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Management of Arnold-Chiari Malformation; After Long Period of Headaches, Syncope and Trauma: A Report of 3 Cases

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Abstract
Chiari malformation type 1 (CM1) is a condition in which the cerebellar tonsils move downward into the spinal canal, affecting the area where the skull meets the neck. When there are signs of a tonsillar obstruction or herniation >5–14 mm, which happens when the tonsillar structures go through the posterior cranial fossa, symptoms start to show up. We report a case of a 65-year-old woman with headache, dizziness, and sensory disturbances in the extremities. Therefore, diagnostic imaging was recommended: MRI revealed a Chiari type I malformation (CM1) with tonsillar descent >5 mm, syringomyelia from C2 to T4, and the presence of syringomyelia. In the second case, we present a 56-year-old woman with persistent headache, neck pain, and dizziness, so diagnostic imaging was recommended: MRI revealed a decrease in the tonsils through the foramen magnum, confirming Chiari type 1 malformation with the presence of syringomyelia in the spinal cavity. In the third case, a 49-year-old woman presented with a history of dizziness, falls, and sensory disturbances. She had experienced worsening symptoms for five years, culminating in a recent fall due to syncope and trauma. Imaging and diagnosis: Magnetic resonance imaging showed that she has Chiari malformation (CM1) with a 6-mm drop in the cerebellar tonsils and signs of increased pressure inside the skull. The diagnoses was confirmed by the presence of syringomyelia in the spinal cavity. These cases demonstrate that CM1 can escalate to a surgical emergency, necessitating timely imaging diagnostics, including MRI or CT scans, to confirm the diagnosis and determine the appropriate surgical course. to enlarge the posterior cranial fossa through laminectomy, thereby alleviating neural compression.

Keywords Arnold-Chiari malformation, Posterior Cranial Fossa, Decompression, Headache, Laminectomy

Introduction

Chiari malformation type 1 (CM1) is a pathological condition involving the displacement of the cerebellar tonsils below the foramen magnum into the spinal canal, affecting the craniocervical junction [1]. Foramen magnum decompression (FMD) was first reported in 1930 as a so-called secondary phenomenon in patients with idiopathic intracranial hypertension, hydrocephalus, CSF leak, pseudomeningocele, or descending hernia with decompression defect. [2]. Arnold-Chiari malformations are classified into four

types, with CM1 and basilar invaginations being the most prevalent forms affecting the craniocervical junction in adults. First described by pathologists Julius Arnold and Hans Chiari in the late 19th century, this condition remains a complex clinical syndrome characterized by cerebellar and brainstem deformities [3]. CM1 has a prevalence of approximately 1 in 1,000 individuals, with 1 in 100 meeting radiological diagnostic criteria. New imaging techniques and diagnostic criteria have significantly improved the understanding of CM1's comorbidities, symptomatology, and surgical treatment approaches [4].

Four principal mechanisms have been proposed for the pathogenesis of CM1:

1. Excessive tissue in the posterior cranial fossa due to tumours.
2. Hemodynamic changes leading to increased intracranial pressure.
3. Insufficient growth of bony structures in the posterior cranial fossa.
4. Displacement or lowering of central nervous system structures due to reduced intrathecal pressure [5].

Case Presentation

Case report 1

We report A 65-year-old woman presented with headaches, dizziness, and sensory disturbances in the extremities. Initial conservative treatments were ineffective, and neurosurgical intervention was recommended. Imaging and Diagnosis: MRI revealed Chiari type I malformation (CM1) with tonsillar descent >5 mm, C2 to T4 syringomyelia, stage 2 hypertension, and intervertebral disk herniations (C5-C7). Confirmed Chiari type 1 with the presence of syringomyelia in the spinal cavity and paresthesia in the right arm, hand, and fingers. Surgical Procedure: Microsurgical decompression of the posterior cranial fossa was performed with a C1 laminectomy. A dural defect was sutured with Prolene, and haemostasis was achieved using a haemostatic patch. During the follow-up, the patient's recovery was stable and without any eventuality; our decision was discharge.

