

Intraventricular Cavernoma

İntraventriküler Kavernom

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Abstract

Cerebral cavernomas are simple vascular malformations that are mostly located in the brain parenchyma and usually remain asymptomatic. With the incidence of 0.5%, they are the most commonly identified cerebral vascular malformations in general population. However, with only 138 reported cases to date, intraventricular cavernomas are exceptionally rare. They may either remain asymptomatic or cause serious clinical conditions depending on their size and location. Sectional imaging methods are essential in their diagnosis. And in symptomatic cases, complete surgical removal is the treatment of choice. Here, we present a 67-year-old lady with an intraventricular cavernoma in the third ventricle of whom the symptoms were mild and ambiguous and therefore we preferred close clinical and radiological follow-up instead of intervening.

Key Words: Intraventricular Cavernoma, Third Ventricle, Magnetic Resonance Imaging

Öz

Serebral kavernomlar, pek çoğu beyin parankiminde yer alan ve sıklıkla asemptomatik kalan basit vasküler malformasyonlardır. Sıklıkları %0,5 olup genel popülasyonda en sık rastlanan serebral vasküler malformasyonlardır. Bununla birlikte, intraventriküler kavernomlar, bugüne dek sadece 138 olgu bildiri ile istisnai derecede nadir lezyonlardır. Asemptomatik olabilecekleri gibi, boyut ve lokalizasyonlarına bağlı olarak ciddi klinik tablolara da yol açabilirler. Tanıda, kesitsel inceleme metotları esastır. Ve semptomatik olgularda tedavi yöntemi, cerrahi olarak çıkarılmalarıdır. Burada, 67 yaşındaki bir bayanda, semptomları hafif ve belirsiz olan ve bu nedenle girişim yapmak yerine yakın klinik ve radyolojik takibi tercih ettiğimiz, üçüncü ventrikül yerleşimli bir intraventriküler kavernom olgusunu sunuyoruz.

Anahtar Kelimeler: İntraventriküler Kavernom, Üçüncü Ventrikül, Manyetik Rezonans Görüntüleme

Introduction

Cavernomas, also referred to as cavernous malformations, cavernous hemangiomas or cavernous angiomas, are simple vascular malformations consisting of thin-walled, ectatic, elastic fiber-free capillaries. While most of the lesions are considered as congenital, cases of cavernoma developed following brain surgery or radiotherapy have also been reported (1). They are usually small lesions measuring 2 mm to a few centimeters and often remain asymptomatic. However, they are also likely to produce symptoms that vary depending on their localization and size. With the incidence of 0.5%, cavernomas are the most commonly identified cerebral vascular malformations in general population. But, accounting for 2.5-10.8% of all cerebral

cavernomas, intraventricular cavernomas (IVC) are exceptionally rare lesions (2,3). Only 138 cases of IVC have been reported to date (1-5). IVCs may be detected at any age with a slight female predilection. And according to the previously reported cases, the lateral ventricles are the most common location of IVCs, followed by the third and fourth ventricles, respectively (5). Here, we present a 67-year-old lady with an IVC in the third ventricle.

Case Report

A 67-year-old female patient with a 14-year history of hypertension and hypercholesterolaemia, which were both under good control, presented with the complaints of headache,

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memory impairment and clumsiness over the last few years. Neurological examination findings were unremarkable. Magnetic resonance imaging (MRI) revealed a well-defined lobulated mass in the interpeduncular cistern measuring 15x14x12 mm (AP x TR x CC) which was protruding into the third ventricle (Figure 1). The mass showed heterogeneous low signal intensity in T1-weighted and high signal intensity in T2-weighted and fluid attenuation inversion recovery (FLAIR) sequences. In T1-weighted, T2-weighted, and FLAIR sequences, there were bright hyperintensities within the lesion. And a peripheral hypointense rim surrounding the mass was evident in all three sequences (Figure 2). A mild heterogeneous enhancement of the lesion was noted following intravenous gadolinium administration (Figure 3). Apart from the mass, the patient's MRI showed numerous atrophic and chronic ischemic changes throughout the brain parenchyma.

Based on the rather specific imaging findings, the patient was diagnosed as having an IVC. However, her complaints were attributed to the nonspecific atrophic and chronic ischemic

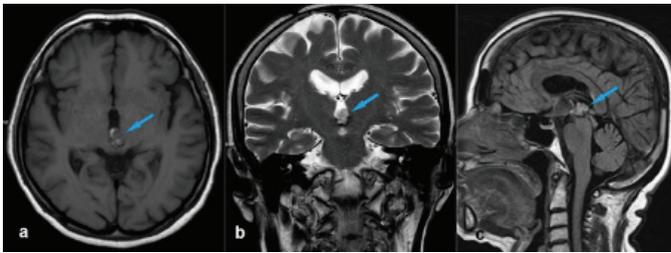


Figure 1: Axial T1-weighted (a), coronal T2-weighted (b) and sagittal FLAIR (c) images demonstrating a well-defined lobulated mass in the interpeduncular cistern (arrows). Note the lesion protrudes into the third ventricle (c)

