Multiple Spinal Meningiomas: A Case Report of a Rare Entity

Çoklu Spinal Menenjiomlar: Nadir Bir Vaka Sunumu

Mazhar Müfit Ünal[1], Ayfer Aslan[2]

[1]Department of Neurosurgery, Edremit State Hospital, Balıkesir, Turkey
[2]Department of Neurosurgery, Hitit University Faculty of Medicine, Çorum, Turkey

ABSTRACT

The occurrence of multiple meningiomas (MMs) in the spinal canal is a rare clinical entity, if neurofibromatosis is absent. Only 17 cases of multiple spinal meningiomas can be found through a search on the database of pubmed. Moreover, MMs removed from the same patient usually show identical histological features. Considering the rarity of MMs with distinct histological features in the spinal compartment, we described an unusual case with two different subtypes of spinal meningiomas at entirely different levels at the same time and briefly reviewed the related literature.A 47-year-old woman presented with pain, numbness and weakness on the left arm and leg leading to the diagnosis of two intradural extramedullary spinal tumors located anterior to the spinal cord at the levels of C7 – Th2 and Th12 – L1 through magnetic resonance imaging. After successfully removal of both tumors, the histopathological examination revealed meningothelial and psammomatous meningiomas, respectively. Although multiple meningiomas in the spinal compartment is fairly uncommon, this entity should be kept in mind and we suggest screening of other regions of the spine in cases of spinal meningioma, even if without signs of neurofibromatosis.

Keywords: Case report; intradural tumor; multiple meningiomas; meningotheliomatous meningioma; psammomatous meningioma; spinal cord

Received: 05.09.2020 **Accepted:** 05.10.2020

ÖZET

Spinal kanalda çoklu menenjiomların görülmesi, nörofibromatozis yoksa çok nadir bir durumdur. Pubmed veri tabanında, sadece 17 çoklu spinal menenjiom vakası yer almaktadır ve aynı hastadan çıkartılan çoklu menenjiomlar özdeş histolojik özellikler göstermektedir. Spinal bölgede farklı histolojik özelliklerdeki çoklu menenjiomların ender görülmesini göz önünde bulundurarak, bu yazıda aynı anda farklı seviyelerde bulunan iki farklı spinal menenjiom alt tipinin görüldüğü nadir bir vakayı sunduk ve bununla ilgili literatürü kısaca gözden geçirdik. Sol kol ve bacakta ağrı, uyuşma ve güçsüzlük şikayetleriyle başvuran 47 yaşında kadın hastada, manyetik rezonans görüntüleme sonucunda C7 – T2 ve T12 – L1 seviyelerinde iki adet intradural ekstrameduller spinal kitle saptanmıştır. Her iki tümörün başarılı bir şekilde çıkartılması sonrasında, histopatolojik çalışmalar bunların sırasıyla meningotelyal ve psammomatöz tip menenjiom olduklarını göstermiştir. Spinal kompartmanda çoklu menenjiomlar oldukça nadir görülse de bu durumun akılda tutulmasını ve spinal menenjiom vakalarında, nörofibromatozis bulguları olmasa da diğer omurga seviyelerinin görüntülenmesini öneriyoruz.

Anahtar Sözcükler: Vaka sunumu; intradural tümör; çoklu menejiomlar; meningotelyal menenjiom; psammomatöz menenjiom; spinal cord

Geliş Tarihi: 05.09.2020 **Kabul Tarihi:** 05.10.2020

ORCID IDs: M.M.Ü.0000-0002-6103-8569, A.A. 0000-0002-5413-7833

Address for Correspondence / Yazışma Adresi: Ayfer Aslan, MD, Department of Neurosurgery, Hitit University Faculty of Medicine, Çorum, Turkey. E-mail: ayferaslan86@gmail.com

doi:http://dx.doi.org/10.12996/gmj.2021.100

INTRODUCTION

Meningiomas account for 20-38% of all primary intracranial tumors (1) and 30.7% of all primary intradural spinal tumors (2) with a female predominance (3–6). Spinal meningiomas can be found in 7.9% of all meningiomas (2), and they tend to be solitary (7). Multiple spinal meningiomas should give rise to thought of neurofibromatosis (8,9). Without evidence of neurofibromatosis, multiplicity of spinal meningiomas is an extremely rare condition.

