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Case Report of a Cardiac Arrest during Surgery Revealing an Arnold Chiari Malformation

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Abstract **Case Report**

Arnold Chiari malformation is a common congenital anomaly of the craniocervical junction. It is defined as a descent of the cerebellar tonsils below the foramen magnum, putting pressure on both the brain and spine causing many symptoms. The patients typically experience no symptoms or chronic slowly progressive symptoms. However, recent reports indicate that a subset of patients with AC may present with acute deterioration, cardiorespiratory arrest and sudden unexpected death. We report a case of sudden unexpected death of a 34 years old woman, married with 3 kids, without any medical history, that was admitted for a heterogeneous multinodular goiter in clinical and biological euthyroidism and scheduled for total thyroidectomy. In intraoperative, the patient presented a cardiorespiratory arrest while positioning her head in hyperextension for a good surgical approach, neck released by anesthesia. Para-clinical tests showed later an AC malformation. Cases of sudden cardiorespiratory arrest in children and adults with AC malformation have been documented, leading to death in some cases. However, these patients who died unexpectedly have experienced a recent trauma. This is a unique example of a sudden unexpected death of a woman undiagnosed AC, in absence of any previous trauma, that occurred during a conventional anesthesia for a thyroid surgery.

Keywords: Cardiac arrest - Arnold Chiari- sudden death - hyperextension of the neck.

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INTRODUCTION

Arnold-Chiari I malformation (ACMI) is a congenital disorder corresponding of the caudal misplacement of the cerebellar tonsils into the upper cervical spinal canal [1].

ACMI may be asymptomatic until adolescence or adulthood, and even when they are symptomatic, symptoms are variable and non-specific. Common symptoms include but are not limited to headache, neck pain, hearing loss, extremity weakness, dizziness, sleep apnea, cardio respiratory arrest, and rarely sudden death.

We report a case of sudden unexpected death of a 34 years old woman, scheduled for total thyroidectomy. In intraoperative, the patient presented a cardio respiratory arrest while positioning her head in hyperextension.

CASE REPORT

A 34-year-old woman, without any medical history, that was admitted for a heterogeneous multinodular goiter in clinical and biological euthyroidism and scheduled for total thyroidectomy. Pre anesthetic consultation concluded on an ASA II patient, stable.

The patient was admitted at the OR after premedication made by Hydroxyzine (1mg/kg), vital and hemodynamic constants at the admission in the OR were: BP 120/75 mmHg, PR 85 bpm and SpO2 100%.

Induction was done, after pre oxygenation, by Fentanyl (3µg/kg) Propofol (2mg/kg) and Rocuronium (0,6mg/kg). Patient was intubated without any incident, entertain of anesthesia was done by Isoflurane (MAC at 1) and bolus of Fentanyl.

The patient was positioned after by a hyperextension of the neck to expose the surgical site, 1 minute after, the patient presented a bradycardia at 25bpm managed by atropine 0,5 mg, the heart rate stabilized at 90bpm, 2 minutes after, a second bradycardia occurred refractory to atropine followed by asystole. The CPR has been performed with 100% of Oxygen, verification of the placement of the tracheal tube and isoflurane was stopped. A normal cardiac activity resumes after 5 minutes of CPR and 2 mg of adrenaline, the surgery was postponed till stabilization of the patient and the patient was transferred to the ICU.

At the ICU, patient was sedated and ventilated (6ml/kg) in CV mode; the BP was 120/55mmHg PR at 89 SpO2 95%, miosis; the Glasgow Coma Score was 3 with absence of the brainstem reflex, EKG and biology samples were normal and also a CT scan was normal.

24 hours after, sedation was stopped without any sign of awakeness, GCS at 3, persistent miosis and absence of the brainstem reflex, EKG shows no conduction or repolarization trouble and electroencephalography performed at day 2 shows diffuse brain distress.

An MRI result was ACMI type 1 (Figure 1 and 2). The evolution was marked by the persistence of the coma with neurovegetative events and cerebral oedema and cerebral death at day 6.

