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CASE REFOPT / *OLGU SUNUMU*

MULTIFOCAL MYXOPAPILLARY EPENDYMOMA OF THE FILUM TERMINALE. CASE REPORT AND REVIEW OF THE LITERATURE

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SUMMARY:

Myxopapillary ependymomas (MPE) are distinctive variant of spinal cord ependymomas, typically arise in the filum terminale and conus medullaris, accounting 40-60% of spinal cord tumors. We report a case of MPE with two discrete foci on the both ends of the filum terminale and discuss solitary and multifocal MPEs with data of other four patients with double MPEs of filum terminale founded in the literature. A 24-year-old previously healthy male presented with a two-month history of back pain radiating to his right leg. He reported no motor, sensory, bowel, or bladder dysfunction. A MRI examination of lumbosacral region which was performed with a prediagnosis of discal pathology, revealed two mass lesions. The patient underwent surgery. Histopathological sectioning of both tumors revealed that tumor tissues showed mucinous stromal changes. There were cuboidal uniform tumor cells which spread papillary structures usually as single-row on the hyalinized stroma. There was no mitosis. The patient was followed for 15 months. Postoperative spinal MRI was performed 6 and 12 months after surgery showed no recurrence of the tumors. Double independent MPEs of filum terminale are rarely reported. Right interpretation and diagnosis are open to discussion with diagnosis of drop metastasis and two different foci of an ependymoma. Cranial and whole spinal MRI with Gadolinium included sacrococcygeal region should be done in patient with MPEs. En bloc total removal of multifocal MPEs gives a good prognosis.

Key words: *Myxopapillary ependymomas, multifocal, surgery.*

Level of Evidence: *Case report, Level IV*

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ÖZET:

Miksopapiller epandimom (MPE), spinal kord epandimomlarının bir varyantıdır. Spinal kord tümörlerinin % 40-60'ını oluşturur ve tipik olarak filum terminal eve konus medullaristen köken alır. Bu olgu sunumu yazısında filum terminalenin iki ayrı bölgesinden köken alan multifocal bir epandimom olgusu literatürdeki benzer olgularla birlikte değerneldirilerek sunulmaya çalışılmıştır. 24 yaşında herhangi bir sağlık problem olmayan bir erkek hasta yaklaşık 2 aydır sağ bacağına yayılan bel ağrısı yakınması ile merkezimize başvurmuştur. Yapılan nörolojik muayenesinde herhangi bir motor, duyusal veya sfinkter kusuruna rastlanmadı. Disk patolojisini verifiye etmek için çekilen lumbosakral MR'da 2 adet kitle lezyonu saptandı. Hastaya cerrahi uygulandı. Çıkarılan kitle lezyonunun histopatolojik incelemesinde tumor dokusunun müsinöz stromal değişiklikler gösterdiği saptandı. Mitoz gözlenmedi. Hasta 15 ay takip edildi ve ameliyat sonrası 6. ve 12. aylarda yapılan MR incelemelerinde nüks görülmedi. Filum terminalenin birbirinden bağımsız 2 ayrı epandimomu çok seyrek olarak görülen bir durumdur. Sorunun kaynağının 2 ayrı yer mi olduğu ya da bir drop metastaz mı olduğu tartışmalı bir konudur. Kranial ve tüm spinal kontrastlı MR incelemesi bu tür hastalarda mutlaka yapılmalıdır. Tümörün en blok çıkartılması sonucu prognoz iyidir.

Anahtar Kelimeler: *Miksopapiller ependimoma, multifokal, cerrahi tedavi*

Kanıt Düzeyi: *Olgu Sunumu, Düzey IV*

INTRODUCTION:

Myxopapillary ependymomas (MPE) are distinctive variant of spinal cord ependymomas, typically arise in the filum terminale and conus medullaris, accounting 40-60% of spinal cord tumors [13,15]. MPE is WHO grade I, often encapsulated, slow-growing tumor, appear to be characterized a benign course and long-term survival [5]. Although MPEs tend to recur locally, dissemination and metastasizing had been occasionally reported [12]. Dissemination is primarily thought in patient with multiple spinal lesions, but concomitant localization of a MPE had been rarely reported [4,6-7,10].

MPEs are potentially curable tumor because they are amenable to complete surgical resection. Gross total en bloc resection should be attempted when possible. If total or subtotal piecemeal removal has been achieved, patients can be followed with serial magnetic resonance imaging (MRI) because of local recurrence and seeding via cerebrospinal fluid [14].

We report a case of MPE with two discrete foci on the both ends of the filum terminale and discuss solitary and multifocal MPEs with data of other four patients with double MPEs of filum terminale founded in the literature.