In overall, the frequency of multiple meningiomas in central nervous system (for both intracranial and spinal compartments) is reported as 1 - 10% of all meningiomas (4); while, for only intracranial compartment, it is 1.7 - 12% among population (5,7,10,11), and 16% in autopsy series (12). Besides, the frequency of multiple spinal meningiomas is unclear in the literature, and our literature search revealed only 17 cases of multiple spinal meningiomas reported up to date. However, those reports mostly presented cases of multiple spinal meningiomas with identical histological subtypes at different levels (13,14). Although there are some studies reporting that the same patient could have intracranial meningiomas of different histologies and grades (4,5); to the best of our knowledge, such reports for spinal compartment is scarce in the literature and this prompt us to report our case which displayed an unusual occurrence of different histological types of multiple spinal meningiomas at separate levels.

CASE REPORT

A 47-year-old woman presented with weakness of the left arm and leg starting in a short while after the pain and numbness on the same side that existed for a couple of weeks. In neurological examination, the patient had left monoparesis of the left arm (4/5 of muscle power scale) and monoplegia of the left leg (1/5 of muscle power scale), along with a hypoesthesia below Th1 on the left side. Magnetic resonance imaging (MRI) disclosed two intradural extramedullary, homogenously contrast enhanced mass lesions located anterior to spinal cord at C7 – Th2 and Th12 – L1 levels, with the sizes of 11 x 18 x 50 mm and 11 x 13 x 18 mm, respectively (Figure 1). Accordingly, the spinal cord has been displaced backwards at both levels. The patient was operated with posterior approach for both tumors at the same stage. C7 – Th1 – Th2 and Th12 – L1 laminotomies followed by dural opening and total resection of tumors were carried out along with intraoperative spinal neuromonitoring (Figure 2). The inner layer of the dura attached to the tumor was cauterized and the dura closed by water-tight sutures at both levels. The excised laminae were reconnected to the remains of vertebrae with mini-plates and screws at both levels, establishing laminoplasty. Immunohistopathological examination with staining for S100, EMA, CEA, progesterone, p63 and Ki-67 resulted in meningotheliomatous meningioma at C7 – Th2 (Figure 3), and psammomatous meningioma at Th12 – L1 (Figure 4). Postoperative course was uneventful, as her pain and hypoesthesia were ameliorated as well as her weakness which was completely resolved on her left arm and improved to 3/5 of muscle power scale on her left leg. Afterwards, she was transferred to the Physical Therapy and Rehabilitation Unit for further assistance and her 2 years of follow-up disclosed no recurrence to date.