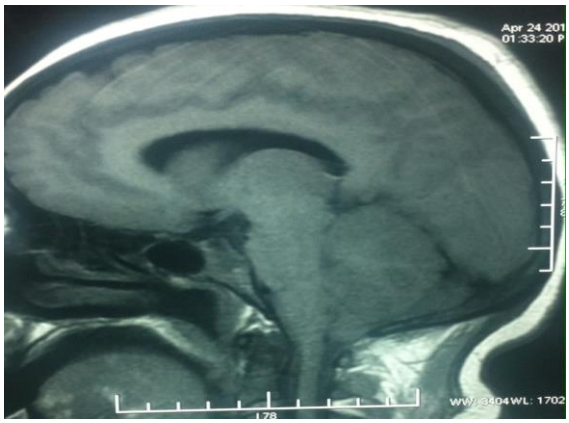

Figure 1: Ptosis of the cerebellar tonsils shown in a cereberal MRI

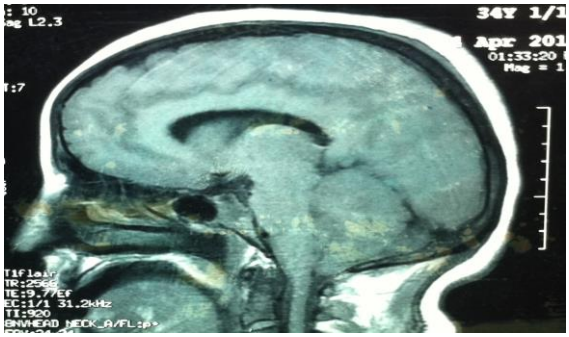

Figure 2: Ptosis of the cerebellar tonsils shown in a cereberal MRI

DISCUSSION

ACMI is defined as hydrocephalus with cerebellar tonsillar herniation of at least 3–5 mm through the foramen magnum into the cervical canal [2]. CMI has been divided into adult and pediatric types, and the adult type usually presents after the second or third decade of life, forming a chronic tonsillar herniation [3].

Type I is the most common form of ACM and has been estimated to occur in 1 in 1000 births and mostly asymptomatic. ACMs cases are often detected coincidently while undergoing unrelated investigations [4, 5]. ACM type I is no longer listed as a rare disorder, the introduction of MRI has increased the frequency of ACM case discoveries.

The prevalence rate of type I ACM is ranged between 0.1%-0.5% and less predominant in males [6]. Genetic basis for type I ACM has been often suggested [7, 8]. Recent studies favor the presence of genetic linkage to chromosomes 9 and 15.

It can also be caused later in life if spinal fluid is drained excessively from the lumbar or thoracic areas of the spine either due to traumatic injury, disease, or infection. This is called acquired or secondary Chiari malformation. Primary Chiari malformation is much more common than secondary Chiari malformation [9].

The herniated cerebellar tonsils restrict cerebrospinal fluid exiting from the fourth ventricle causing increased pressure upon the upper cervical spinal canal resulting in a wide variety of associated symptoms. Common symptoms include but are not limited to headache, neck pain, hearing loss, extremity weakness, dizziness, sleep apnea, cardiorespiratory arrest, and rarely sudden death [10].

Our case describes a cardiac arrest occurring after positioning the head of the patent to expose the surgical area, Alegre and al was the first to describe a sudden cardiac arrest and death in an adult patient with ACMI like our case the death was preceded by a head movement causing a transient compression of the brainstem precipitating cardiac arrest, we believe that we are reporting the first case of Chiari I malformation causing a cardiac arrest in the Operating room while positioning a patient for non-neurosurgical surgery, and we also notice the importance of the collaboration between surgeons and anesthetists while positioning a patient before surgery.

CONCLUSION

ACMI is no more a rare disease, symptoms are uncommon but remains dangerous, we should consider it in case of unexplained symptoms (neurological, cardiovascular or respiratory….), the MRI makes the diagnosis easy and the only treatment is surgical.

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Conflict of interest: None

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