



CASE REPORT

COLLOID CYST WITH SEPTUM PELLUCIDUM AGENESIS: A CASE REPORT AND REVIEW OF THE LITERATURE

Deniz Konya¹, Arzu Gerçek², Serdar Özgen¹, M. Necmettin Pamir¹

¹Department of Neurosurgery, School of Medicine, Marmara University, Istanbul, Turkey ²Department of Anesthesiology and Reanimation, Institute of Neurological Sciences, Marmara University, Istanbul, Turkey

ABSTRACT

A 37-year-old man was admitted to our clinic with sudden onset of severe headache and vomiting. Fundoscopic ophthalmic examination revealed papiledema as a sign of increased intracranial pressure. Cranial magnetic resonance imaging revealed homogenous gadolinium, enhanced cystic lesion in the third ventricle and agenesis of the septum pellucidum. Transcallosal interhemispheric approach was performed and the cystic lesion was excised totally. Pathological diagnosis was a colloid cyst. After one-day in intensive care, the patient was discharged on postoperative day 4. His neurological examination was normal after a six-month follow-up. This report represents the second report of colloid cyst and septal agenesis, and a first report of treatment of the colloid cyst with this syndrome.

Keywords: Colloid cysts, Septum pellucidum agenesis, Treatment modality

SEPTUM PELLUCIDUM AGENEZİNİN EŞLİK ETTİĞİ KOLLOİD KİST: VAKA SUNUMU VE LİTERATÜR TARAMASI

ÖZET

37 yaşında erkek hasta ani başlayan şiddetli baş ağrısı ve kusma şikayetleri ile kliniğimize başvurdu. Gözdibi muayenesinde intrakraniyal basınç artışının bir bulgusu olarak papilödem saptandı. Kraniyal magnetik rezonans görüntülemeye 3. ventrikülde, gadolinyum tutan kistik lezyon ve septum agenezi saptandı. Transkollazal interhemisferik yaklaşımla kist tamamen çıkarıldı. Patoloji olarak kolloid kist tanısı kondu. Yoğun bakımda bir gün takip edilen hasta, postoperatif 4 günde taburcu edildi. Altıncı ayda yapılan kontrolünde nörolojik muayenesi normal sınırlarda bulundu. Bu vaka literatürdeki 2. kolloid kist ve septal agenezis vakası ve ilk intrakraniyal basınç artışı ile bulgu veren vaka sunumudur.

Anahtar Kelimeler: Kolloid kist, Septum pellucidum agenezi, Tedavi yaklaşımı

INTRODUCTION

Colloid cysts (CC), having a fibrous epithelial shaped capsule and insert mucous or hyaloids substance, account for 0.3-2 % of intracranial tumors¹. They are benign tumors that can be surgically cured. They are mostly located in the anterosuperior portion of the 3rd ventricle and rarely, in the septum pellucidum¹. They may block the outflow pathways, especially at foramen Monro, for the cerebrospinal fluid, causing acute hydrocephalus². If untreated,

they may lead to sudden neurological deterioration and death.

Septum pellucidum (SP) separates the left and right lateral ventricle from each other and is made up of two thin sheets of mostly glial-like elements that have a potential space between them³. It consists of an ependymal lining toward the ventricles and contains neuronal and glial cell elements. These cell elements have connections to the hypothalamus and the hippocampus. Rakic

Corresponding author:

.Deniz Konya, M.D.

Department of Neurology, School of Medicine, Marmara University,
Altunizade, Istanbul, Turkey

e-mail: drdkonya@hotmail.com

Marmara Medical Journal 2007;20(2);110-113



and Yakovlev⁴ suggested that the layers of septum form as a result of cavitation of the medial inferior commissural plate during the formation of corpus callosum. The two layers of SP are separate at birth. In most people, they typically start fusing posteriorly. In case of absence of the SP, the frontal horns if not distorted by a cleft, have a square appearance⁵.

SP agenesis may be primary or secondary to hydrocephalus. In cases with other accompanying neurological disorders, SP agenesis can be detected in childhood. This report represents the second report of CC and septal agenesis, and a first report of treatment of CC with this syndrome.

CASE REPORT

A 37-year-old man presented with sudden onset of severe headache and vomiting. His consciousness progressed to stupor. Fundoscopic ophthalmic examination revealed papilledema as a sign of increased intracranial pressure. Cranial magnetic resonance imaging (MRI) revealed a homogenous, gadolinium enhanced cystic lesion in the third ventricle and agenesis of the septum pellucidum (Fig.1). The results of laboratory studies were within reference values.

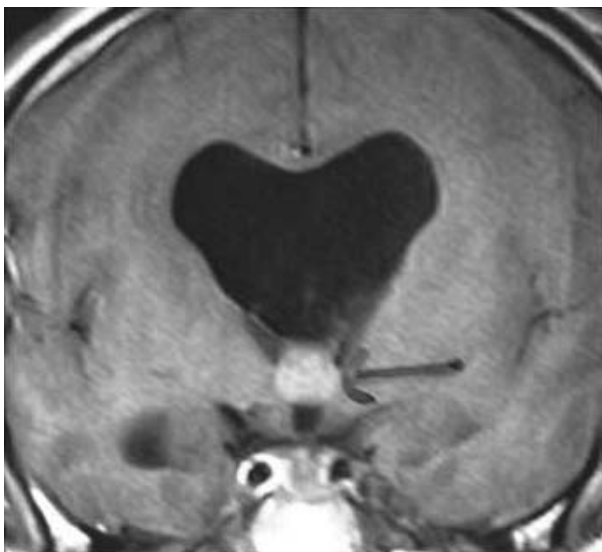


Figure 1: Preoperative T1 weighted coronal cranial MRI demonstrates homogeneous contrast enhanced colloid cyst (black arrows) ventricular enlargement and septum pellucidum agenesis.

