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Case Report

Apnoeic episodes in a patient with Chiari type I malformation

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The Chiari type I malformation is a rare condition that generally occurs in young adults. It consists of a caudal displacement of the cerebellum and brain stem to a variable extent through the foramen magnum.[1]

There can be a wide variety of symptoms in these patients, including headache, weakness, numbness of the limbs, unsteadiness and loss of balance.[2] A variety of respiratory disorders have been described including acute respiratory failure,[2] respiratory arrest [3] and sleep apnoea.[1] It is extremely unusual for a patient with the condition to have no abnormal neurological signs on presentation, if apnoeic problems are a feature.

We describe such a patient in whom, curiously, a transient pyrexia accompanied his apnoeic episodes.

CASE REPORT A 31 year old farmer arrived in Casualty at Mid-Ulster Hospital with a history of several apnoeic episodes associated with unresponsiveness, witnessed by his wife. Rectal diazepam had been administered about an hour prior to admission by the general practitioner, as the patient had appeared to be anxious and shivering on coming round from the apnoeic episodes.

There was a past medical history of similar symptoms, also with a high initial temperature, 18 months and two years previously, requiring hospital admission for two days on each occasion. The diagnosis on each admission was "viral illness". Hyperventilation had been noted on a couple of occasions. Arterial blood gases during one of these episodes showed a pH of 7.64, (normal range 7.36-7.44kPa) pO2 13.7, (normal range 11.3-14.6 kPa) and pC02 2.42,(normal range 4.7-6.0 kPa).

On examination on his third admission, his temperature was 38.6 degrees centigrade. He had 10 apnoeic episodes in Casualty associated with loss of consciousness, during which he was noted to be haemodynamically stable. Full neurological examination revealed no significant abnormalities, apart from his unresponsiveness whilst apnoeic.

Investigations showed that full blood picture, urea and electrolytes, thyroid function and blood sugar were normal. ECG and chest x-ray were

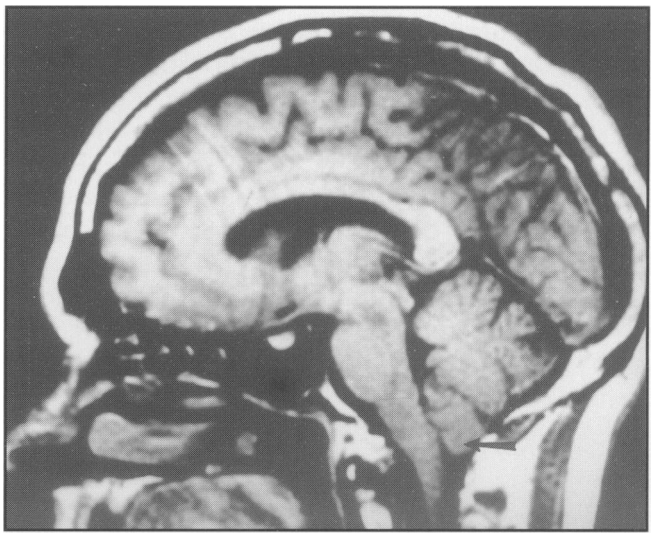

Figure MRI image showing ceribillar tonsilar discent (Chiari type I malformation).

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normal. CT scan brain and EEG with hyperventilation were normal.

He was readmitted one week later with further apnoeic episodes, and two weeks later with severe generalized headache. Full neurological examination was invariably normal.

MRI scan was performed in order to evaluate the brain stem more accurately than CT scanning permits. This showed a Chiari type I malformation with herniation of the cerebellar tonsils through the foramen magnum to the level of the upper border of the posterior arch of C1. (Figure) MRI of the thoracic spine was normal.

He was transferred to the Regional Neurosciences Unit for assessment with a view to foramen magnum decompression. It was decided to proceed with surgery. A sub-occipital craniectomy was performed and the posterior arch of C1 was removed. The dura was then opened and the tonsils were found to have herniated to just below the level of the foramen magnum, but they did not reach the arch of C1. Some fine adhesions between the tonsils and the brainstem were noted; these were divided by sharp dissection. The fourth ventricle was entered and a Surgicel patch was placed over the dural opening. Post-operative recovery was unremarkable apart from some headaches, which gradually resolved. There have been no further problems with apnoeic episodes in the 30 months that have elapsed since surgery.

DISCUSSION

Although the original description of the Chiari malformations was in 1891,[4] respiratory failure as a presenting feature in adults has only been described in the past decade.[2, 3, 5, 6]

Conventionally, the diagnosis should be considered in adults when symptoms and signs of damage to the cerebellum, medulla and lower cranial nerves appear.

Our case illustrates that it is important to think of the diagnosis in any patient with unexplained apnoeic episodes, even though neurological examination may be normal.

The exact pathogenesis of the apnoea is not fully understood – possibly compression of the medulla or compromise of the vascular supply to the brain stem may be responsible.[7]

Chiari type 1 malformation is a condition that is potentially treatable by surgical decompression.[3] The fact that our patient has not relapsed during the 30 month follow-up period is encouraging.

The unusual features in this case are the absence of neurological signs or florid evidence of respiratory failure, and the presence of pyrexia with the apnoeic episodes.

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