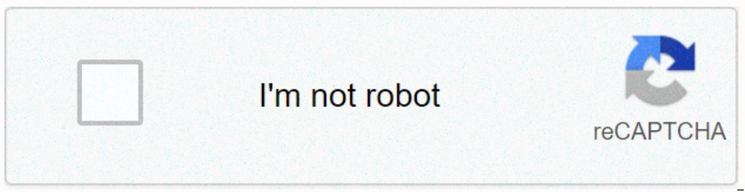

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Chiari malformation cluster headaches

Cluster headache (CH) is considered a primary headache syndrome and therefore neuroradiological central nervous system studies should be normal in primary CH patients (1). However, symptomatic cluster-like headaches (CLH) have been reported in conjunction with various disorders such as aneurysms, arteriovenous malformations, cervical trauma, medullary infarctions, inflammatory/infectious diseases, and tumors (2). CLH is described in only one case report in the relevant literature (3) in relation to Chiari type 1 malformations (CM-1). Here we describe a case diagnosed with CLH associated with symptomatic CM-1. A 28-year-old female patient had been hospitalized with recurrent headaches since the age of 20. She had severe, recurrent, pain in the right frontal and periorbital areas and headaches, lasting about 20-90 minutes. She also suffered from ipsilateral tearing, rhinoceros and ptosis during headaches. These pain attacks occurred irregularly two to four times a day. Although this attack usually appeared spontaneous, it was precipitated by head or neck movements. Additionally, she also suffered from occasional pain in the occipital-suboccipital area, dizziness, blackout of the eyes, palpitations, sweating; which disappears in 5-10 minutes. Neurological examination revealed vertical and down-beat nystagmus, positive Romberg's test, and hyperreflexia. Her cardiology tests were normal. Cranial and cervical MRIs revealed CM-1 (Figure 1). She responded well to methyl prednisolone 60 mg/day (10 days), which was tapered in a month period. We believe that CM-1 can be interpreted as symptomatic in this patient. While CM-1 may be asymptomatic, clinical manifestations, which typically begin in adolescent adulthood, may include headaches, visual disturbances, neuro otological complaints, lower cranial nerve dysfunction, and sleep apnea (4). Among manifestations of CM-1, headache is one of the most common symptoms, occurring in 15-98% of patients. The known headache spectrum in CM-1 includes cough headaches, strenuous headaches, low cerebrospinal fluid pressure headaches, prolonged headache attacks, suboccipital headaches, and migraine attacks (4). While CM-1 and CH seemed like independent diseases, determining the actual cause of CH is obviously difficult when these two pathological processes co-occur. This raises the question of whether the pathogenesis of these diseases may be linked. With the current precision of central nervous system imaging, CM-1 can be a random finding when MRI is performed for other reasons. In CM-1, variable tail displacement of cerebellar tonsils occurs in the upper cervical canal. These structural abnormalities in CM-1 may include stretching of cranial nerves or direct compression of the brain stem nuclei, compression of the posterolateral part of the medulla and the upper cervical spinal cord, vascular distortion in the areas watered by vertebral and posterior inferior cerebellar arteries, on the root plains of C1, C2 and vagus nerves (4,5). Although ch's pathophysiology remains indeterminate, it has a neuronal component with the involvement of the trigeminal nerve (1). The pain of CH may be associated with dysfunction in an area of the brainstem and/or craniospinal pressure exociation, stimulating pain-sensitive structures in patients with CM-1. Therefore, we believe that CM-1 may be a factor associated with symptomatic cluster headache.1. Domitrz I, Gawel M, May E. Cluster headache a symptom of various problems or a primary form?. A case report. Neurol Neurochir Pol. 2013;47:184-188. [PubMed] [Google Scholar] 2. Mainardi F, Trucco M, Maggioni F, Palestini C, Dainese F, Zanchin G. Cluster-like headache. A comprehensive review. Cephalalgia. 2010;30:399–412. [PubMed] [Google Scholar] 3. Seijo-Martinez M, Castro del Río M, Conde C, Brasa J, Vila O. Cluster-like headache: association with cervical syringomyelia and Arnold-Chiari malformations. Cephalalgia. 2004;24:140–142. [PubMed] [Google Scholar] 4. Kaplan Y, Oksuz E. Chronic migraine associated with Chiari type 1 malformations. Clin Neurol Neurosurg. 2008;110:818–822. [PubMed] [Google Scholar] 5. Khurana RK. Headache