Case report 2

A 56-year-old woman presented with persistent headaches, neck pain, and dizziness, along with stage III hypertension. Conservative treatment failed, and surgery was necessary. Imaging and diagnosis: MRI findings revealed indirect signs of increased intracranial pressure. With decreased tonsils through the foramen magnum. Chiari malformation type 1 was confirmed with the presence of syringomyelia in the spinal cavity. Surgical procedure: Microsurgical decompression of the PCF was performed, with a C1 laminectomy. A dural defect was sutured with Prolene, and haemostasis was achieved using a haemostatic patch, achieving reversibility of symptoms. During the follow-up for ICU, the patient was stable and we decided to discharge them.

Case report 3

A 49-year-old woman presented with a history of dizziness, falls, and sensory disturbances. She had experienced worsening symptoms over five years, culminating in a recent fall for syncope and trauma. Imaging and diagnosis: MRI confirmed Chiari malformation (CM1) with 6 mm cerebellar tonsillar descent and indirect signs of increased intracranial pressure. It was confirmed by the presence of syringomyelia in the spinal cavity. A postoperative MRI scan of the brain. The procedure was performed with decompression of the posterior cranial fossa at the level of C1. Postoperative follow-up of the surgery: decompression of the craniovertebral junction. The midline structures are not displaced. There are no data of acute intracranial pathology. The patient was discharge and follow by consultation.

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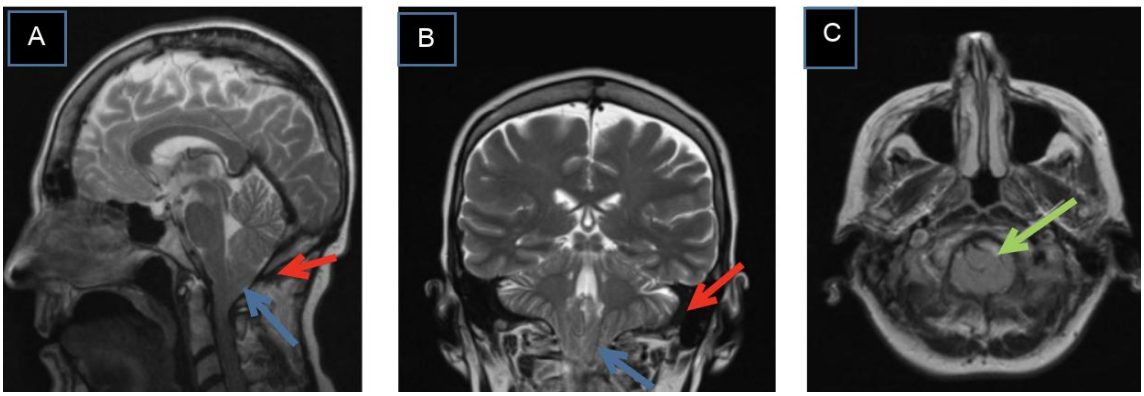

Figure 1. a) Sagittal MRI, with Arnold-Chiari malformation, with a tonsillar herniation below the foramen magnum (McRae's line); red and blue patients with severe headaches with paraesthesia of the right arm, hand and fingers. b) Coronal imagen, visualizing displacement of cerebellar tonsillar structures; blue and red arrow. c) Axial image showing posterior displacement of the tonsils downwards from the foramen magnum.