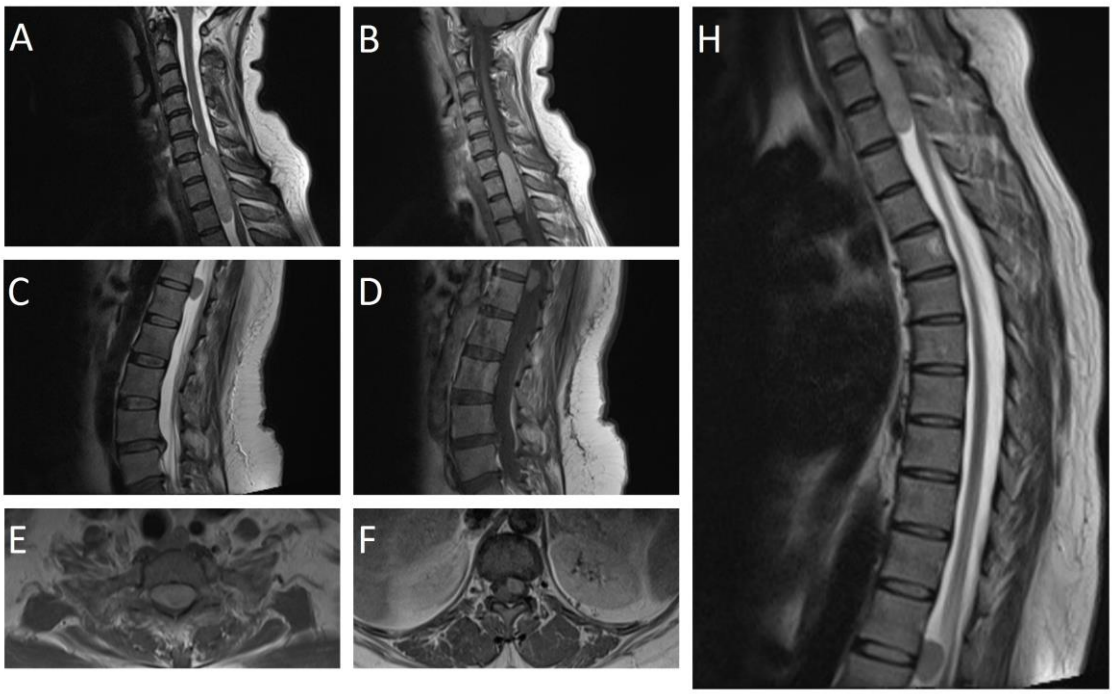

Figure 1.T2-weighted and contrast enhanced T1-weighted sagittal and axial MRI scans showed two intradural extramedullary spinal meningiomas lacated anterior to the spinal cord at C7-T2 (A, B, E and H) and T12 – L1 (C, D, F and H) levels displacing the spinal cord posteriorly at both levels.

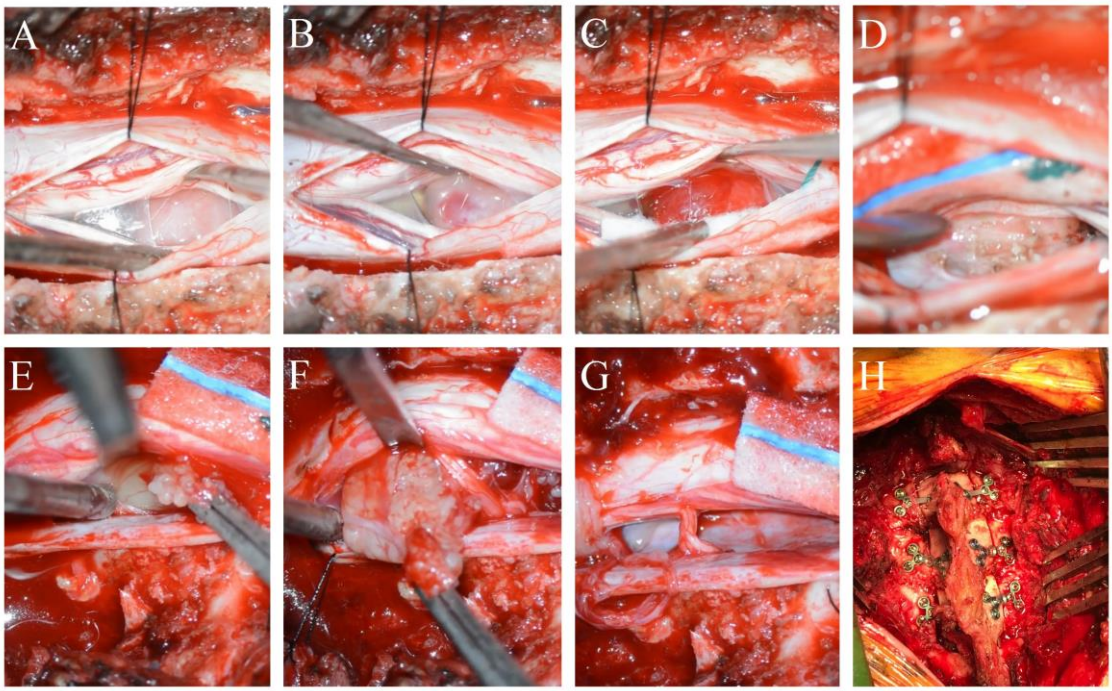

Figure 2. Exposure and removal of ventral intradural extramedullary meningiomas from Th12 – L1 (A, B, C and D) and C7 – Th2 (E, F and G) levels followed by laminoplasty (H).

