Case Report

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Arnold Chiari Malformation Mimicking Postdural Puncture Headache in a Postpartum Female

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ABSTRACT

Arnold Chiari malformations are a group of structural defects ranging from herniation of the hindbrain to skeletal deformities and neurological dysfunction. Type I Arnold Chiari is the most common type of malformation with a prevalence of 1 in 1000 births. Patients are frequently asymptomatic; however, if symptoms do develop, it is typically during the reproductive age. Suboccipital headaches and neck pain are the most common symptoms that one with type 1 Arnold Chiari will experience. Here we present a case of a 29 year old, Afican American female, G4P1021, who presented to our labor and delivery unit at forty weeks and two days of gestation complaining of regular painful uterine contractions. She delivered a healthy neonate vaginally with vacuum assistance and developed an occipital headache associated with neck pain on postpartum day 1. Her symptoms did not resolve after 2 blood patches at which point she was evaluated by neurology. Head imaging was performed and notable for an Arnold Chiari malformation.

Keywords:

Menstrual period, Abortion, Pregnancy, Postdural puncture.

Introduction

Arnold Chiari malformations are a group of structural defects ranging from herniation of the hindbrain to skeletal deformities and neurological dysfunction. There are four types of malformations: type I occurs when there is a displacement of the cerebellar tonsils beneath the foramen magnum, usually diagnosed in adolescents or adults. Type II occurs when there is a downward displacement of the medulla, fourth ventricle, and cerebellum into the cervical spinal canal, as well as elongation of the pons and fourth ventricle. Type III occurs when a portion of the cerebellum and/or brainstem herniates through a defect in the back of the head or neck, and type IV occurs when there is a hypoplasia of the cerebellum.

Type I Arnold Chiari is the most common type of malformation with a prevalence of 1 in 1000 births [1]. Patients are frequently asymptomatic; however, if symptoms do develop, it is typically during the reproductive age. Suboccipital headaches and neck pain are the most common symptoms that one with type 1 Arnold Chiari will experience. Other symptoms that an individual with type 1 Arnold Chiari may experience are dizziness, tinnitus, vertigo, double or blurred vision, hypersensitivity to bright lights, gait ataxia, and muscle weakness, just to name a few [2]. These symptoms are typically made worse with maneuvers that increase intraabdominal pressure, namely the Valsalva maneuver.

There have been few case reports documented in the literature of women experiencing new-onset symptoms of Arnold Chiari after childbirth [3-7] and there is much debate regarding possible iatrogenic causes of this malformation in the postpartum period [8]. Here, we present a case report of a young female who developed persistent headache and neck pain on postpartum day one status post a vacuum-assisted vaginal delivery.

Case Presentation

A 29-year-old African American female, G4P1021, presented to our labor and delivery unit at forty weeks and two days of gestation complaining of regular painful uterine contractions. Her last menstrual period was January 3, 2019, which gave her an estimated due date of October 10, 2020. She reported adequate fetal movement, no loss of fluid from the vagina, and no vaginal bleeding. The patient had no other complaints at that time. Her prenatal course was unremarkable- she was Rh-negative, received Rhogam at twenty-eight weeks gestation, her one-hour glucose challenge test was 112, she was immune to rubella and varicella, and had reactive antibodies to hepatitis B. Her group b *streptococcus* status was unknown. Urine cultures were negative throughout pregnancy.

The patient had no past medical or surgical history. She had a benign gynecologic history with menses beginning at age thirteen, with regular thirty day menstrual cycles. She denied any recent travel and denied tobacco, alcohol, or drug use. She had a past obstetric history of one normal spontaneous vaginal delivery of a 3317-gram female in 2011 accompanied by epidural analgesia, as well as one first-trimester spontaneous abortion, and one elective termination of pregnancy.

Upon admission to the labor and delivery unit her hemoglobin and hematocrit were 10.8g/dL and 31.9g/dL respectively, her platelets were 292,000 and her white blood cell count was 9.92. Her calcium, sodium, potassium, chloride, blood urea nitrogen, creatinine, and glucose were: 9.5, 134, 4.4, 103, 22, 10, 0.58, and 113 respectively. Her coagulation profile was normal with INR: 0.91, PT: 10.6, and PTT: 26.8. Her vital signs were stable, blood pressure was mildly elevated ranging from 120-140s/70-80smmHg, and heart rate ranging from 70-80 beats per minute.