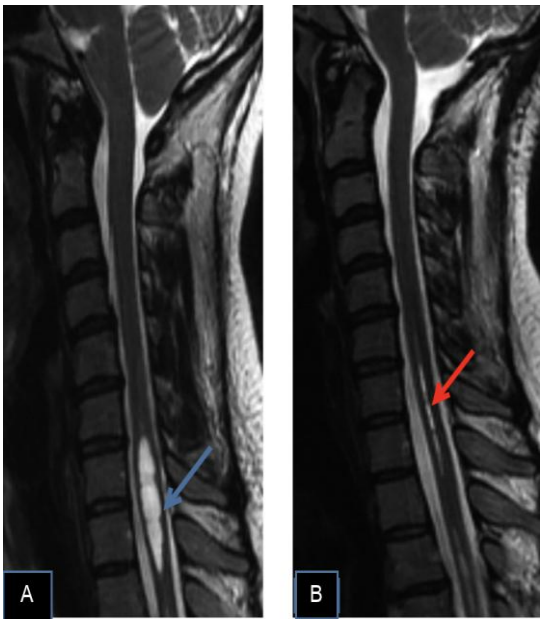

Figure 2. a) Chiari malformation type 1, spinal syringomegaly with tonsillar herniation 6 mm below the foramen magnum (blue arrow). b) Descending tonsils through the foramen magnum and spinal syringomegaly (red arrow).

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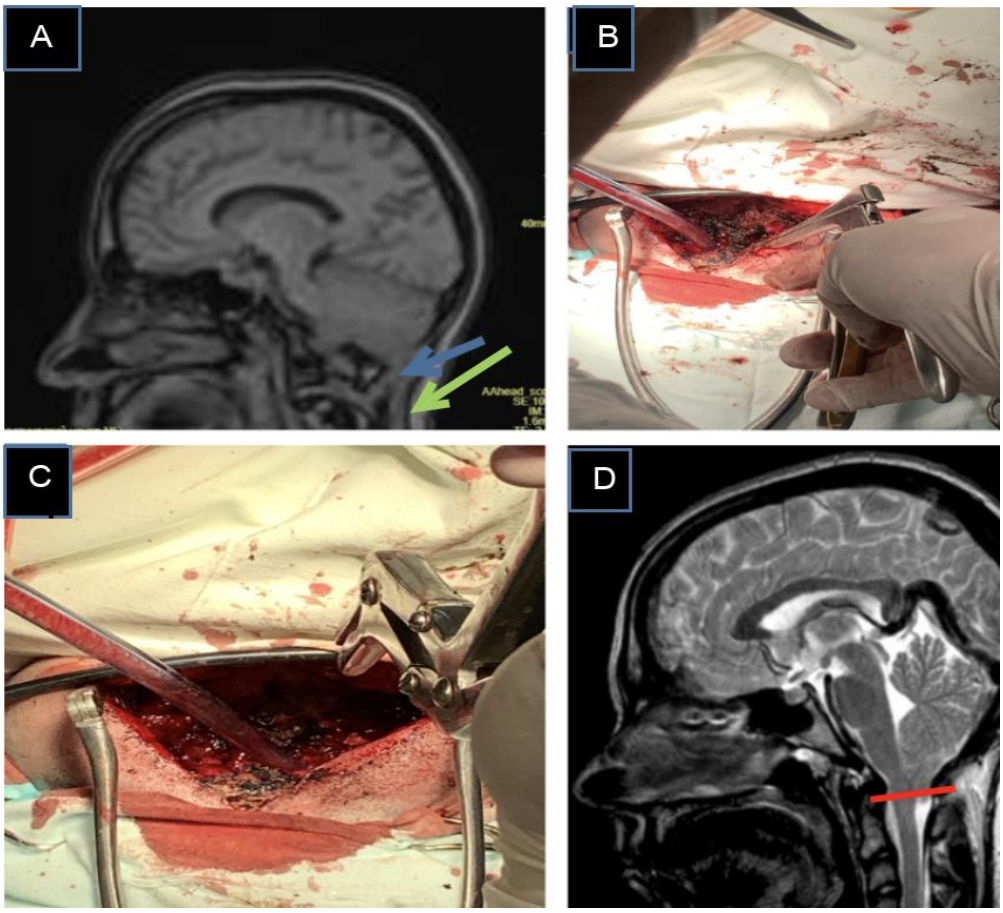