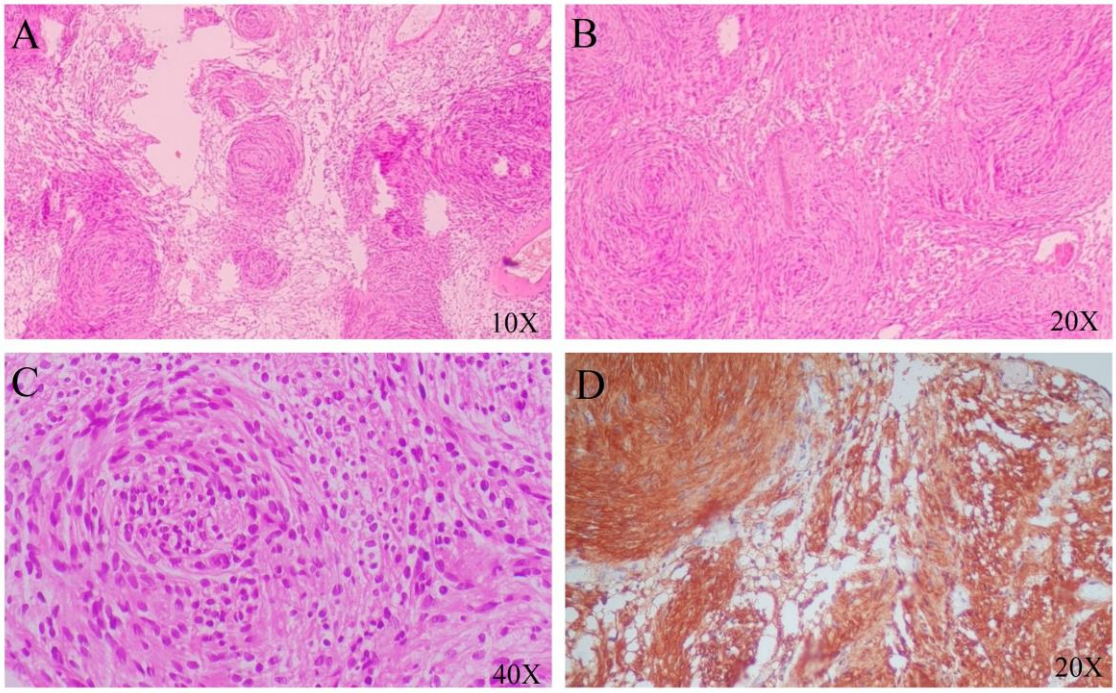

Figure 3. Histopathological examination of the tumor from C7 – Th2; meningothelial cells forming whorls stained with hematoxylin and eosin (A, B and C); and S-100 positivity (D) showing characteristics of meningotheliomatous meningioma.

