of the neck were obtained. The MRI revealed a Type 1 Chiari malformation (Fig. 1). Radiological diagnosis of a Chiari malformation requires the cerebellar tonsils to be lying at least 5mm below the foramen magnum. CT of the cervical spine demonstrated a congenital fusion of C1, occipital condyles and posterior arches with occiput. There was mild cerebellar herniation into the posterior spinal canal.

Neurosurgical team was consulted and the patient went on to have a posterior fossa decompression, C1 laminectomy and fascia lata duraplasty for repair. A tracheostomy was done concurrently to help patient protect his airway. Patient made a good post-operative recovery but has tracheostomy in situ 4 months post procedure. A percutaneous endoscopic gastrostomy (PEG) was inserted to assist with long-term nutritional support.

Discussion

In 1891, the Austrian pathologist Hans Chiari described Chiari malformation as a group of congenital abnormalities involving the cerebellum and spinal cord. [1] The Type 1 Chiari malformation or adult Chiari malformation has its primary feature herniation of the cerebellar tonsils through the foramen magnum into the cervical spinal canal with obliteration of the cerebellomedullary cistern but no meningocele associated [3]. The Type 2 Chiari malformation has a meningocele associated with it and typically presents early in the paediatric population. Julius Arnold described a similar case of a Type 2 Chiari malformation in 1894 [3].

The term Arnold Chiari malformation was later coined to incorporate those Chiari malformations associated with

a meningocele [3]. This has to some extent, created confusion in the literature. Strictly speaking this term should only be used in patients with a Type 2 Chiari malformation and not to be confused with adult Chiari malformation or Type 1 Chiari malformation.

Chiari Malformation is described in the literature as a potential cause of vocal cord paralysis. However, this is more commonly seen in the Type 2 Chiari malformation typically occurring in the newborn. As these cases have a meningocele associated with it, the symptoms usually present early and hydrocephalus is common. The patient may also be at risk of coning with respiratory compromise, obstructive sleep apnoea and even death [4].

Adult Chiari or Type 1 Chiari malformation has no associated meningocele. There is caudal displacement of cerebellar tonsils through the foramen magnum but the symptoms are often insidious. Patients are often asymptomatic till adulthood.

In this case report, we present a man who is 68 years of age who has never demonstrated any symptoms and signs of syringomyelia or headaches. The only symptom was aspiration pneumonia from bilateral vocal cord palsies. Our literature search found only 2 other such reported cases in adults where vocal cord palsy and aspiration pneumonia were the initial presenting complaint for Type 1 Chiari Malformation. However the patients reported were much younger, being 44 and 54 years old respectively [5, 6].

Chiari malformation is a rare cause of bilateral cord palsy in the adult population. In any elderly patient presenting with aspiration and vocal fold palsy, a cerebrovascular event, space-occupying lesion or malignancy is high on the list of the differential diagnosis. The experience of this case highlights the importance of MRI imaging and the need to consider other rare causes, as the management and outcome were significantly altered.

Fig. 1 Sagittal MRI showing type 1 Chiari Malformation.

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