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Primary Fourth Ventricular Meningioma: Case Report and Review of the Literature

Primer 4. Ventrikül Meninjiomu: Olgu Sunumu ve Literatür Derlemesi

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ABSTRACT

Primary fourth ventricle meningiomas are extremely rare, and they are defined as meningiomas arising from the choroid plexus and lying strictly within the fourth ventricle. In this report we present a 61-year-old man with progressive worsening vertigo and gait disturbance, and new onset of diplopia. Neurological examination revealed bilateral abducens nerve paralysis, horizontal nystagmus, and gait disturbance with truncal ataxia. Neuroimaging revealed a mass lesion in the fourth ventricle with brain stem compression, and obstructive hydrocephalus. The patient was operated in the prone position with suboccipital craniotomy and splitting the lower vermis. Total resection of the tumor was achieved with no intra- or post-operative complications. Histopathologic examination revealed fibroblastic type meningioma (WHO grade I).

KEYWORDS: Fourth ventricle, Meningioma, Posterior cranial fossa, Surgical treatment

ÖZ

Primer 4. ventrikül meninjiomları nadir görülmektedir ve koroid pleksustan kaynaklanan ve 4. ventrikül içinde sınırlanan meninjiom olarak tanımlanmaktadır. Bu sunumda 61 yaşında erkek hasta ilerleyici vertigo, yürüme bozukluğu ve yeni başlayan çift görme şikayetleri ile başvurdu. Nörolojik muayenede bilateral abdüsens sinir felci, horizontal nistagmus, yürüme bozukluğu ve trunkal ataksi saptandı. Görüntülemelerde 4. ventrikül içinde yer alan ve beyin sapına ve obstrüktif hidrosefaliye neden olan kitle lezyonu saptandı. Hasta suboksipital kraniotomi ile ve alt vermis ayırma ile yaklaşarak opera edildi. Perop komplikasyon gelişmeden tümör total olarak rezeke edildi. Histopatolojik değerlendirme sonucunda fibroblastik tip meninjom (WHO grade I) saptandı.

ANAHTAR SÖZCÜKLER: Dördüncü ventrikül, Meninjiom, Posterior kranyal fossa, Cerrahi tedavi

INTRODUCTION

First described by Sachs in 1983 (9), primary fourth ventricle meningiomas are extremely rare at 6.6% of all intraventricular meningiomas and only a few cases reported in the literature describing these tumors (2,3,4,6,7,11,16,17,19). They are clearly defined as arising from the choroid plexus and lying strictly within the ventricular cavity (13,21). In this report, the authors present an additional case of a primary fourth ventricular meningioma. Radiological, clinical and pathologic data and intraoperative findings are discussed, with a brief review of similar cases in the literature.

CASE REPORT

A 61-year-old man was admitted with a 2-year history of progressively worsening vertigo and gait disturbance, with new onset of diplopia. Neurological examination revealed bilateral abducens nerve paralysis, horizontal nystagmus, and gait disturbance with truncal ataxia. The Romberg test was negative. Brain computed tomography (CT) scan showed a mass lesion in the posterior fossa obstructing the fourth ventricle, with secondary triventricular hydrocephalus. The mass lesion showed no calcifications. Cranial magnetic resonance imaging (MRI) with gadolinium enhancement revealed a mass lesion in the fourth ventricle with brain stem compression, and obstructive hydrocephalus. The lesion was round with clear margins, hypointense in T1, and iso-/hypointense in T2-weighted images, with homogenous dens contrast enhancement (Figure 1A-D). Surgical intervention was decided, and surgery was performed in the prone position. The lesion was approached by a suboccipital craniotomy, with bone removal and opening of the posterior border of foramen Magnum. Under the magnification of the operating microscope, the dura was opened, and the lower vermis was found to be edematous. After cerebrospinal fluid drainage from the cisterna magna, the lesion was exposed by splitting the lower vermis. The tumor was found to be firm, with clear cleavage planes and little vascularization, and was solely located in the fourth ventricle. Intracapsular debulging of the tumor was performed, followed by dissection of the capsule from the surrounding tissue until the exposure of the

