Case Report

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Type 1 Arnold Chiari Malformation With Syringomyelia: A Case Report

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Abstract

Objective: Arnold Chiari Malformation is a pathological herniation of the hindbrain through the foramen magnum into the cervical canal, characterized by herniation of the inferior cerebellar tonsils of 3-5mm 30-70% accompanied by syringomyelia.

Methods: This case presents a 16-year-old woman who came to the Neurophysiology Section of Wahidin Sudirohusodo Hospital with complaints of weakness and numbness in her left arm since last year. It all started with pain in the back of the head and neck after a heavy sneeze three years ago, and since then, the pain has been inconsistent. The patient has gone to the doctor and was given medication, but his symptoms are not improving. Clinical manifestations get worse after the patient uses a computer or mobile phone. Physical examination revealed atrophy of the thenar and hypothenar muscles and sensory dissociation in the left arm's 5th cervical toa first thoracic dermatome. NCV and F-wave examination showed no signs of radiculopathy, plexopathy, or neuropathy. Furthermore, cervical magnetic resonance imaging (MRI) showed Arnold Chiari Malformation Type-1 with syringomyelia extending from cervical spinal cord segment-2 to thoracic-1.

Result: This patient was diagnosed based on history, physical examination, nerve conduction study, and MRI spine. There was no clinical improvement after therapy.

Conclusion: The clinical picture of Arnold Chiari Malformation Type-1 is so varied that it requires detailed information to diagnose.

Keywords: Arnold Chiari Malformation Type-1, Sensory Dissociation, Syringomyelia.

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Introduction

Arnold Chiari Malformation is a pathological herniation of the hindbrain through the foramen magnum into the cervical spinal canal. Arnold Chiari malformation is easier to recognize with the increasing availability of MRI in health facilities.[1] Arnold Chiari malformation is classified into four types. Classification is made based on the morphology of the malformations that occur.[2] Type 1 with 5mm herniation of the caudal end of the cerebellar tonsil through the foramen magnum. Type 2 is characterized by herniation of the brainstem, fourth ventricle, and caudal end (>5mm) through the foramen magnum with spina bifida. Type 3 is characterized by cerebellar herniation, with or without brainstem herniation through the posterior encephalocele. Type 4 is characterized by cerebellar hypoplasia or aplasia with a normal posterior fossa and no herniation of the hindbrain.[1,2]

Arnold Chiari Malformations Type-1 are characterized by cerebellar tonsillar herniation inferiorly into the cervical canal of about 3-5mm, which can be seen on a sagittal MRI. The pressure of the tonsils on the foramen magnum anatomically and physiologically obstructs cerebrospinal fluid, which usually flows between the posterior fossa and the cervical subarachnoid space.[1] Arnold Chiari Malformation can be a congenital disorder and also Clinical symptoms of Arnold Chiari Type 1 malformations generally include headache or neck pain that worsened with Valsalva activity or maneuvering or signs of brain stem compression.[3] Syringomyelia can be associated with Type 1 Arnold Chiari malformations, and these patients usually present with signs of spinal cord dysfunction. The most common malformation type is believed to have cerebellar tonsillar dislocation more than 5mm below the foramen magnum in adults and 6mm in children.[4] This case report shows treatment of anterior crossbite with fixed orthodontic appliance. The brain stem is normal, and a syrinx can accompany this malformation, but sometimes it does not.[1,3,4,5]

Among patients with this type of malformation, about 14% -30% of these patients are asymptomatic until they reach adolescence and adulthood.[5] The onset of symptoms can occur spontaneously or due to trauma.[6] The prevalence of congenital type 1 Arnold Chiari malformations reaches 1 per 1,000 births but seems to be higher.[5-8] Many theories have explained the development of Chiari malformations without hydrocephalus. Currently, the development theory focuses on the difficulty in balancing the cerebrospinal fluid pressure waves during the Valsalva Manoeuvre.[1] Prolonged intra-

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cranial hypertension can result in downward migration of cerebellar tonsilla, which results in obstruction of normal cerebrospinal fluid flow between the posterior cranial fossa and the cervical subarachnoid space.[1] Conditions that inhibit the physiological flow of cerebrospinal fluid in Magendie's foramen trigger the formation of malformations, for example, arachnoid veils or adhesions. The clinical manifestations of the Arnold Chiari malformation can be classified into three categories: signs or symptoms related to brain stem compression, signs or symptoms related to cerebellar compression, and spinal cord dysfunction due to syringomyelia.[1]