Upon physical examination, the patient appeared well developed, well-nourished, and in no acute distress. The neck was supple, no thyromegaly, no masses. The external nose

had no mass or deformity, External ears showed no mass or deformity. The patient presented with normal respiratory effort, clear to auscultation, and regular sinus rhythm, S1 normal, S2 normal, with no murmurs, rub or gallop. She had a gravid abdomen with a fundal height of 40 centimeters. On vaginal examination, there were no lesions, or blood noted, her cervix was found to be 4-5 centimeters dilated, 60% effaced with the fetus at the -3 position. External fetal monitoring showed a category I fetal heart tracing with the fetal heart baseline of 140 beats per minute with moderate variability, positive accelerations and no decelerations. Uterine contractions were occurring every 3-5 minutes. The biophysical profile was 10/10. The fetus had an estimated gestational weight of 3863g.

The patient requested an epidural, which was slightly difficult due to her body habitus, however, it was successfully placed after two attempts. Status post epidural placement, the physician was called to bedside due to fetal bradycardia. She was examined and her cervix was found to be fully dilated with the fetus at the 1+ station. Artificial rupture of membranes was performed at that time and clear fluid was noted. The patient was instructed to push with her uterine contractions. Due to recurrent fetal decelerations, the decision was made to deliver the fetus with vacuum assistance. The vacuum was applied and after one pop-off, the head delivered followed by the shoulders. The pediatricians were present for the delivery and assigned APGARS of 9 and 9 at 1 and 5 minutes respectively. The placenta was then delivered with intact membranes. Upon inspection, a small first-degree laceration was noted and repaired with 3-0 chromic. Hemostasis was excellent at that time. The total estimated blood loss was 350cc.

The patient and neonate were transferred to the postpartum unit in stable condition. On a postpartum day one, the patient began to develop an occipital headache associated with neck pain that was worsened by movement. She was given intravenous fluid hydration as well as Fioricet which did not alleviate her pain. The patient was evaluated by anesthesia who recommended a blood patch for possible postdural puncture headache. The blood patch procedure was performed by anesthesia with some difficulty however it transiently relieved her symptoms.

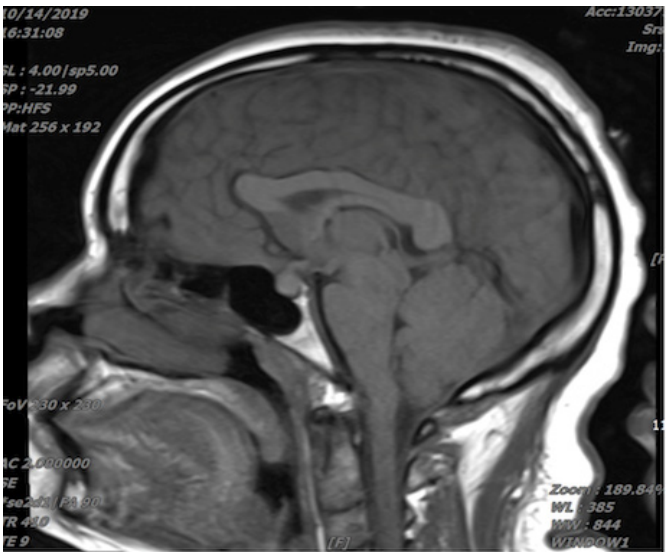

Figure 1: The cerebellar tonsils lie approximately 3 mm below the foramen magnum.

On postpartum day two, the patient was still reporting an occipital headache as well as neck pain that was worsened by movement. She was also complaining of photophobia and preferred to lay flat with the lights off. Due to the worsening nature of the headache, the obstetric team ordered brain imaging. Findings from the CT with and without contrast (figure 1) were as follows: "no intra-axial or extra-axial fluid collection or hemorrhage. There is no hydrocephalus or midline shift. Relative effacement of the cisterns may be age-related. There is no evidence of territorial infarction. There is a hypodensity of the brainstem at the pontomesencephalic junction. The cerebellar tonsils lie approximately at 3 mm below the foramen magnum. The mastoid air cells and imaged paranasal sinuses were clear. The imaged orbits were normal configurations. The impression from the radiologist was, that the hypodensity of the brainstem at the pontomesencephalic junction is likely artefactual, however, given the clinical context a follow-up MRI may be of benefit. The low-lying cerebellar tonsils and relatively effaced cisterns, could not be determined if it was an acute or chronic finding since no prior study was available for comparison. No other evidence of intracranial hypotension, however, CT is insensitive. An MRI with contrast should be obtained if clinical concerns for hypotension persisted."

