Case Report

Bagcilar Medical Bulletin 2017;2(4):104-107
DOI: 10.5350/BMB20171216055549

Isolated Fourth Ventricle Associated with Treated Hydrocephalus and Chiari type 2 Malformation

Murat Karacan[1], Feyza Karagoz Guzey[1], Ozgur Yusuf Aktas[1], Mustafa Safi Vatansever[1], Azmi Tufan[1], Burak Eren[1], Ilker Gulec[1], Ebru Doruk[1]

[1]Health Sciences University, Bagcilar Training and Research Hospital, Neurosurgery Clinic, Istanbul - Turkey
[2]Health Sciences University, Kanuni Sultan Suleyman Training and Research Hospital, Beylikduzu State Hospital, Neurosurgery Clinic, Istanbul - Turkey

Corresponding author:
Feyza Karagoz Guzey,
Health Sciences University, Bagcilar Training and Research Hospital, Neurosurgery Clinic, Istanbul - Turkey
E-mail address: fkarag@yahoo.com

Date of submission: December 16, 2017
Date of acceptance: December 19, 2017

ABSTRACT

Isolated fourth ventricle is a rare condition seen as a result of anatomical or functional occlusion of the inlet and outlet holes of the fourth ventricle. It may be seen in cases with Chiari type 2 malformation, too. We report a 20-month-old boy with isolated fourth ventricle presenting with weakness in his arms. He had been treated for myelomeningocele and hydrocephalus in the neonatal period and followed for Chiari type 2 malformation. The isolated fourth ventricle developed in the presence of a well-functioning lateral ventricle shunt. He was treated with a new shunt into the fourth ventricle.
Keywords: Chiari malformation type 2, hydrocephalus, isolated fourth ventricle

ÖZET

Tedavi edilmiş hidrosefaliye ve Chiari tip II malformasyonuna eşlik eden izole dördüncü ventrikül

İzole dördüncü ventrikül, dördüncü ventrikülün giriş ve çıkış deliklerinin anatomik veya fonksiyonel tıkanması sonucu ortaya çıkan nadir bir durumdur. Chiari tip 2 malformasyonlu olgularda da görülebilir. Kollarda güçsüzlükle belirti veren izole dördüncü ventrikül saptanan 20 aylık bir erkek çocuk sunuldu. Hasta yenidoğan döneminde meningomiyelosel ve hidrosefali nedeniyle tedavi edilmiş ve sonrasında Chiari tip 2 malformasyonu nedeniyle izlenmişti. Olguda izole dördüncü ventrikül iyi çalışan bir yan ventrikül şantı varlığında gelişmişti. Olgu dördüncü ventriküle yeni bir şant takılarak tedavi edildi.
Anahtar kelimeler: Chiari tip 2 malformasyonu, idrosefali, izole dördüncü ventrikül

Introduction

Isolated fourth ventricle is caused by various factors that lead to an obstruction in the entry and exit holes of the fourth ventricle such as meningeal infection, intraventricular hemorrhage, and ventriculitis (1). This rare condition can also be seen in patients with Chiari type 2 malformation. It can emerge as a result of shunt surgery (2). Cranial computerized tomography (CT) and magnetic resonance imaging (MRI) examinations are used in its diagnosis. In the cranial CT, it is seen as an isolated large round dilatation of the fourth ventricle. In addition to this finding, compression of the brain stem, ascending tentorial herniation, downward displacement of the occipital lobe, and septum formation can be seen on MRI. Clinical findings may be on a wide scale from nausea and vomiting to coma and even paraplegia/tetraplegia. Classical ventricular shunt surgery or endoscopic procedures can be used in its treatment.

Case report

A 20-month-old boy with Chiari type 2 malformation, who had undergone surgery in his neonatal period because of lumbar myelomeningocele and hydrocephalus, was admitted to our outpatient clinic with complaints of decrease in his arm movements and recurrent vomiting for a few days.

In his neurological examination, he was conscious and responsive to the environment. Cranial nerve examination revealed no abnormal findings. Increased muscle tone was noticed in the upper extremity. The patient could take objects by his arms but his spontaneous movements in the upper extremity were reduced and there was a paresis in both his arms and hands with 3/5 muscle tone. In the lower extremity, decreased muscle tone and paraplegia were seen. The child's bladder was catheterized.