Generally, patients manifest pain in the occipital or cervical area, which is localized in nature. Clinical symptoms usually include dysesthesia in cervical dermatomes 2.[5,7] Neck pain or headache often results from an activity or after coughing or sneezing (Induction of Valsalva).[1,4,5,7] MRI can easily confirm the diagnosis of type 1 Arnold Chiari malformations. The presence of syringomyelia in about 30% - 70% of patients should be determined by MRI in all spinal cord segments.[7] CT scan can further determine abnormalities in the bones, and explicit photographs can help evaluate tissue stability.[1,5,7]

Medical therapy has proven to be unable to overcome Chiari malformations. Various therapeutic paradigms have emerged.[1] The first decision to make is whether the lesion is genuinely symptomatic. Observation is needed in asymptomatic patients without a syrinx. Surgical intervention is recommended in symptomatic or asymptomatic patients with syringomyelia.[1] The degree of brainstem compression and tonsillar herniation must also be noted. The most common surgical procedure for this malformation is the decompression of the posterior fossa.[8] The goal of surgery is to enlarge the posterior fossa and reconstruct the Magna cisterna so that normal cerebrospinal fluid flow from the posterior fossa to the cervical subarachnoid space can occur. In most patients with Chiari malformations with a syrinx, syrinx size will be reduced and does not require immediate treatment because the decompression of the posterior fossa has overcome the pathological process caused by syringomyelia.[1,8]

Case Report

A 16 years old woman came to the neurology clinic of Wahidin Sudirohusodo Hospital on May 8, 2019, with complaints of weakness and cramps in her left hand, which was experienced about one year ago. These complaints were felt, mainly if the patient used a cellphone or computer for a long time. The patient also complained of pain that felt like a puncture. It spread from the left shoulder to the tip of her left hand. This complaint began with head and neck pain that occurred suddenly after the patient sneezed loudly about three years ago. After that, the pain in the head andneckdisappeared.Itwouldbereappeared,especially if the patient was coughing or sneezing. There was no history of fever, head trauma, nor family history of the same complaint.

General physical examination results were within normal limits. Neurological examination showed a decrease in the movement of the left superior extremity, and the strength was also decreased (4+). We also found atrophyoftheleftthenarmuscleandthelefthypothenar muscle. From the sensory examination, we found a sensory dissociation as high as left cervical dermatome 5 to thoracic 1 (C5-T1), proprioceptive function within normal limits.

Nerve Conduction Velocity (NCV) and F-wave examinations did not show any radiculopathy, plexopathy, or neuropathy.Magnetic resonance imaging (MRI) without contrast on cervical vertebrae showed tonsillar herniation accompanied by syringomyelia, and it is suitable for the image of Arnold Chiari Malformation Type-1.

This patient was diagnosed as Arnold Chiari Malformation Type-1 according to radiological imaging (MRI). The patient was given Meloxicam 7.5 mg/12 hours/orally and Gabapentin 300 mg/24 hours/oraly to treat pain in her left hand. The patient was also consulted to the Neurosurgery Department and was advised to undergo surgery, but the patient and family refused.

Discussion

In all cases of Chiari malformations Type-1, about 30-70% of patients have bone abnormalities at the craniocervical junction. In addition, small and narrow posterior fossa are also often found. It limits the space for normal cerebellum development before causing 'overcrowding' and tonsillar herniation under the foramen magnum.[9]

Physiological activities such as coughing or sneezing or Valsalva maneuvers exert a pulsatile pressure effect on the cerebellar tonsils, give a pistol-like force to obstructive cerebrospinal fluid, and direct the fluid into the interstitial, spinal cord, or even into the canal spinal cord, or even into the central canal, where the liquid empties.[9] The 'water hammer' mechanism provides a rational explanation of the clinical symptoms experienced by the patient. Based on the history, the patient only complained of neurological symptoms after loud sneezing. Sneezing has a mechanism similar to that of the Valsalva maneuver, and pulsatile pressure due to repeated sneezing produces a 'water hammer' effect that causes signs of spinal cord dysfunction.[9]

Initially, the patient was suspected of suffering from plexopathy due to clinical symptoms experienced by the patient, such as neck pain, weakness, and cramps in the left hand. After physical examination, there was a

sensory dissociation as high as cervical dermatome 5 to thoracic 2. In addition, this patient also underwent Nerve Conduction Velocity (NCV) and F-wave examination and found no signs of radiculopathy, plexopathy, or neuropathy, so that we thought about a central lesion of the complaints experienced by the patient.