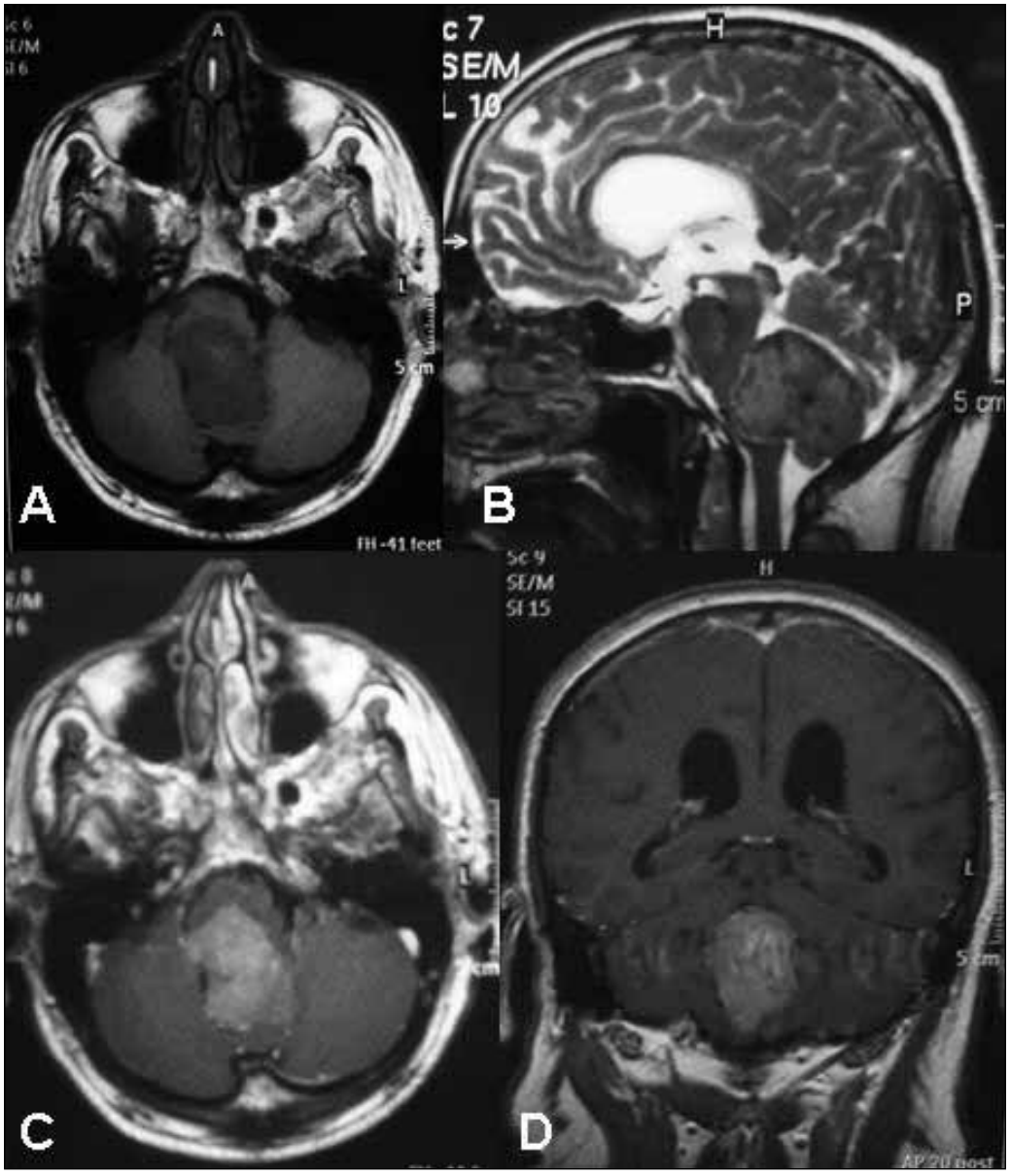

Figure 1: Pre-operative cranial MRI of the patient in our case, demonstrating a mass lesion in the fourth ventricle with brain stem compression, and obstructive hydrocephalus. **A)** axial T1-weighted, **B)** sagittal T2-weighted, **C)** axial post-contrast, and **D)** coronal post-contrast images.

rhomboid fossa. No intraoperative complications occurred, and the tumor was totally resected, with bleeding control before closure Postoperatively, the patient's complaints showed improvement with no additional neurological deficits, and the patient was discharged on postoperative day 7 without complications. Histopathological examination revealed fibroblastic type meningioma (WHO grade I). There was minimal hypercellularity, pleomorphism and pattern loss, with no signs of malignancy. There was no necrosis nor invasion of neighbouring neural parenchyma, and the mitotic activity was found to be 0-1/10 BBA. The Ki-67 proliferation index was lower than 1%. Radiological follow-up with contrast enhanced cranial MRI showed no residual tumor and decompression of the brain stem, with regression of the hydrocephalus (Figure 2A, B).