Figure 3. a) Sagittal T1 MRI of a patient with a diagnosis of Arnold-Chiari malformation type 1; b) head fixation with a Mayfield clamp in the occipitocervical region. Decompression of the craniovertebral junction posterior fossa by Kerrison Cervical Rongeur. c) Partial decompressive resection of the occipital bone and vertebral arches of C1, suction of the bleeding and haemostasis with wax, and decompression of the neural structures by the atlantooccipital membrane. Stille bone Rongeur. d) T2 MRI, with descending tonsils through the foramen magnum.

Discussion

In an analysis of 130 articles, 23 patients met inclusion criteria, examining CM1 cases associated with syringobulbia. Symptoms such as headache, neck pain, cranial nerve palsies, limb weakness, dysesthesia, Horner syndrome, cerebellar ataxia, and breathing disorders were commonly observed. The medulla oblongata was identified as the most affected structure, with syringomyelia frequently present. Surgical decompression of the PCF or foramen magnum, often combined with cervical laminectomy and duraplasty, was the most effective intervention [8, 9]. Patients who underwent early surgical intervention prior to spinal cord compromise experienced significant symptomatic improvement, even in cases with syrinxes ≥6 mm. The decision to operate is largely dependent on the stabilization of spinal cord damage and the potential for postoperative complications [10, 19].

Basilar invagination, often secondary to repetitive microtraumas, contributes to musculoskeletal instability and compression of adjacent neural structures. Decompression and stabilization effectively mitigate these effects. The pathophysiology of basilar invagination highlights the importance of addressing repetitive mechanical stressors to prevent further deterioration [11]. The first documented modern case of CM1 utilizing advanced imaging techniques was reported by Gebarski and Greenberg in 1984. A 40-year-old woman presented with cerebellar atrophy, gait disturbances, and visual impairment and was ultimately

diagnosed with olivopontocerebellar degeneration. Imaging revealed cerebellar tonsillar herniation, and angiography identified displacement of the posterior inferior cerebellar arteries (PICA). The patient underwent suboccipital craniectomy and C1–C2 laminectomy with a dural graft, resulting in significant recovery of gait and vision within two months [12]. Decompressing the C1 vertebra is particularly effective in expanding the PCF, restoring cerebrospinal fluid (CSF) flow, and reducing syrinx volume in CM1 patients. Duraplasty further enhances these outcomes by preventing complications such as CSF leakage or fistula formation. However, failed duraplasty may compromise CSF to optimize the benefits of decompression [13].

- CM1 (classic CM1): This is a type of malformation that manifests in the central nervous system at the caudal level of the cerebellar tonsils, with a displacement greater than 5 mm toward the foramen magnum, with or without the presence of syringomegaly. [20, 21]
- CM2: This is a congenital malformation associated with spina bifida and other brain abnormalities, such as aqueductal kinking and polymicrogyria. [22] Spina bifida is accompanied by myelomeningocele, which is a congenital malformation due to incomplete closure of the neural tube of the central nervous system, considered serious due to its embryonic development. [23]
- CM0 is simply an obstruction of the cisterna magna by arachnoid adhesions. The posterior cranial fossa will have a smaller volume due to compression of the cerebellar tonsils through the foramen magnum and the presence of syringomegaly in the cervical spinal cord. [24] Chiari malformation 1.5 (CM 1.5), such as the presence of this malformation between Chiari type 1 and Chiari type 2, with a neural tube defect [25].