spectrum in Arnold-Chiari malformations. Headache. 1991;31:151–155. [PubMed] [Google Scholar] Chiari malformations are a neurological disorder that occurs from an abnormality in the brain's small brain, which causes it to push through the opening of the skull into the spinal area. This malformation causes pressure on both the brain and spinal canal, which can cause headaches, neck pain and other symptoms. Although Chiari malformations are rare, there are different subtypes. The most common is identified as type I and is congenital (present at birth). Symptoms for this type typically develop in adolescence or early adulthood as the brain and skull grow. Other subtypes such as type II are also innate, but are detected in vitro or at birth and most often affect children with neural tube defects such as spina bifida. Chiari malformations are more common in women than men. What causes chiari malformations? Chiari malformations are caused by the faulty development of the brain during pregnancy. While present at birth, it often goes undetected, because symptoms usually do not present themselves until later stages of growth in adolescence. In rare cases, acquired Chiari malformations can develop in adults as a result of trauma, injury or infection. Symptoms Many patients with Chiari malformations type I never experience symptoms, and the condition is detected during diagnostic imaging tests. For those with more severe malformations, symptoms include: Headache, neck and lower back pain, which intensifies during physical exertion Sleep apnecibility and difficulty swallowing Dizziness, loss of balance and limb weaknessPresidentens of a syrinx – a fluid-filled sac on the spinal cord that is closely associated with Chiari malformation diagnosis on the face and Headache Clinic, we believe the best physical and emotional outcomes for our patients begin with our expert diagnostic team. The board-certified neurological specialists at UTHealth Neurosciences are trained across several medical disciplines, including neurology and neurosurgery, and are highly qualified in rapidly and thoroughly diagnosing patients with Chiari malformations through a series of tests and procedures, which may include: Neurological exams in the eyes, vision, muscle strength and reflexesMRI (magnetic resonance imaging - a test that uses magnetic waves to create images of structures inside the head) CT (computed tomography - a series of X-rays taken from different angles to produce cross-sectional images) treatment for Chiari malformations Each patient's treatment options are unique and will vary based on their overall health, age and other factors such as the severity of symptoms and the degree to which they affect quality of life. Many patients with Chiari malformations do not experience any symptoms, and no treatment will be required other than watchful monitoring through regular exams. Common treatments for Chiari malformations include: Posterior fossa decompression-a surgical procedure to relieve pressureElectroe cautery to narrow the lower cerebellumSpinal laminectomy to increase the size of spinal canalMedications to manage pain Related conditions and treatments Contact us at UTHealth Neurosciences, we offer patients access to specialized neurological care at clinics throughout the greater Houston area. To ask us a question, schedule an appointment or learn more about us, please call (713) 486-8000 or click below to send us a message. In case of emergency, call 112 or go to the nearest emergency room. More than a tongue-twister, Arnold-Chiari malformations are a well-described innate (present at birth) abnormality of the small brain and brainstem. While four different types have been described, types 1 and 2 are most common. Type 2, 3, and 4 are typically diagnosed in childhood due to severe abnormalities involving the skull or spinal cord. However, type 1, considered a milder form of the condition, may not cause symptoms until adulthood, if ever. In all types, changes affect the posterior part of the brain and spinal cord. Part of the small brain (balance center of the brain) squeezes through the base of the skull next to the spinal cord. This can cause changes in the spinal cord and compression of the small brain and its circulation. Headaches and other symptoms are common. Headaches associated with Arnold-Chiari can take various forms, including an intermittent form or a persistent one. The pain can be frontal (as in the forehead area) or in the neck or back of the head area. Characteristically, but not always, the pain can worsen with neck movement, coughing, or straining movements. Associated is a frequent complaint. When investigated, other problems may be found lack of coordination, difficulty swallowing, impaired eye movements, sensory disturbances and others. Arnold-Chiari is diagnosed with an MRI scan of the brain and spinal cord, but cannot be identified by a CT scan. If a severe malformation is present, surgery may be needed to remove pressure on different parts of the brain and spinal cord and relieve associated symptoms. If surgical intervention is not required, treatment is typically based on symptom control or relieving pressure. Pres.