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Unusual association of diseases/symptoms

Arnold–Chiari malformation type 1 complicated by sudden onset anterior spinal artery thrombosis, tetraparesis and respiratory arrest

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Summary

Chiari in 1891 described a constellation of anomalies at the base of the brain inherited congenitally, the characteristic of which are (1) extension of a tongue of cerebellar tissue posterior to the medulla and cord that extends into the cervical spinal canal; (2) caudal displacement of the medulla and the inferior part of the fourth ventricle into the cervical canal; and (3) a frequent but not invariable association with syringomyelia or a spinal developmental abnormality. Chiari recognized four types of abnormalities. Presently, the term has come to be restricted to Chiari's types I and II, that is, to cerebellomedullary descent without and with a meningomyelocele, respectively. The association of Arnold–Chairi malformation and high cervical cord infarction is unusual. The most common syndrome, anterior spinal artery syndrome (ASAS), is caused by interruption of blood flow to the anterior spinal artery, producing ischaemia in the anterior two-thirds of the cord, with resulting neurologic deficits. Causes of ASAS include aortic disease, postsurgical, sepsis, hypotension and thromboembolic disorders. The authors present an interesting case of cervical cord infarction due to anterior spinal artery thrombosis in a patient of type 1 Arnold–Chiari malformation without any of the above predisposing factors.

BACKGROUND

Type 1 Arnold–Chiari malformation presenting as sudden onset tetraparesis with anterior spinal artery thrombosis is reported for the first time.

CASE PRESENTATION

A 31-year-old Saudi female not known to have any medical illness in the past presented to a peripheral hospital with sudden onset neck pain that radiated to both arms. Within less than half an hour, the patient collapsed and was apneic. She was intubated and electrocardiography performed was normal. Blood pressure was 90/70 mm Hg and pulse rate 76/min. Patient was shifted to our hospital for further workup and management. Examination in the emergency room revealed patient intubated with Glasgow coma scale of 3/15. Limbs were flaccid and plantar response was unequivocal. Repeat electrocardiography showed sinus rhythm. X-ray of the chest was normal as were complete blood counts and biochemistry. Cardiac markers were negative. CT brain revealed diffuse brain oedema. Cerebrospinal fluid examination revealed normal protein of 23.82 mg/dl (range up to 45 mg/dl) and normal cell count. Patient was admitted to the intensive care unit and MRI with contrast revealed an elongated intramedullary T2-T1 high-signal intensity lesion from C2 to C5 spinal level with swollen cord suggestive of ischaemic infarction (figures 1 and 2). There was cerebellar tonsillar herniation through the foramen magnum behind the upper cervical cord. Anterior spinal artery thrombosis was diagnosed on MRA (figure 3). Cervical spine was normal. Blood glucose was normal as was lipid profile. Immune markers were negative and anticardiolipin antibody was negative. Patient was diagnosed as Arnold–Chairi malformation type 1 with anterior spinal cord syndrome due to anterior spinal artery thrombosis resulting in sudden respiratory arrest and anoxic brain damage due to spinal shock and dysautonaumia. Patient developed ventilator-associated pneumonia, followed by multiorgan failure and is still living in a vegetative state.

INVESTIGATIONS

MRI with contrast.

DIFFERENTIAL DIAGNOSIS

- High cord compression
- Intramedullary cervical cord tumour
- Haematomyelia
- Atlantoaxial dislocation
- Venous thromboembolism.

OUTCOME AND FOLLOW-UP

Developed anoxic brain damage and is living in vegetative state on tracheostomy.

DISCUSSION

Arnold–Chiari malformation is a heterogenous entity and no embryological or genetic theory has explained the exact