Headaches are very common in Chiari malformation type 1, even in other regions or locations, among which suboccipital headaches show no relationship with the Valsalva maneuver, but there are these types of cough-induced suboccipital headaches, as a characteristic of (CM1) [26]. A retrospective study conducted between 2007 and 2019 comprised 107 patients with Arnold-Chiari malformation type 1 (CM1), with hernia displacements measuring an average length of 13 ± 5 mm (range: 5–24 mm). Among these patients, 38 had duraplasty at the C1 level as part of their surgical therapy. Syrinx volumes were analyzed using magnetic resonance imaging (MRI) and three-dimensional visualization software. Following surgical decompression, a 20% expansion of the posterior cerebral fossa (PCF) at the inferior medial level was detected, coinciding with a reduction in syrinx volume. At the C1 level, an additional 3% increase in volume was attained, which further expanded to 5% with the inclusion of duraplasty ($p = 0.029$). Notably, the procedure exhibited low problems ($p = 0.526$). Monthly increases in syrinx volume varied between 5.9% and 7.5% in 10 patients who received solely C1 laminectomy ($p = 0.040$), reinforcing the efficacy of combining this surgery with duraplasty. [6, 27].

Another study examined the connection between craniovertebral junction (CVJ) abnormalities and syringomyelia in CM1 patients treated at a university hospital between 2012 and 2017. The study covered 64 patients, of whom 59% were women. Among these patients, CVJ abnormalities were more prominent in those with basilar invagination (71%, $p = 0.012$), followed by platybasia (44%) and cervical kyphosis (33%). Additionally, CVJ abnormalities were more frequently related to syringomyelia ($p = 0.045$), excluding individuals with concurrent hydrocephalus [7]. [28]. [29]. Our case collection emphasizes a substantial link between the delayed onset of symptoms in adults with CM1 and the severity of cerebellar tonsillar descent. In one of our patients, the development of symptoms was precipitated by a traumatic concussion, which aggravated the underlying disease over an extended period. demonstrating that many adult CM1 patients remain asymptomatic until the cerebellar tonsils drop more than 5-6 mm beyond the foramen magnum, causing displacement and compression of central nervous system components.

Table 1. Patients with Chiari type 1 (CM 1), with more than 3-14 mm of tonsillar herniation, treated with posterior cranial fossa decompression plus C1 laminectomy and/or cranioplasty.

Author	Study	Year	Diagnose	No. patients	Main symtoms	Tonsillar herniation	Procedure	P value
Gündağ -Papaker M. et al. [14].	Retrospective	2021	CM1	45	Headache		posterior fossa decompression and duraplasty	p<0.05
Kumar A. et al. [15].	Prospective longitudinal study	2019	CM1	30	Neck pain and weakness	10.4 ± 4.64 mm that reduced to 7.35 ± 3.10 mm	suboccipital decompression	N/A
Feghali J. et al. [16].	retrospective	2020	CM1	175	Headache	>10 mm of herniation (*n* = 18	suboccipital decompress	0.016
Saindane M. et al. [17].	Retrospective	2014	CM1/unilateral or bilateral transverse sinus stenosis	30	Headache	IQR, 7.0 mm below to 14.0 mm below) -basion	suboccipital decompress	*p* < 0.001
Zarrin D. et al. [18].	Retrospective	2024	CM1	46	Headache	13.03 mm ± 5.7 mm	C1 laminectomy	< 0.05.