On previous imaging examinations made just before and

after shunt surgery in the neonatal period, no dilatation was seen in the fourth ventricle (Figure 1a-e). However, a mild dilatation of the fourth ventricle was seen on the CT taken 4 months before the onset of weakness of his arms (Figure 2 a-b). Brain CT and MRI taken after the emerging of complaints revealed a prominent tubular-shaped dilatation of the fourth ventricle (Figure 3 a-d). Lateral ventricles were normal in size (Figure 3 b and d). It was ascertained from his medical history that the patient had undergone neither meningeal infection nor intraventricular hemorrhage before and after neonatal surgery. Further, last examination revealed no meningeal infection. Without any intervention to the old shunt device, a new shunt was placed into

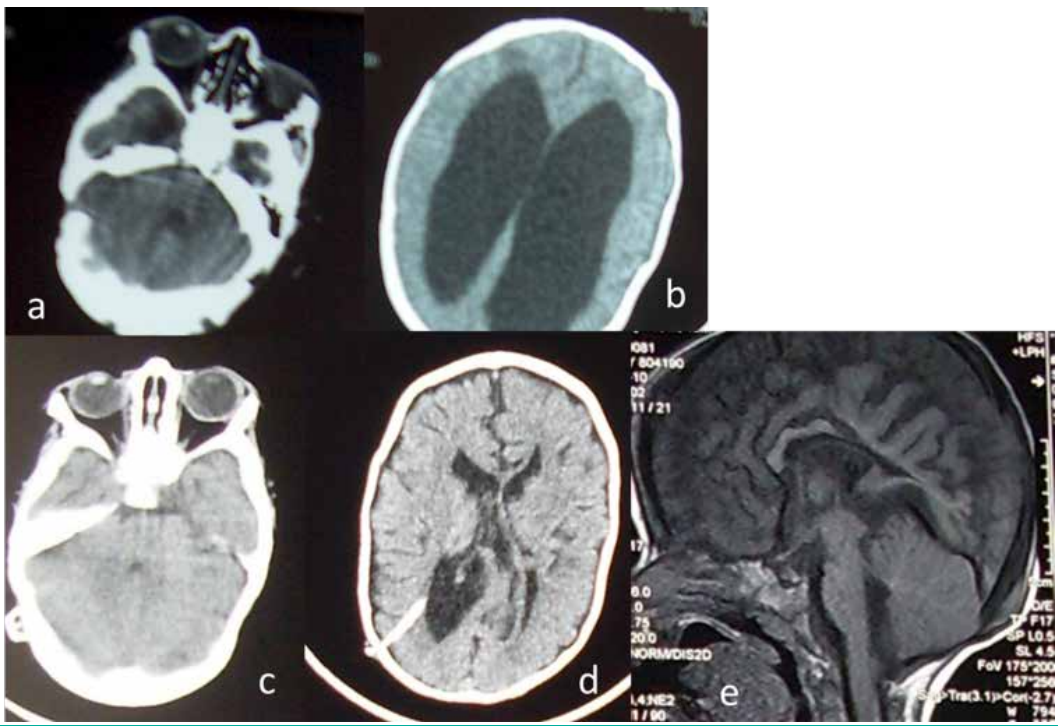

Figure 1: Brain CTs before (a and b) and after (c and d) shunt placement and midsagittal brain MRI after shunting (e) during the neonatal period showing no fourth ventricle dilatation (a, b and e), and dilated lateral ventricles before shunt surgery (b) diminished in size after shunting (d).

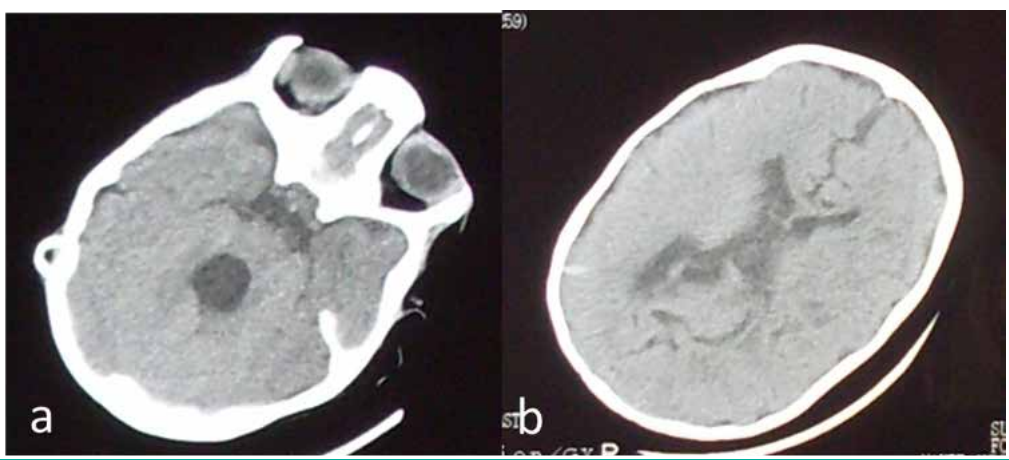

Figure 2: Brain CT showing a mild dilatation of the fourth ventricle (a) 4 months before the onset of weakness in the patient's arms. There was no dilatation of the lateral ventricles (b).