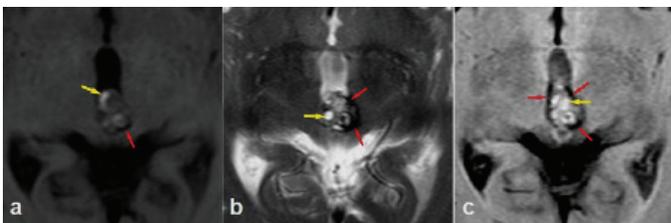


Figure 2: Axial T1-weighted (a), T2-weighted (b) and FLAIR (c) images showing a mass with intralesional bright hyperintensities (yellow arrows) and a perilesional hypointense rim (red arrows)

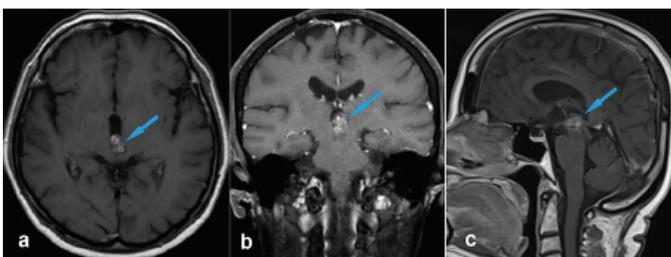


Figure 3: Post-contrast axial (a), coronal (b) and sagittal (c) images show that the lesion shows mild heterogeneous enhancement following gadolinium administration (arrows)

changes, not to the IVC. So, no intervention was performed. But the patient was called for a third month MRI follow-up for IVC.

Discussion

According to the previously reported cases, the mean maximum diameter of IVCs is about 2.6 cm and the larger ones are usually located in the lateral ventricles (5). Shirvani et al. (5) have pointed out that as a result of the absence of surrounding brain tissue as a barrier factor and frequent intralesional bleeding, IVCs grow more quickly compared to their parenchymal counterparts. While small IVCs usually remain asymptomatic, larger ones may cause serious clinical conditions depending on their location. It has been shown that the most common presentation of IVCs is the mass effect on the adjacent brain tissue. And IVCs, most commonly those located in the third ventricle, may cause hydrocephalus. Seizures, hemorrhage and neurological disorders are the other possible presentations of these rare vascular malformations (1-5). We presented a case of a relatively small IVC that was located in the posterior aspect of the third ventricle. The calibration of the ventricular system was within normal limits. The symptoms of the patient were rather mild and ambiguous. So, we attributed these long-standing unclear symptoms to the atrophic and chronic ischemic changes in the brain and therefore we preferred close clinical and radiological follow-up instead of intervening.

Sectional imaging methods are essential in the diagnosis of cerebral cavernomas. The parenchymal and intraventricular cavernomas show similar computed tomography (CT) and MRI characteristics. On CT, they appear as hyperdense masses with possible scattered calcifications. And they typically appear as popcorn-like lesions on MRI. Hyperintense foci which are usually evident on T1-weighted images represent methemoglobin. On T2-weighted images, a hypointense peripheral rim as the result of the paramagnetic effect of hemosiderin is usually present. This paramagnetic effect of hemosiderin can also be demonstrated as a marked low-signal area on T2-weighted gradient echo sequences in equivocal cases. Cavernomas usually show no or mild enhancement following intravenous gadolinium administration (6). Other intraventricular masses such as central neurocytoma, subependymoma, meningioma, subependymal giant cell astrocytoma and metastasis should be taken into consideration as the imaging differentials of IVC. In our case, typical popcorn-like appearance along with the intralesional hyperintense foci and hypointense rim were present. So, we easily established the diagnosis on routine brain imaging sequences.

Complete surgical removal is the treatment of choice in symptomatic cases of IVC (7,8). In recent years, endoscopic surgical approach to these cases has increasingly been used

(1,3,5,7). Endoscopic resection is recommended for IVCs located in the lateral ventricles, the interventricular foramina and to some extent the third ventricle (1). In our case, we preferred to follow the lesion without any intervention. However, the lesion in our case is located in a critical location for hydrocephalus. Considering the possibility of a rapid change in the clinical picture due to the increase in the mass volume due to bleeding in the following period, we planned close monitoring of the patient.

Conclusion

While cavernomas are the most commonly identified cerebral vascular malformations in general population, their intraventricular forms are extremely rare. IVCs should always be taken into consideration in the imaging differentials of an intraventricular mass.

Ethics

Informed Consent: Informed consent for publication was obtained from the patient.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.Ö., A.D., Concept: A.D., Design: M.Ö., Data Collection or Processing: A.D., R.P.K., T.A., E.S.G., Analysis or Interpretation: R.P.K., T.A., E.S.G., Literature Search: M.Ö., Writing: M.Ö.

Conflict of Interest: The authors declare that they have no conflict of interest.

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