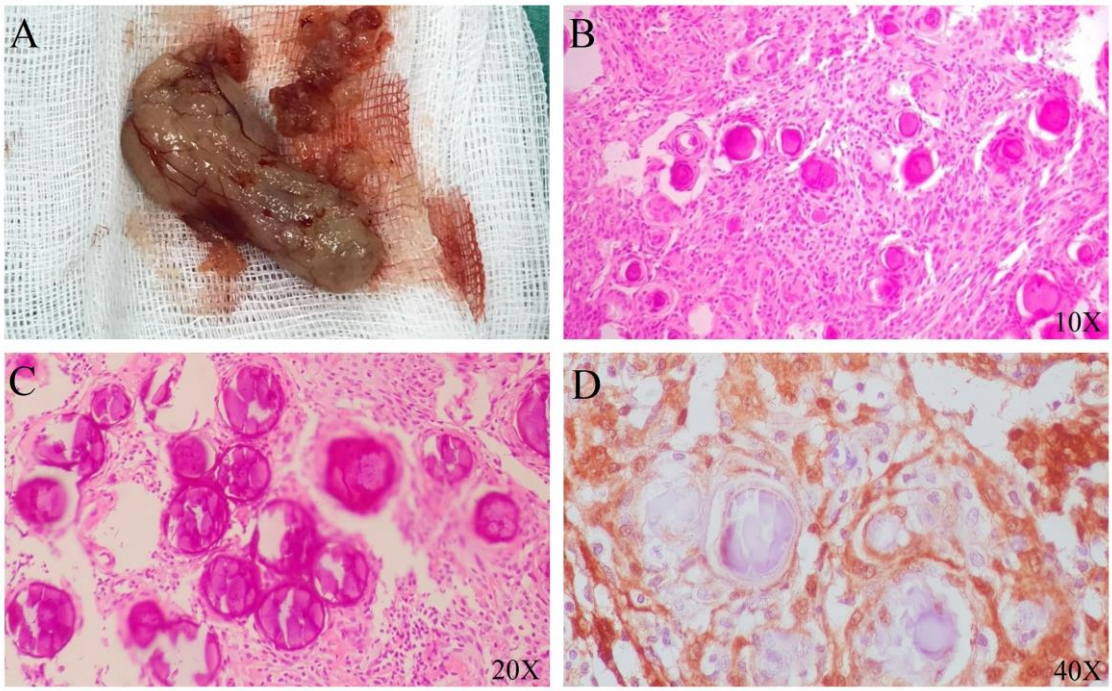

Figure 4. Histopathological examination of the tumor from Th12 – L1; macroscopically appearance of grayish solid tumor (A); numerous psammomatous calcifications among meningothelial cells stained with hematoxylin and eosin (B and C); and S-100 positivity (D) showing characteristics of psammomatous meningioma.

DISCUSSION

The occurrence of multiple spinal meningiomas without clinical signs of neurofibromatosis is extremely rare. A few cases of multiple spinal cord meningiomas in the absence of neurofibromatosis have been reported in the literature so far (Table 1).

Regrettably, exhaustive studies reporting the real incidence of spinal multiple meningiomas, except some case reports and limited reviews of literature, do not exist; therefore, current knowledge regarding frequency of multiple meningiomas is exclusively confined to the intracranial compartment. Andrioli, et al. (7) reported their case series of 934 patients operated for central nervous system (CNS) meningiomas, and found 14 cases of MMs in the intracranial compartment (an incidence of 1.5%), but none in the spinal canal. In the study of Ortaeskinazi, et al. (15), there was only one patient with multiple spinal meningiomas among total of 14 patients operated for spinal meningiomas during 10 years, between 1985 and 1995. Namer, et al. (16) found a rate of 3.5% (1 in 29 patients) for multiple spinal meningiomas among surgically treated spinal meningiomas during the years of 1970 – 1982. Nonetheless, the incidence of MMs among spinal meningiomas is claimed to be 1.6 – 2.8% in some reports (16).

There are various hypotheses regarding pathogenic mechanisms of MMs, however, the pathogenesis of this issue remains unclear in the literature and it is beyond the aim of this paper (5). The risk factors for multiplicity of meningiomas were introduced as genetic predisposition (especially for familial meningiomatosis and neurofibromatosis) and prior radiation exposure in previous studies (4).

Generally, the most frequent histological type of meningiomas is reported as meningothelial, followed by psammomatous (3). The coexisting of histologically different meningiomas at the same patient is infrequent and is mostly reported in the cranial compartment rather than the spinal canal (4,5).

Accordingly, cases of multiple spinal meningiomas reported in the literature up to date mostly illustrated concurrent meningiomas with the identical histopathological features at separate levels (13, 14, 17–24, 25). Therefore, we speculated that multiple spinal meningiomas with different histological features are an even rarer condition. The case reported in this paper had two different subtypes of meningioma (meningotheliomatous and psammomatous) at the cervicothoracic and thoracic spinal regions. Apart from our case, only 3 of 17 case reports in the literature showed multiple spinal meningiomas with distinct histological features at separate levels (26–28). Besides, due to the lack of case series on this topic, there is no evidence of the tendency of MMs removed from the same patient to have the same histological features (5).

The majority of spinal meningiomas are found intradural extramedullary, while purely extradural spinal meningiomas are extremely rare (1,9,10,29,30). The most frequent site of spinal meningiomas was reported as the thoracic region, followed by the cervical and lumbar regions (3,6,15,16,30,31). In terms of anterior-posterior location to the spinal cord, meningiomas are mostly found lateral to the cord, while the next common location is posterior, followed by anterior which is rarest and mostly found in meningiomas of cervical region (3,6,15,16,30). In our case, both cervical and thoracic tumors were anterior despite of its rareness. As ventral attachment is generally considered difficult for complete resection, the successful operation of two ventral meningiomas in our case is, no doubt, deeply gratifying.