the fourth ventricle, from the opposite side of the shunt located in the lateral ventricle and it was connected to a separate peritoneal catheter. After this intervention, the patient's symptoms of muscle weakness and increased muscle tone improved and his CT examination demonstrated a decrease in fourth ventricular size (Figure 4a). After 57 months, he had no residual paresis in his arms and hands, and CT examination revealed that there was no dilatation of the fourth ventricle (Figure 4b).

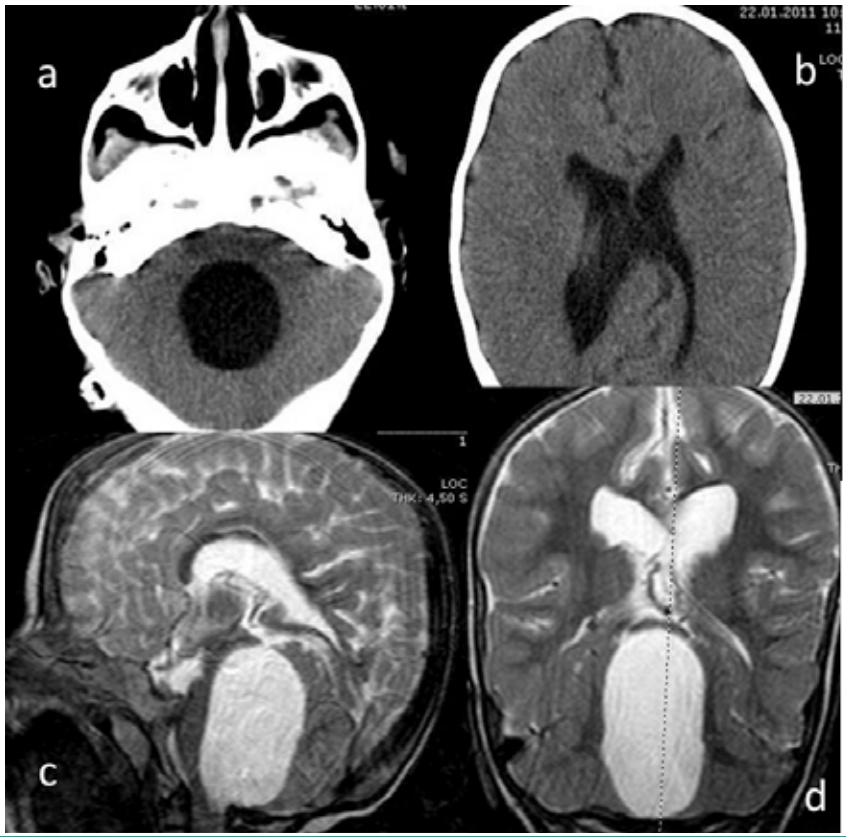

Figure 3: Brain CT (a and b) and MRI (c and d) performed after onset of the symptoms revealing a huge dilatation of the fourth ventricle (a, c and d) without dilatation of the lateral ventricles (b and d).

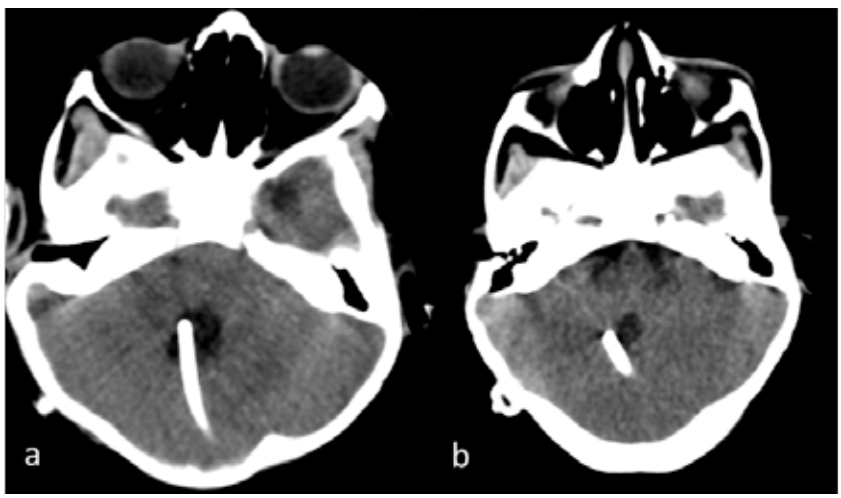

Figure 4: a) Brain CT performed after fourth ventricle shunting revealing that the size of the fourth ventricle had significantly diminished; b) Brain CT performed 57 months after fourth ventricle shunting revealing normal-sized fourth ventricle.