Craniotomy was scheduled. Transcallosal interhemispheric approach was performed, and the tumor was excised totally (Fig. 2). Pathological diagnosis was CC. After one day in intensive care, the patient was discharged on postoperative day 4.

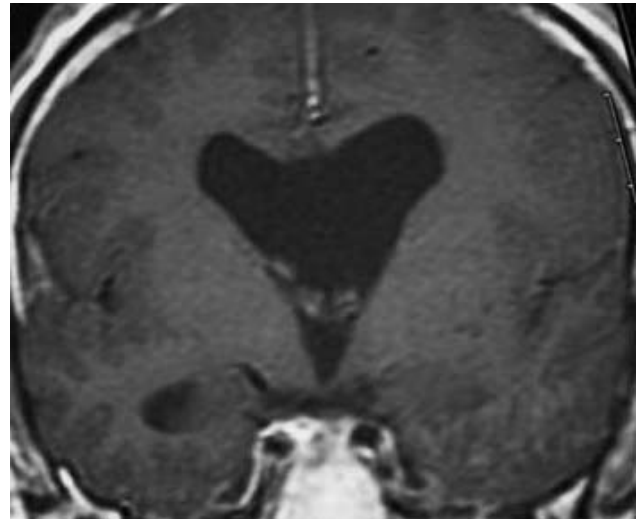


Figure 2: Postoperative T1 weighted coronal cranial MRI. Colloid cyst was excised totally.

DISCUSSION

The human brain is divided into two hemispheres by the septum or septum verum. The septum verum is a combination of nerve cells and the SP⁶. The evolution of both the SP and the corpus callosum is the same. However, SP anomalies may accompany corpus callosum anomalies. The well-known anomalies of the SP are cavum SP, cavum verge cysts and agenesis of the SP. Septo-optic displasia, schizencephaly, corpus callosum agenesis, hydrocephalus, porencephaly, basal encephalosele and hydrancephaly are usually associated with SP anomalies⁷. The SP is a part of the limbic system. Thus, limbic system anomalies may also be accompanied by SP anomalies. Schizophrenia is frequently seen with SP agenesis because of the relation with the limbic system⁶. Our patient's psychological status was previously normal. Congenital hemiplegia or diplegia is another entity that may occur in SP agenesis and porencephaly.



Barkovich and Norman⁸ reviewed 35 patients with SP agenesis. These patients were divided into seven groups: septo-optic dysplasia, schizencephaly, holoprosencephaly, agenesis of the corpus callosum, chronic-severe hydrocephalus, basilar encephaloceles, and porencephaly/hydrancephaly. They concluded that the absence of SP could provide a valuable clue to the diagnosis of malformations of the brain. Sargon et al⁹ have reported 5-layer durameter and agenesis of SP and corpus callosum in a 55-year-old man postmortem. Menezes et al⁷ studied 12 patients with absence of the SP associated with porencephalies. Literature reveals only one report of SP agenesis without additional malformation¹⁰. In this case, there were no other brain anomalies except SP agenesis.

A colloid cyst located in the third ventricle was first described by Wallman¹¹ in 1858 postmortem of a patient having incontinence and gait disturbance. Developmental midline cysts constitute a separate entity and are usually associated with malformation disorders of the brain. Embryologic origins of the colloid cysts are still unclear. Recently, immunohistochemical and ultrastructural trials convinced scientists that the colloid cyst has endodermal structure. Ectopic endodermal elements are supposed to migrate into the velum interpositum during central nervous system development¹². The cyst contains collagen and fibroblast and is surrounded by a simple or pseudo capsule. The highly vascular collagen capsule contains hemosiderin and calcium. Colloid cysts are usually gelatinous, viscid, or semisolid and rarely contain liquid¹³. In this case, the cyst was mucous.

Del Carpio et al¹⁴ reported a colloid cyst of the third ventricle with agenesis of the corpus callosum. They concluded that a colloid cyst, generally a tumor with an origin of maldevelopment, was the initiating cause of callosal agenesis. Its location, anteroinferior to the foramen Monro, coincides with that of the first callosal fibers and if present during the critical period of embryogenesis, may preclude the formation of the CC. Kantarci et al¹⁵ described a 22-year-old man with

Gorlin's syndrome having unusual findings of thin corpus callosum and colloid cyst.

A colloid cyst is easily detected by cranial MRI and Computed tomography because of its localization, shape, or structure. Contrast enhancement is rarely seen on computed tomography, and the cyst is usually isodense or hyperdense, less frequently hypodense with cerebrospinal fluid. The density of a colloid cyst is dependent on the increases in viscosity¹⁶. The density of the cyst is important in planning the surgery. Stereotactic approach or endoscopic procedures should not be the choice for highly viscous cysts. A colloid cyst may present hypointensity, isointensity or hyperintensity in T1 sequence MRI. These appearances of colloid cysts depend on the amount of blood, hemosiderin, cholesterol crystals, cerebrospinal fluid, and paramagnetic ions in the cysts¹⁶. The most common appearance is a mass that is hyperintense on T1 and hypointense on T2 weighted studies.

There are several considerations for the treatment of CC. Ventriculoperitoneal shunting, stereotactic cyst aspiration, neuroendoscopic surgery, transcortical-transcallosal approach, and anterior transcallosal microsurgery are all treatment procedures for the colloid cyst^{1,13,16-21}. The posterior transcallosal approach was first used for the colloid cyst by Dandy²² in 1921. Camacho¹⁸ recommends no intervention in the absence of hydrocephalus and a cyst size of less than 15 mm diameter, which is opposed by many authors because of the risk of sudden death^{1,2,16,23}. In our case, total excision was achieved by anterior transcallosal approach without neurological deficit.

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