A decrease in pain and temperature characterizes sensory dissociation in this patient, but the sensation of light touch, vibration, and position are within normal limits. This clinical finding is due to the location of the syrinx, which is located in the central part and disrupts the spinothalamic pathway, which carries a sensation of pain and temperature, and does not interfere with the posterior column, which carries light touch sensation, vibration, and position.[4] Syringomyelia is a cystic sac filled with cerebrospinal fluid located within the spinal cord. Syringomyelia is a common complication of Arnold Chiari Malformations Type-1.[4]

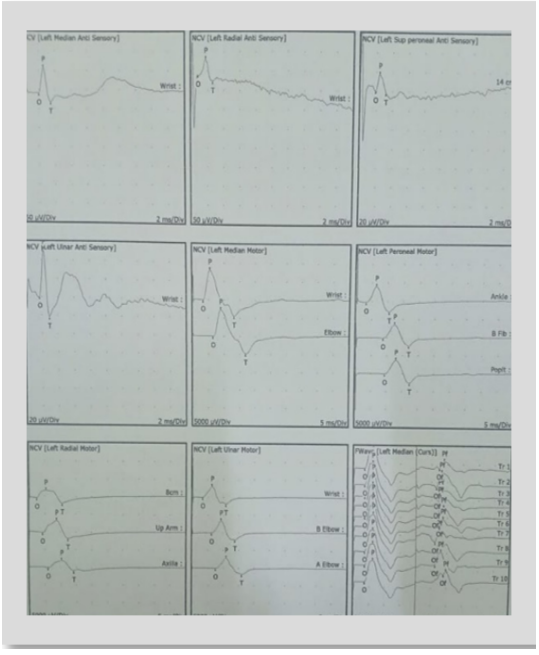

Figure 1 **NCV Studies & F-wave Show Normal Results.**

Figure 2 **C-Spine MRI without contrast shows an irregular hypointense lesion in T1Wi and hyperintense in T2Wi suggested Intramedullary from the C2 - T1 level. Cerebellar tonsillar that herniated inferiorly through the foramen magnum as far as 1.1cm from the basion episthion line.**

Gardner's hydrodynamic theory often explains the formation of a syrinx. Gardner points out that the abnormal development of the fourth ventricular outlet causes excessive distention of the neural tube in the embryonic period, which will encourage cerebrospinal fluid to flow along the path of least resistance, flowing into the central canal or parallel tract fibers.[9] Instead, obstruction of the foramen magnum interferes with the relationship between intracranial and spinal subarachnoid spaces, limiting cerebrospinal fluid compliance to oscillations if the intracranial space is under high pressure.[9]

In other words, cerebrospinal fluid from the fourth ventricle forms a syrinx in the spinal cord due to arterial pulsation or pressure dissociation between the intracranial subarachnoid space and the spinal cord. Another theory explains that the cerebrospinal fluid that comes directly from the subarachnoid space and forms a syrinx directly affects anatomic abnormalities in the cerebellar tonsil.[10] There is also a theory that explains that the liquid in syrinx comes from the accumulation of extracellular fluid. The pathomechanism of the syrinx formation is believed to be suitable with the theory explained by Gardner.[9,10]

The patient was consulted to the Neurosurgery department for posterior fossa decompression. However, patients and families refused to do the surgery. In symptomatic patients who do not undergo surgery, headaches can increase in intensity and frequency. The sensory disturbance can get heavier as the size of the syrinx increases in the spinal cord.[5,6]

Conclusion

Arnold Chiari Malformation Type 1 is a disease that has varied and non-specific clinical symptoms so that the diagnosis can resemble other diseases. In-depth history and investigations are needed to make a diagnosis of this disease. It must always be remembered that central neurological processes can mimic peripheral neurological diseases, so the possibility of a central neurological diagnosis must be considered if the patient presents with general and non-specific clinical manifestations and resembles peripheral neurological disease but

does not show improvement in symptoms after being given therapy for some time.

Aknowledgment

None.

Conflict of Interest

The authors affirm no conflict of interest in this study.

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