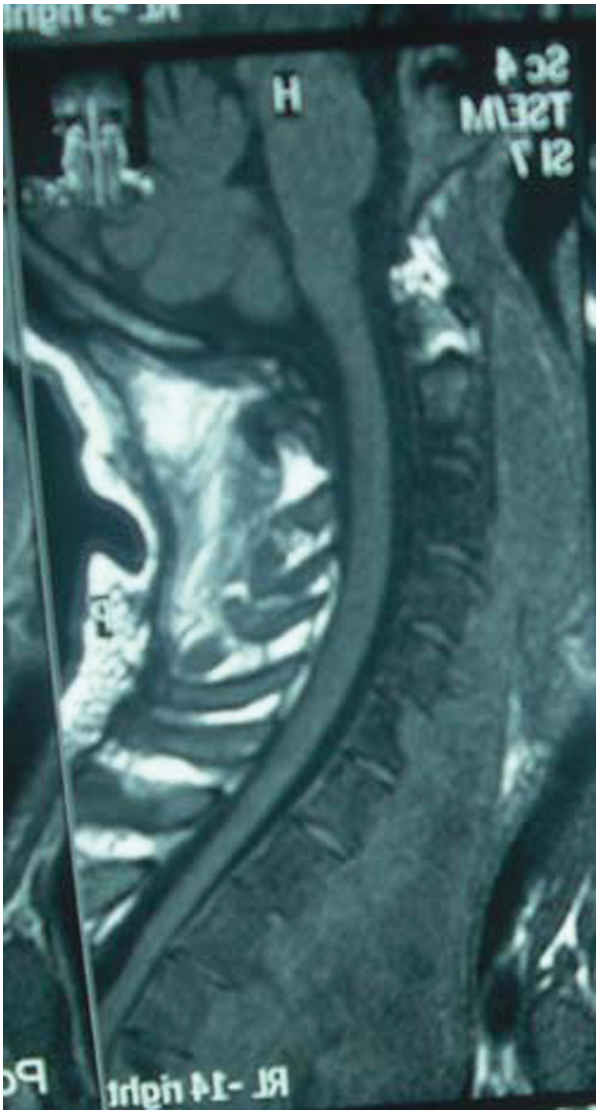

Figure 1 MRI showing tonsillar herniation with normal looking cervical spinal cord.

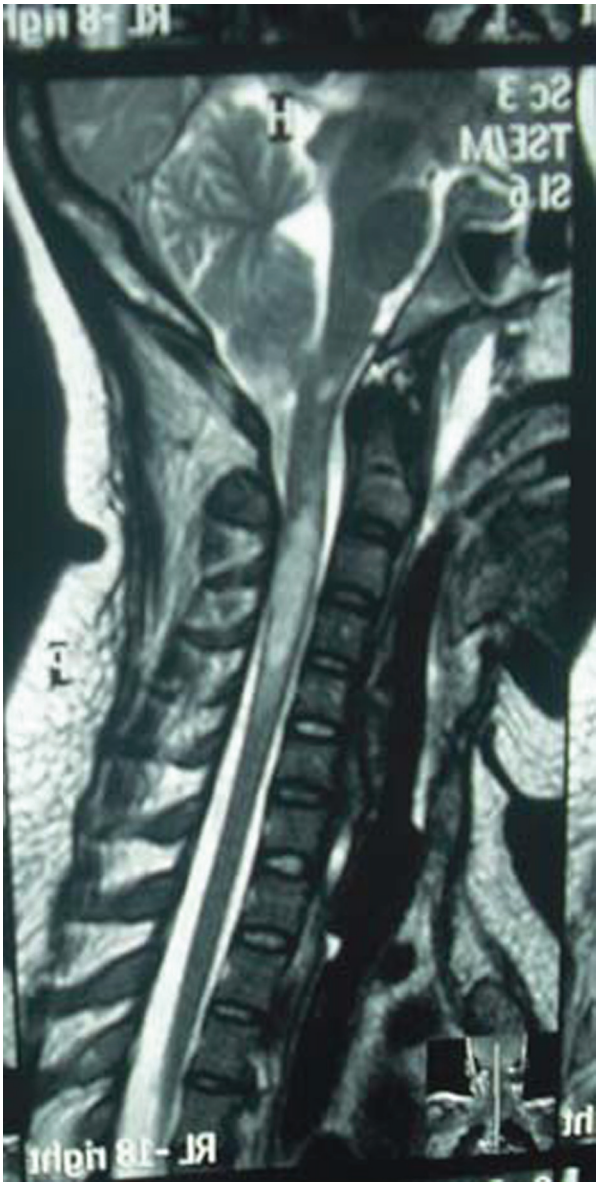

Figure 2 MRI showing cervical cord infarction on contrast enhancement.

pathogenesis or genetic mutation of all forms of malformation.[1] At present, the two main types are type I and type II per revised classification.[2]