DISCUSSION

Meningiomas account for approximately 15% of all intracranial neoplasms (19). They are known to occur in various locations within the brain, originating from the arachnoid cap cells. The primary occurrence of meningiomas in the ventricular system without dural attachment is extremely rare, with an incidence of 0.5 to 3% among all intracranial meningiomas. Of these tumors, 77.8% occur in the lateral ventricular trigone (more

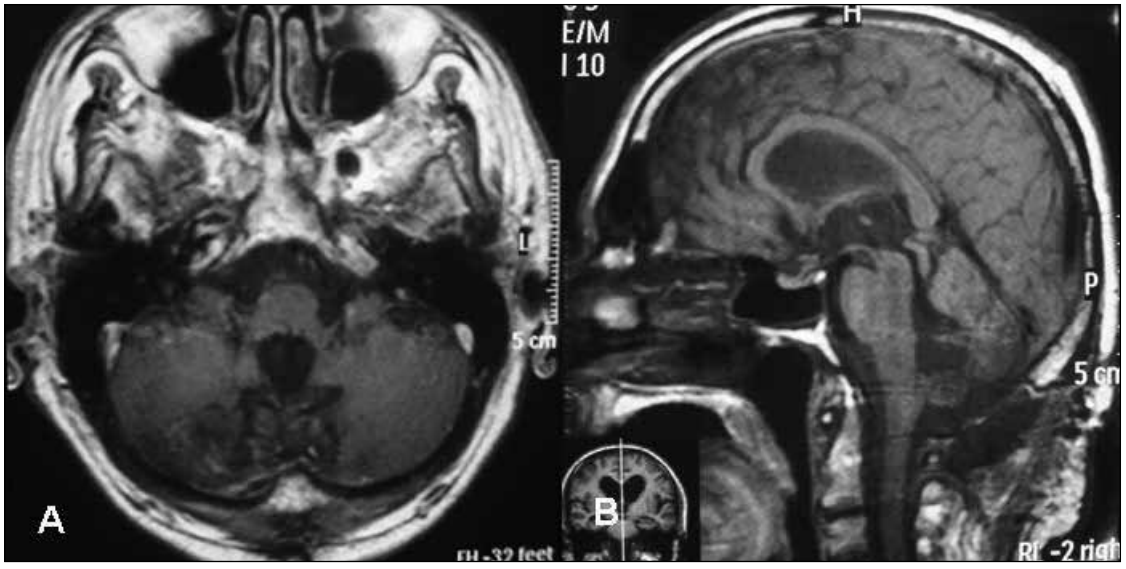

Figure 2: Post-contrast enhancement MRI demonstrating no residual tumor and decompression of the brain stem, with regression of the hydrocephalus. **A)** axial and **B)** sagittal images.

commonly on the left side for unexplained reasons), 15.6% in the third ventricle and 6.6% in the fourth ventricle (19)

First described by Sachs in 1983 (9), primary fourth ventricle meningiomas are extremely rare, with only 28 cases reported in the literature describing these tumors (2,3,4,6,7,11,16,17,19). These reports, including our case, are summarized in Table I.

Abraham et al. (1) classified posterior fossa meningiomas without dural attachment as Type I: meningiomas that arise from the choroidal plexus of the fourth ventricle and lie entirely in it, Type II: meningiomas that arise from the inferior tela choroidea and are located in both the fourth ventricle and cerebellar hemisphere, and Type III: meningiomas located in the cisterna magna. Fourth ventricle meningiomas correspond to the Type I of this classification. These tumors have slight female predominance. Reviewing the cases in the literature, including our case, 67% of patients were female and 33% were male, with sex ratio of 2:1 female to male. The average age of the patients was 45 years, ranging from 14 to 72. However, the age of the female patients was younger (mean = 36.9 years old) compared to male patients (mean = 60 years old).

