# Multiple Hydatid Cysts of the Aqueduct of Sylvius: A Case Report With MRI Study

# Aqueductus Sylvius'da Multipl Kist Hidatik: Olgu Sunumu

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Abstract: One to two of all Echinococcus granulosus infections involve the brain, and the finding of a hydatid cyst in the aqueduct of Sylvius is extremely rare. We describe the case of a 10-year-old girl with a symptomatic hydatid cyst in the aqueduct of Sylvius. The patient exhibited signs of elevated intracranial pressure and also had left hemiparesis, ataxia and cerebellar signs. Neuroradiological imaging revealed obstructive hydrocephalus and a cystic lesion in the aqueduct of Sylvius. Magnetic resonance imaging (MRI) was used to evaluate the patient pre- and postoperatively, and surgery was carried out via a suboccipital approach using Dowling's technique. Multiple cysts were removed intact without rupture, and the patient survived and recovered fully. This is the first case with a lesion at this location to be documented by MRI, and only the second of its kind in the literature. MRI is important in terms of diagnosing and planning surgical treatment for these cysts. Total removal without rupture should be the surgical goal in all cases.

Key words: Aqueduct of Sylvius, cerebral hydatid cyst, magnetic resonance imaging

Özet: Echinococcus granulosus enfeksiyonları % 1-2 oranında beyinde hidatid hastalığı şeklinde görülür. Aquaductus Sylvius'da hidatik kist görülmesi çok enderdir. 10 yaşındaki bir kız çocuğunda aqueductus sylvius da semptomatik hidatik kist olgusu sunulmuştur. Olguda kafa içi basınç artırı bulguları, solhemiparazi, ataksi mevcuttu. Nöroradiolojik incelemelerde obstruktifhidrosefali ve aqueductus sylvius da kistik lezyon saptandı. Magnetik rezonans görüntüleme (MRG) preoperatif ve postoperatif olarak kullanıldı. Hasta oturur pozisyonda suboksipital yaklaşımla Dowling tekniği kullanılarak opere edildi. Çoklu sayıda kistler rüptüre edilmeden tamamıyla çıkarıldı. Bu olgu MRG ile dokümente edilen ilk aqueductus sylvius kist hidatik olgusudur. Literatürde ise ikinci aqueductus sylvius kist hidatik olgusudur. MRG, tanıda ve cerrahi planlamada önemli bir araçtır. Cerrahide amaç ise kisti rüptüre etmeden çıkarmaktır.

**Anahtar kelimeler:** Aqueductus sylvius, beyin, kist hidatik, magnetik rezonans görüntüleme

## INTRODUCTION

Hydatid disease in humans begins with infestation of the larval form of the canine tapeworm Echinococcus granulosus. This organism is found

worldwide, with particularly high populations where sheep are raised, and is the most common cause of the human form of the disease (1). Hydatid disease can affect almost any tissue in the body, and the most common sites of cyst development in

humans are the lungs, liver and central nervous system. Only 1-2% of all E. granulosus infections involve the brain. These cases are usually solitary cysts that arise in the brain parenchyma (1,2). Cysts have also been found in the ventricles or the posterior fossa (6), but such findings are rare.

Here we report on a patient who was diagnosed with hydatid cyst of the aqueduct of Sylvius, representing only the second such case in the literature. We also discuss the importance of magnetic resonance imaging (MRI) in this case, and how this technique helped us plan the surgical strategy.

# CASE REPORT

A 10-year-old girl was admitted to our hospital with a 1-month history of headache, nausea and vomiting. Her neurological examination revealed papilledema, left-sided palsy of cranial nerve VI, left hemiparesis, ataxia, cerebellar signs and hyperactive muscle stretch reflexes of the lower extremities. The circumference of the patient's head was 60 cm. Plain radiographs of the skull showed signs of intracranial hypertension. MRI demonstrated triventricular hydrocephalus together with enlargement of the aqueduct of Sylvius (Figure 1). T1-weighted images demonstrated multiple hypointense cystic lesions in the aqueduct of Sylvius. A subsequent whole-body radiological investigation revealed multiple hydatid

Figure 1: Preoperative sagittal and axial T1-weighted MR images show multiple cysts in the aqueduct of Sylvius, and enlargement of the third and lateral ventricles.

cysts in the liver, pancreas and left kidney. The hemagglutination test for hydatid disease was positive.

At surgery we performed a suboccipital craniectomy with the patient in the semisitting position, and opened the dura with a Y-shape incision. After the vermian incision was made, we were able to visualize the cyst in the upper part of the fourth ventricle. Once the cyst was totally removed using Dowling's technique, we could see five more cysts in the caudal part of the aqueduct of Sylvius. All these cysts were removed intact.

The early postoperative course was uneventful and the patient developed no further neurological deficit or any seizures. She was prescribed 12-mg/kg/day albendazole for 9 months as treatment (intermittently) for systemic hydatid disease. A neurological examination at routine follow-up 2 months postsurgery revealed only mild hemiparesis on the left side, and MRI results were normal (Figure 2). At this stage, the patient was transferred to the general surgery clinic to be treated for abdominal cysts.

### DISCUSSION

Hydatid cysts are usually acquired in childhood and grow slowly over time. The most common presentation is that of a child or adult with signs and



Figure 2: Postoperative sagittal T1-weighted MRI confirms total cyst removal, and shows frontal pneumocephalus and a residual cavity filled with cerebrospinal fluid.

symptoms of increased intracranial pressure. Clinically, patients tend to have problems of headache, vomiting, hemiparesis, papilledema, ataxia and seizures. Ventricular hydatidosis usually causes obstructive hydrocephalus (4,6).

The literature contains only one other case of a hydatid cyst in the aqueduct of Sylvius (5). The authors discussed the appearance of the lesion on computed tomography (CT), and reported that they had not been able to remove the cyst without rupture. Our case involved multiple hydatid cysts that arose in the aqueduct of Sylvius and grew into the fourth ventricle. This is the first case in which MRI has been used to document cysts originating at this location.

MRI is the procedure of choice for diagnosing hydatid cysts. The signal from the cyst is nonhomogeneous and of medium-to-low intensity on T1-weighted images, and of high intensity on T2-weighted images. MRI appears to be more useful and reliable than CT with regard to defining the nature of these cysts.

The pericystic region shows contrast enhancement after Gd-DTPA injection. The sensitivity of MRI, in addition to its multiplanar imaging capabilities, allows for accurate detection and assessment of affected structures in cases of cerebral hydatid cysts. In summary, MRI is of diagnosis in hydatid disease, and has also proven extremely beneficial with regard to surgical planning (7).

The treatment of choice for cerebral hydatid cyst is total surgical removal, and this can be achieved using Dowling's technique. We noted several important features of this method, as follows: 1) the patient's semisitting position helps the surgeon approach and manipulate the cyst(s) with greater ease; 2) a large bone flap is necessary; 3) monopolar

coagulation should not be used; 4) the cyst can best be eased out of the site by lowering the head end of the operating table and instilling warm saline solution between the cyst membrane and the surrounding neural tissue.

Preoperative diagnosis is key in terms of planning the operation and taking adequate measures to prevent rupture of daughter cysts at surgery. When a cystic lesion is detected on a CT scan, MRI should always be done as well because the latter is superior for diagnosing and characterizing hydatid lesions. Finally, in countries where this disease is endemic, hydatid cysts should always be included in the differential diagnosis for cystic masses.

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