In the more common type I Chiari malformation (without meningocele or other signs of spinal dysraphism), neurologic symptoms may not develop until adolescence or adult life. The symptoms are those of (1) increased intracranial pressure, mainly headache, (2) progressive cerebellar ataxia, (3) progressive spastic quadriparesis, (4) downbeating nystagmus or (5) the syndrome of cervical syringomyelia (segmental amyotrophy and sensory loss in the hands and arms, with or without pain). The patient may show a combination of disorders of the lower cranial nerves, cerebellum, medulla and spinal cord (sensory and motor tract disorders), usually in conjunction with headache that is mainly occipital. This combination of symptoms is easily mistaken for multiple sclerosis or a tumour at the foramen magnum. The MRI is a useful tool to differentiate these conditions.[2] The symptoms are usually chronic but may have an acute onset after sustained or forceful extension of the neck, as, for example, after a long session of dental work, hairdressing in women or chiropractic manipulation. The physical habitus of such patients may be normal, but approximately 25% have signs of an arrested hydrocephalus, or a short 'bull neck'. The nature and severity of headache, reasonably attributable to a Chiari malformation, are somewhat unclear. Occipitonuchal pain with coughing, position change or the Valsalva manoeuvre is the most dependable association.[3] Sudden onset nuchal pain and respiratory arrest in our case was due to cervical cord infarction and spinal cord swelling due to anterior spinal artery thrombosis. There have been case reports of breathing disorders following anterior spinal artery thrombosis.[4] The reported causes of anterior spinal artery thrombosis are genetic coagulation disorders,[5 6] postsurgical, thromboembolism, sepsis and systemic lupus erythematosus.[7] There is a case report of aspergillosis and anterior spinal artery thrombosis.[8] That Arnold–Chiari malformation type I is a cause or association of anterior spinal artery thrombosis in

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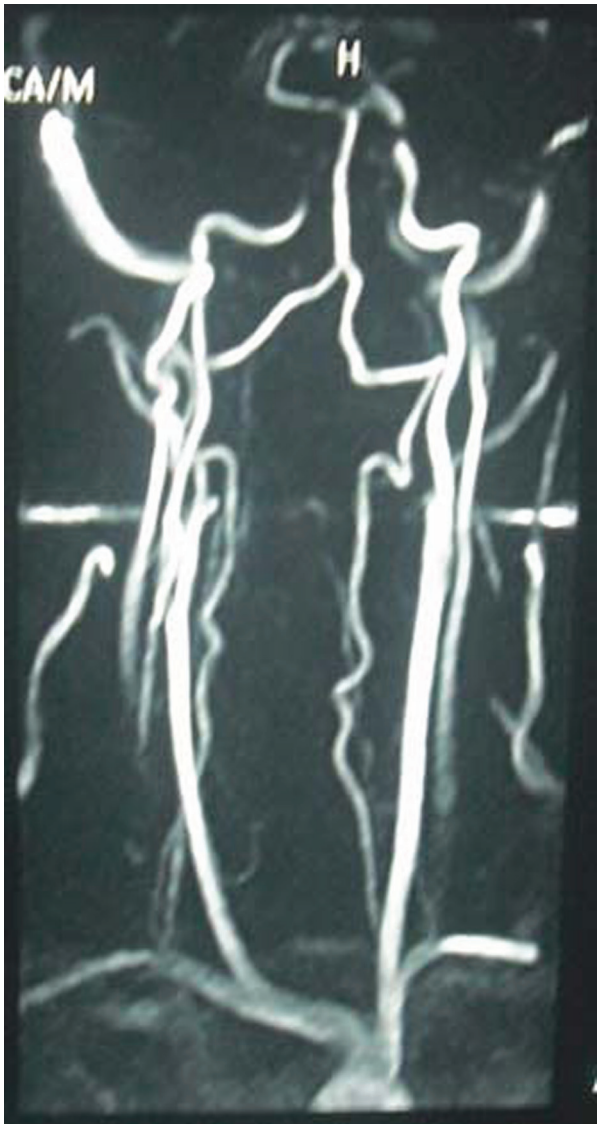

Figure 3 Magnetic resonance arteriogram showing absence of anterior spinal artery signal.

the cervical cord is not reported so far, nor is catastrophic presentation as tetraparesis and respiratory arrest known. Thus, our case emphasises the awareness of these presentations and associations in type I Arnold–Chiari malformation in adult life.

Learning points

- Catastrophic presentation of type I Arnold–Chiari malformation as respiratory arrest and tetraparesis is not unusual and awareness of the condition can avoid anoxic brain damage by early intervention.
- Association of cervical anterior spinal artery thrombosis and Arnold–Chiari malformation type I is reported; whether the latter is causal is not known.
- MRI is a useful tool for diagnosis of the anomaly as well as anterior spinal artery thrombosis and spinal cord infarction especially with contrast enhancement.
- Early recognition of spinal shock would have ameliorated the anoxic insult to brain by support with vasoconstrictors rather than intravenous fluids.

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Competing interests None.

Patient consent Obtained.

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