Patients with fourth ventricular meningioma generally present with the clinical features of intracranial hypertension related to obstructive hydrocephalus (7,13,21), with some cases presenting with cerebellar dysfunction (4,8,15), and focal neurological deficits, such as diplopia (10,22), peripheral hemifacial nerve paralysis (12) and long tract signs (18). On the other hand, the patient in our case presented with cerebellar dysfunction and diplopia, with no significant clinical signs of hydrocephalus, although observed radiologically. Some reports described fluctuating of the symptoms due to the intermittent occlusion of the fourth ventricle in the supine position, however, this may not be the rule, as our patient demonstrated progressive worsening vertigo and gait disturbance, with acute development of diplopia.

Reviewing the histological types of primary fourth ventricle meningioma shows that they are most commonly of fibroblastic (40%), meningothelial (24%) and transitional (16%) subtypes. However, 2 cases of clear cell (6) and single cases of psammomatous (13), choroid (11) and endotheliomatous (14) subtypes have been also reported. Moreover, there is an interesting finding about the relation between the subtype of the primary fourth ventricle meningiomas and the gender of the patients. A review of the literature, including our case, revealed that of the 10 patients of fibroblastic subtype, 9 with known gender, most of the patients (5 patients) were male to female with a ratio of 55.6% to 44.4%. This finding is opposite to the general female predominance in the primary fourth ventricle meningioma series.

Surgical resection is the primary treatment modality of these tumors (7). Cushing has reported the first successful resection performed by Sachs (9). A total removal was achieved in all (including our case) but one operated patient (20). Splitting the lower vermis, the tumor was easily removed after intracapsular debulking as it is only attached to the choroid plexus. In contrast to posterior fossa meningiomas lying partially within the fourth ventricle, the total removal of purely intraventricular meningiomas can be achieved successfully with low operative risk and an excellent outcome (7).

CONCLUSION

Primary fourth ventricle meningiomas are extremely rare, arise from the choroidal plexus of the fourth ventricle and lie entirely in it. Microsurgical total removal of primary intraventricular meningiomas can be achieved successfully with low operative risk and an excellent outcome.

Table I: Summary of the Reported Cases of Primary Fourth Ventricle Meningioma

Author	Sex/Age	Pathology	Treatment
Sachs, 1938	F/38	Fibroblastic	T
Vogel and Stevenson,1950	M/65	Meningothelial	Autopsy finding
Haas and Ritter, 1954	M/41	Fibroblastic	Autopsy finding
Schaerer and Woolsey, 1960	F/42	Fibroblastic	T
Hoffman, 1970	F/44	Transitional	T
	M/61	Transitional	T
Rodriguez-Carbajal and Palacios, 1974	F/32	Meningothelial	T
	F/49	Meningothelial	ST
Gökalp et al., 1981	F/30	Psammomatous	T
Giromini et al., 1981	F/14	Endotheliomatous	T
Tsuboi et al., 1983	F/30	Fibroblastic	T
Nagata et al., 1983	F/52	Fibroblastic	T
Matsumara et al., 1988	M/62	Fibroblastic	T
Ceylan et al., 1992	M/48	Meningothelial	T
Lima-de-Freitas et al.,1994	F/32	Meningothelial	T
Iseda et al., 1997	F/47	Transitional	T
Cummings et al.,1999	M/72	Fibroblastic	T
Chaskis et al., 2000	M/72	Fibroblastic	T
Akimoto et al.,2001	F/72	Transitional	T
Carlotti et al., 2003	F/28	Clear cell	T
	F/23	Clear cell	T
Makhmudov et al., 2003	F/NA	NA	T
Bhatoe et al., 2006	NA	Fibroblastic	T
	NA	Meningothelial	T
Bertalanffy et al., 2006	NA	NA	T
Liu et al., 2006	NA	NA	T
Epsari et al.,2006	F/20	Chordoid	T
Present report	M/61	Fibroblastic	T

*(**F:** female, **M:** male, **NA:** no information available, **ST:** subtotal resection, **T:** total resection)*

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