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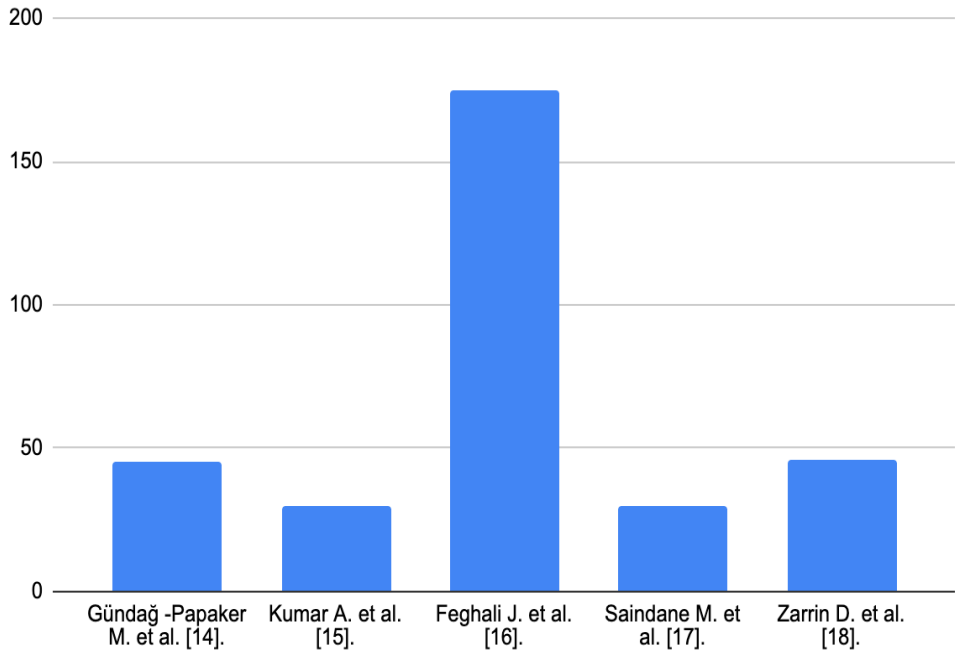

Figure 4**.** Graphs of studies of Chiari syndrome, with management and surgical intervention

Surgical Approach

The patient was in the prone position, and after fixing the head in a Mayfield clamp and treating the operative field using the standard technique, a linear incision was made in the projection of the occipitocervical region. Decompressive partial resection of the occipital bone and C1 vertebral arches was performed. Profuse bleeding from the bone tissue was noted. Haemostasis with wax. Compression of the neural structures by the atlantooccipital membrane was revealed. The latter was opened, compression of the neural structures was eliminated, and their distinct pulsation appeared. A defect of the dura mater was revealed, which was sutured with an atraumatic 6/0 prolene thread. Haemostasis using the haemostatic agent "HemoPatch" (45x90 mm). Haemostasis control: dry. A drainage drain was installed through a counter-opening in the soft tissue bed. Layer-by-layer sutures on the wound (Figure 1).

Our research on the symptoms of Chiari malformation is contingent upon the extent of tonsillar displacement into the foramen magnum; symptoms will be attenuated and will only emerge when the tonsils descend, resulting in compression of the anatomical structures within the posterior cerebral fossa. Consequently, the existence of this illness may remain undetected for years. The initial symptom is a common headache, frequently disregarded by the patient, but it escalates in frequency to two to three occurrences per week, prompting them to seek urgent assistance in the emergency department. Consequently, imaging investigations will provide clearer confirmation for visibility and the determination of surgical intervention.

Conclusion

This case series highlights the prolonged asymptomatic nature of CM1 in many patients and its eventual progression to symptomatic stages following triggers such as trauma. For instance, one of our patients developed symptoms after a traumatic concussion, while another presented with longstanding extremity pain that prompted emergency intervention. These cases demonstrate that CM1 can escalate to a surgical emergency, necessitating timely imaging diagnostics, including MRI or CT scans, to confirm the diagnosis

and determine the appropriate surgical course. Surgical decompression, particularly at the C1 level, is strongly recommended to enlarge the posterior cranial fossa through laminectomy, thereby alleviating neural compression. Incorporating duraplasty with grafts, when feasible, is critical to preventing CSF leakage and associated complications such as fistula or infection. Postoperative care, including conservative treatment and regular follow-up, is essential for monitoring recovery and rehabilitation. Follow-up intervals of one to six months are suggested to assess the progression of recovery and address any emerging complications. This study underscores the importance of early intervention and a multidisciplinary approach in the management of CM1 after the manifestation of the symptoms to ensuring the optimal patient outcomes through surgical precision and postoperative care.

Competing interest

The authors have no competing interest in this report.

Ethical Approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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No

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No

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