Neurology was then consulted to which the patient denied any significant past medical history, or history of aneurysms, intracranial hypertension, headaches, migraines in her or her family. The examination performed by the neurologist revealed alert and oriented to time, place and person, with mild distress from the global headaches. She reported that lying flat on her stomach provided mild relief. Her cranial nerves II-XII were grossly normal and intact. Her motor strength was 5/5 in all four extremities. Her sensation was intact. Deep tendon reflexes were normal 2+. She did not want to stand up and walk due to pain. Neurology recommended MRI of the brain and MRV of the head with and without contrast as well intravenous hydration with normal saline and caffeine. Neurology checks were to be performed every four hours and if abnormal to re-consult the team.

The MRI of the brain was done on multiplanar and multisequence MR images of the brain without gadolinium. The findings were on brain parenchyma were as follows: "No MRI evidence of hemorrhage. No evidence of acute infarct. No intracranial mass or mass effect. There is the preservation of the gray/white matter interface. Borderline size pituitary gland with mild upper surface convexity. The internal auditory canals are patent. Posterior fossa structures are unremarkable. The CSF spaces were appropriate for age. No hydrocephalus. Basal cisterns are patent. The vascular system showed normal fluid voids in the major intracranial circulation. The calvarium, skull base, paranasal sinuses, and mastoid air cells, showed small mucous retention cysts in the maxillary sinus. No discrete lytic or blastic abnormalities. Orbits showed that both globes, extraocular muscles optic nerves, and retrobulbar fat appear unremarkable. Their impression of the MRI was no acute intracranial abnormality. Upon MRA no filling defects were found. No complete occlusion. No MRV evidence of dural venous sinus thrombosis."

On postpartum day four she had reported the headache had slightly improved but was still present. Anesthesia was

re-consulted and a decision was made to place another blood patch. After placement of the blood patch, she reported immediate relief. Neurology had also seen the patient and recommended continuing the IV hydration as well as beginning IV caffeine. They also suggested putting the patient in the Trendelenburg position when in bed and providing an abdominal binder. On postpartum day five and six, the patient had reported occipital headache that was worse with sitting up and ambulating despite the blood patch. Anesthesia and neurology were re-consulted. On postpartum day six and seven, neurology had examined the patient, which was unchanged since the previous exam. Neurosurgery was consulted regarding the Arnold Chairi malformation and there were no acute recommendations. Neurology had recommended discontinuing the IV medication as well as analgesics and beginning magnesium oxide 400mg ×1 maintained by magnesium oxide 200 mg BID as well as Naprosyn 250mg. On postpartum day eight, the patient's blood pressures were noted to be elevated: 130s-150s/80-90s, with an associated headache. Preeclampsia labs were drawn and her protein creatinine ratio was found to be 0.8. The decision was made to start magnesium sulfate therapy for postpartum preeclampsia. She received twenty-four hours of magnesium sulfate therapy without complications. She stayed in the hospital for an additional forty-eight hours after magnesium sulfate therapy with normal blood pressures. Neurosurgery saw the patient again and had no acute recommendations. The patient was eventually discharged home in stable condition with a resolving headache.

Discussion

Diagnosing Type 1 Arnold Chiari Malformation and reasoning it as a differential diagnosis in postpartum women with a headache may prove to be quite challenging. This is due to its rarity and vague symptomatology. When presented with a case of persistent headache status post vaginal delivery with failure of conservative measures and blood patch, care must be taken to consider Arnold Chiari Malformation as a potential cause.

The most common symptoms that one with type 1 Arnold Chiari will experience include sub-occipital headaches and neck pain. However, women with symptomatic Arnold Chiari can also experience less sought out symptoms such as dizziness, tinnitus, vertigo, double or blurred vision, hypersensitivity to bright lights, gait ataxia, and muscle weakness [2]. It is important to ask about these symptoms when evaluating women with persistent headaches in the postpartum period. Diagnostic imaging could also prove to be useful in seeking out a diagnosis.

The literature has hypothesized that type 1 Arnold Chiari malformations can be acquired iatrogenically, especially during neuraxial procedures. Although debates are still ongoing [8], this is very important to consider when discussing methods of pain relief in the intrapartum course. With acquired Arnold Chairi malformations being a potential postpartum complication, it should remain on one's differential when evaluating persistent headaches and neck pain.

Data Availability

The data used to support the findings of this study are available from the corresponding author upon request.

Conflicts of Interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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