Paroxysmal rage as a presenting symptom of the Chiari I malformation

Report of two cases

ROGER J. HUDGINS, M.D.

Department of Pediatric Neurosurgery, Scottish Rite Children’s Medical Center, Atlanta, Georgia

Signs of meningeal irritation including occipital and cervical pain are common in symptomatic children with the Chiari I malformation. The author reports on two children with Chiari I malformations who presented with a previously undescribed symptom presumably caused by intermittent meningeal irritation: paroxysmal rages. In both cases the rages stopped after decompressive surgery. Evaluation including magnetic resonance imaging should be considered in children with new onset of paroxysmal behavioral disorders.

KEY WORDS • Chiari malformation • rage • behavioral disorder • suboccipital craniectomy

The Chiari I malformation is characterized by herniation of the cerebellar tonsils below the foramen magnum. This has been called the “adult” Chiari malformation, but with the advent of magnetic resonance (MR) imaging, as well as a better understanding of the clinical presentation, most Chiari I malformations are now being diagnosed and treated during childhood. Children with a Chiari I malformation present with a variety of symptoms, including headaches, ataxia, dysphagia, crying, nonradicular shoulder or extremity pain, and loss of strength or sensation in the extremities. A presentation with paroxysmal rage has not been reported in the literature. Recently, two children who presented with intermittent violent rages were found to have Chiari I malformations. In both cases the rages ceased after decompression of the malformation.

Case Reports

Case 1

This 2.5-year-old boy presented with a several-month history of rage episodes in which he would suddenly and without warning become extremely irritable, upsetting furniture and biting and kicking those around him, including his parents. Prior to this he had been developing normally. These spells would last for seconds to minutes and then end as rapidly as they began. He had been treated with Mellaril and clonidine without effect. He underwent neurological evaluation, with the only significant finding being unsustained clonus and mild lower-extremity hypertonicity. Electroencephalographic findings were normal. An MR image revealed a Chiari I malformation in which the tonsils ended 20 mm below the foramen magnum. The patient’s brainstem was deviated anteriorly, and there was central lucency within the cerebellar tonsils (Fig. 1). No other abnormalities were noted on the MR image; specifically, the temporal lobes were normal. On the day before surgery the patient had attacked his mother, causing a large bruise on her upper arm from a bite. He underwent a suboccipital craniectomy and C1–2 laminectomy without duraplasty. He has had no episodes of rage in the 12 months since his surgery and is now developing normally.

Case 2

This 8-year-old girl presented with a 6-month history of episodic violent behavior in which she would run wild, scream, and be uncontrollable. During these outbursts she had kicked and punched people around her. These episodes were sudden in onset, but could last most of the day, and would occasionally end only when the child was exhausted. There was no history of headaches, weakness, or dysphagia. Examination was difficult because of her lack of cooperation, but no neurological abnormalities were detected. She had been treated earlier for acute lymphocytic leukemia and had been in remission for 3 years. Treatment for the acute lymphocytic leukemia included systemic and intrathecal chemotherapy (vincristine, methotrexate), but no cranial irradiation. The patient was admitted for psychiatric assessment, and an MR image was obtained, on which the cerebellar tonsils were demonstrated to be 18 mm below the foramen magnum (Fig. 2). The MR image, specifically the temporal lobes, was otherwise normal. The patient underwent a suboccipital craniectomy and C1–2 laminectomy without duraplasty. She has had no further rage episodes in the 8 months since surgery, and she now has a pleasant and engaging person-
ality. Although she has been considered to have a learning disability, her school grades have improved from mostly failing before surgery to mostly average following decompression of her Chiari malformation.

Discussion

The most common presenting symptom of the Chiari I malformation is pain, usually occipital or cervical, but occasionally poorly localized. The pain is believed to be caused by irritation of nerve fibers in the meninges of the foramen magnum and upper cervical dura due to the presence of the abnormally low cerebellar tonsils. This pain is commonly induced or exacerbated by Valsalva maneuvers or coughing, both of which cause transient elevations of intracranial pressure. This syndrome is probably the cause of the irritability and crying seen in infants with the Chiari malformation.

Episodic cervical discomfort or pain can manifest in various ways, depending on such factors as maturity, personality, and social setting. The paroxysmal rages and antisocial behavior exhibited by these two children appear to have been a reaction to the pain caused by the displaced cerebellar tonsils. The intermittent nature of these episodes may have been caused by transient increases in intracranial pressure. This syndrome is probably the cause of the irritability and crying seen in infants with the Chiari malformation.

Distinguishing rage attacks caused by the Chiari malformation from those associated with acting out or psychiatric problems is important. In both of these children, the rages were sudden in onset: the child was normal one moment and kicking and biting the next. Additionally, in both cases the parents stated that the behavior developed suddenly and without prior behavioral abnormalities. It might be prudent, therefore, to consider MR imaging in cases of paroxysmal rage or violent behavior.

It should be noted that in both cases the surgery performed did not include dural opening or expansion. The marked clinical improvement seen postsurgery confirms that compression in the Chiari I malformation is related to bone and not to the dura.

References


Manuscript received January 25, 1999.
Accepted in final form March 29, 1999.
Address reprint requests to: Roger J. Hudgins, M.D., 5455 Meridian Mark Road, Suite 540, Atlanta, Georgia 30342. email: Rogerhud@aol.com.