

About the case of a Chiari 1 malformation discovered after several syncope on a sport ground

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Case report

In February 2011, a 22 year-old man came to see us to recover his ability to practice rugby as an amateur 3rd line. The patient's medical history present a cardio circulatory arrest that took place during a minor surgical intervention, after his anesthetic induction in dorsal decubitus, as he was manipulated to be placed in ventral decubitus.

He experienced transitory dizziness when he was lying down or during fast cephalic position changes. During a game, he suffered a cranial trauma, he had fainted for 2 minutes with 20 minutes amnesia. He also reported having briefly passed out in a match during a tackling that caused a scapular impact without head trauma.

The clinical examination, notably the neurological one, was normal one week after the last episode. We faced difficulties to advance in the diagnosis due to these atypical neurological events. It was decided to make a morphological imaging. His doctor had asked him to stop his sport activity and had requested a brain MRI.

Imaging

A brain MRI discovered a Chiari 1 malformation :

- A disappearance of the peri- bulbar liquid spaces related to the filling of the posterior part of the occipital hole by the cerebellar tonsils was noticed in the foramen magnum shot (there is an hypoplasia of the posterior fossa).
- The cerebellar tonsils are abnormally in low position, the protrusion was 28 mm long under the line joining the two most inclined points of the occipital bone (Figure).
- The large cistern was filled in the T1 sagittal sequences compared to normal subjects whose images show a free aspect of the the large cistern and cerebrospinal fluid around the brain stem
- A medullary MRI showed no syringomyelia .

Figure : Cerebral MRI T1 Sagittal



Chiari 1 malformation

Discovered in 1891 by Hans Chiari, comprises 3 types : Type 1 can long remain pauci-symptomatic, more frequent among women (53/2), its prevalence can be getting close to 5,6‰.

Due to an insufficient development of the posterior fossa, inducing a protrusion of the cerebellar tonsils and of the brain stem through the occipital hole in the cervical canal. In type 1 the protrusion is limited to the lower brain stem and cerebellar tonsils that can reach C2 (25%) or C3. This hernia reduces the required medullary space and disturbs the circulation of the cerebrospinal liquid. It favours the formation of syringomyelic cavities (including very distal ones 32-74%) and hydrocéphalus.

The symptoms appear on average around the age of 25 :

- Headaches 50% significant when they are posterior (caused by coughs, Valsalva maneuver, laughter or efforts).
- Dizziness feelings (caused by quick cephalic position changes), downbeat nystagmus (25%), dysphonia or a dysphagia (5-15%), tinnituses, ataxia, dysesthesia of the trunk or extremities.
- Other more serious manifestations : apnea sleep disorder, syncope, cardiopulmonary arrests for minor traumas, transitory tetraparesias.

Discussion

Suspicion of the
malformation



Brain and medullar
MRI



Neurosurgic
al advice

Contraindica
tion to

- Our patient showed significant symptoms (positional dizziness) and unusual history (cardio circulatory arrest, fainting fit after a scapular contact).
- The normal position of the cerebellar tonsils is above the occipital hole (but not pathological if the protrusion remains ≤3mm for an adult and 5 mm for a child).

- Identify this malformation as soon as possible to avoid serious complications. The examination led for participation in sports activities must try to look into the significant signs, especially in case of consultation for the ability to practice contact sports.
- Cessation of the sport practice must be imposed until the results of the brain and medullar MRI are revealed (reference imaging).

- Rugby is very practiced in our region, which has evolved over the last 10 years of practice avoidance to that of a deliberate collision of the opponent. Technicians have therefore adapted rugby fitness training and the rules also reduce the risk of a serious accident and the ability of front row players has been the subject of specific regulations.

sports

Conclusion

The discovery of a Chiari 1 malformation on a pauci - or asymptomatic subject requires a neurosurgical advice. This malformation constitutes an absolute and permanent contraindication to a contact sports practice, or to sports which might cause falls or collisions.

Acknowledgement

Written consent to publish was obtained. JL, MB and CH wrote the report. We thank French Society of Sports Traumatology for their support.