Discussion

Isolated fourth ventricle may occur for several reasons. Generally it is reported to be seen among patients who have subsequent complications of intraventricular hemorrhage or ventriculitis after a shunt operation in the neonatal period (3). Oi et al. (2) suggested that the isolated fourth ventricle develops as a result of rapid drainage of the cerebrospinal fluid (CSF) by the shunt system. According to their opinion, overdrainage via the shunt causes a functional blockage of the Sylvian aqueduct, and if it persists for a long time, structural and an isolated fourth ventricle will develop. Ang et al. (4) reported that the reasons for the isolated fourth ventricle are different from those of the isolated lateral ventricles. They suggested that this is caused by the occlusion of both entrance and exit holes of the fourth ventricle. In their opinion, the exit holes are usually structurally blocked due to bleeding or infection first, and subsequently structural or functional obstruction of the entrance holes develops.

According to these theories, non-structural aqueduct occlusion may occur as a result of overdrainage via the shunt system in cases with Chiari type 2 malformation where ventriculoperitoneal shunting had been performed previously (3).

However, in our case there was neither any evidence suggesting overdrainage via the shunt system nor a history of infection or bleeding. Anegawa et al. (3) presented a similar case, whose fourth ventricle initially was normal in size and gradually dilated. They suggested that this may be due to an increase in tectal beaking deformity pressing on the aqueduct. In fact, this deformity is one of the characteristics of Chiari type 2 malformation, and Adeloye showed that beaking increases with age in these patients (5). This suggestion can explain the etiopathogenesis of gradual dilatation of the fourth ventricle with a normally working shunt system as in our patient.

Different approaches have been described in the treatment of isolated fourth ventricle such as correction of overdrainage if present, shunt placement into the fourth ventricle, or opening of the exit holes of the fourth ventricle with open surgery, with endoscopic aqueductoplasty, or with endoscopic fenestration (3). Each of these methods has its own advantages and disadvantages. In our case, an endoscopic procedure was not considered because the lateral ventricles were not wide, and open surgery for fenestration was avoided due to the age of the patient. The old shunt was not removed because it was intact and functional, and as the simplest approach, a new VP shunt was placed into the fourth ventricle through the suboccipital route. Although this treatment approach seems easy, frequent complications such as multiple cranial nerve palsies, shunt dysfunction, and infection have been reported in literature (6). In our patient, no complications developed and he has shown clinical and radiological improvement for more than 57 months.

Contribution Categories	Name of Author
Follow up of the case	F.K.G., O.Y.A., M.S.V., A.T.
Literature review	M.K., F.K.G., O.Y.A., M.S.V., A.T., B.E., I.G., E.D.
Manuscript writing	M.K., F.K.G., O.Y.A., M.S.V., A.T., B.E., I.G., E.D.
Manuscript review and revisation	M.K., F.K.G., O.Y.A., M.S.V., A.T., B.E., I.G., E.D.

Conflict of Interest: Authors declared no conflict of interest.

Financial Disclosure: Authors declared no financial support.

References

1. Mohanty A, Manwaring K. Isolated fourth ventricle: To shunt or stent. Oper Neurosurg (Hagerstown) 2017; opx 136 doi:10.1093/ons/opx136. [CrossRef]
2. Oi S, Matsumoto S: Slit ventricles as a cause of isolated ventricles after shunting. Childs Nerv Syst 1985;1(4):189-193. [CrossRef]
3. Anegawa S, Hayashi T, Torigoe R, Ogasawara T. Dilated fourth ventricle in Arnold-Chiari malformation type II: Isolated fourth ventricle as sequelae of shunt? Case report. Neurol Med Chir (Tokyo) 1993;33(8):575-578. [CrossRef]
4. Ang BT, Steinbok P, Cochrane DD. Etiological differences between the isolated lateral ventricle and the isolated fourth ventricle. Childs Nerv Syst 2006;22(9):1080-1085. [CrossRef]
5. Adeloye A: Mesencephalic spur (beaking deformity of the tectum) in Arnold-Chiari malformation. J Neurosurg 1976;45(3):315-320. [CrossRef]
6. Spennato P, O'Brien DF, Fraher JP, Mallucci CL. Bilateral abducent and facial nerve palsies following fourth ventricle shunting: two case reports. Childs Nerv Syst 2005;21(4):309-316. [CrossRef]