Levy, et al. (3) emphasized the significant correlation between anterior-posterior location and spinal axis level. According to this report, while cervical meningiomas incline to site anteriorly in the canal; from C7 down, the location was twice as likely to be posterior (3). There are also some other reports pointing the inclination of spinal meningiomas to site posterior to the spinal cord at thoracic region, and anterior or lateral at cervical region (30). Apart from spinal axis level, there is no data on any other factors affecting the location of meningiomas in the literature. Whether different histological patterns of meningioma incline to arise from specific surfaces of the spinal canal is unknown and might need to be investigated in the future studies.

Table 1 The case reports of multiple spinal meningiomas without neurofibromatosis published in the literature to date

No	Study & Year	Age (yrs) & Gender	Number of spinal meningiomas	Spinal Levels	Location	Histological type
1	Rand et al., 1952[17]	34, F	2	Th3; Th5-6	Anterolateral intradural extramedullary	Psammomatous
2	Rath et al., 1966[26]	20, F	2	C3-6; Th8-10	Ventral extradural; Posterolateral intradural extramedullary	Meningotheliomatous; Psammomatous
3	Resnikoff et al., 1971[27]	37, F	2	Th8; C4	Dorsal intradural extramedullary	Psammomatous; fibroblastic
4	Di Rocco et al., 1984[18]	8, F	2	L4; L2	Posterior intradural extramedullary; anterolateral intradural extramedullary	Meningotheliomatous
5	Kandel et al., 1989[14]	17, F	2	Th6; Th8	Both dorsal intradural extramedullary	Meningotheliomatous
6	Weil et al., 1990[28]	41, M	2	C2-3	Dorsal extradural; ventral intradural extramedullary	Fibroblastic; meningotheliomatous
7	Roda et al., 1992[19]	50, F	2	Th3; Th6	Posterior intradural extramedullary; anterior intradural extramedullary	Meningotheliomatous
8	Makiuchi et al., 1993[13]	73, F	2	Th7; Th10	Intradural extramedullary	Psammomatous
9	Makiuchi et al., 1993[13]	52, M	3	Th7-Th9	Intradural extramedullary	Meningotheliomatous
10	Chaparro et al., 1993[31]	32, M	47	Th2-3; Th3-4; Th5-6; Th6-7; Th9; Th9-10; Th12; L4	Anterior, anterolateral, posterior, lateral intradural extramedullary	Grade 1
11	Lee et al., 1999[20]	77, F	2	Th9; Th12	Intradural extramedullary	Psammomatous
12	Colazza et al., 2002[21]	74, F	3	C6-7; Th6-7; Th9	Anterior; posterior	Meningotheliomatous
13	Shukla et al., 2011[22]	13, F	3	Th4-5; Th9-10; L1-2	Ventrolateral intradural extramedullary	Meningotheliomatous
14	Jain et al., 2015[23]	62, F	4	Th7-8; Th11-12; Th12-L1;	Intradural extramedullary	Psammomatous
15	Kumar et al., 2016[24]	17, M	5	C2-5; Th3-5; Th6; Th8; Th10	Intradural extramedullary	Transitional meningioma
16	Ghanchi et al., 2018[29]	40, M	2	Th6; L1	Lateral extradural	Grade 1
17	Eghbal et al., 2018[25]	57, F	4	C4-6; Th2; Th3-5; Th7	Anterior intradural extramedullary; anterolateral intradural extramedullary	Whorling-sclerosign variant meningioma
18	Current study, 2020	47, F	2	C7-Th2; Th12-L1	Anterior intradural extramedullary	Psammomatous; meningotheliomatous

CONCLUSION

Meningiomas are rarely found to be multiple, especially in the spinal canal, in the absence of neurofibromatosis. We described an unusual case of two spinal meningiomas with different histological features arising from the anterior surfaces of cervical and thoracic spinal region and successfully removed at the same operation stage. To our belief, these sporadic concurrent meningiomas should be reported to support the literature and prompt future meta-analysis investigating the frequency and etiopathogenesis of this entity.

Conflict of interest
No conflict of interest was declared by the authors.

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