CASE REPORT:

A 24-year-old previously healthy male presented with a two-month history of back pain radiating to his right leg. He reported no motor, sensory, bowel, or bladder dysfunction.

Plain lumbosacral radiography showed no abnormality. A MRI examination of lumbosacral region which was performed with a prediagnosis of discal pathology, revealed two mass lesions. First lesion was in L1-L2 and isointense on T1 and T2-weighted images. The tumor was enhancing heterogeneously. Second lesion was in S1-S2 and had same image characteristics with lumbar lesion (Figure-1). Radiological features of lumbar lesion were concordant with ependymoma, but existing of a second lesion caused doubt and confusion. It is thought that both lesions were (might be) metastatic tumors or sacral lesion was a metastasis of upper lumbar lesion. Therefore, the patient underwent cranial and whole spinal MRI in order to evaluate the neuraxis, and no other lesion was identified.

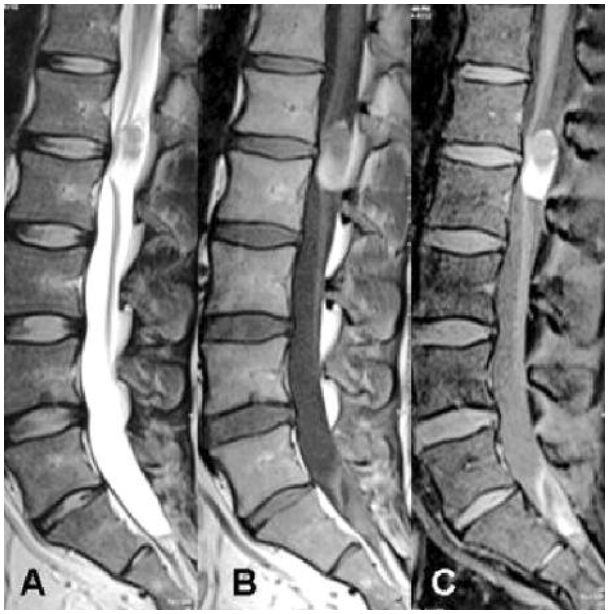

Figure-1. A MRI examination of lumbosacral region revealed two mass lesions. First lesion was in L1-L2 and isointense on T1 and T2-weighted images. The tumor was enhancing heterogeneously. Second lesion was in S1-S2 and had same image characteristics with lumbar lesion.

The patient underwent surgery, with intraoperative recording of somatosensory evoked potentials and sphincteric EMGs. Two separate laminectomies tailored to the extent of the tumors were performed. First, inferior half of L1 and superior half of L2 laminas were removed. When the dura was opened, encapsulated and reddish tumor was seen. The tumor was well-circumscribed and with no signs

of rupture on the pseudocapsular wall. The tumor was resected en bloc with attached division of filum terminale. Second, S1 and S2 laminectomies were performed and at the intradural exploration, reddish, encapsulated tumor was removed en bloc with attached distal end of filum terminale.

The postoperative course was uneventful. Histopathological sectioning of both tumors revealed that tumor tissues showed mucinous stromal changes. There were cuboidal uniform tumor cells which spread papillary structures usually as single-row on the hyalinized stroma. There was no mitosis (Figure-2).

The patient was followed for 15 months. Postoperative spinal MRI was performed 6 and 12 months after surgery showed no recurrence of the tumors.

DISCUSSION:

Ependymomas are the most common intramedullary tumors in adults and arise from ependymomal cells on the central canal of the spinal cord, the filum terminale, and the white matter adjacent to a ventricular surface [8,13]. MPE is a specific subtype of ependymomas and account 90% of primary filum terminale tumors[17]. The designation "myxopapillary" is based on the tendency of this variant ependymoma to produce mucin and, by virtue of its arborizing vasculature, to form papillae [15]. MPE is characterized by distinctively abundant supporting fibrous connective tissue stroma, mucinous degeneration of this supporting stroma and mucin secretion by the tumor cells [15]. Although other histological types also occur in the conus medullaris, none demonstrate the predominant mucinous degeneration [16]. The distinctive histologic feature of MPE may be due to direct apposition of ependymal cells to the connective tissue in the filum [6,16].

MPEs have been considered benign tumors with rare cases of tumor dissemination. Plans et al. [12] reported 23 cases of subarachnoid dissemination. Although, reported cases of tumor dissemination and systemic metatases have increased significantly after the advent of MRI, MPEs are characterized by a benign course and good long-term survival [5]. Local recurrence is still the main expression of the malignant behavior of MPEs.

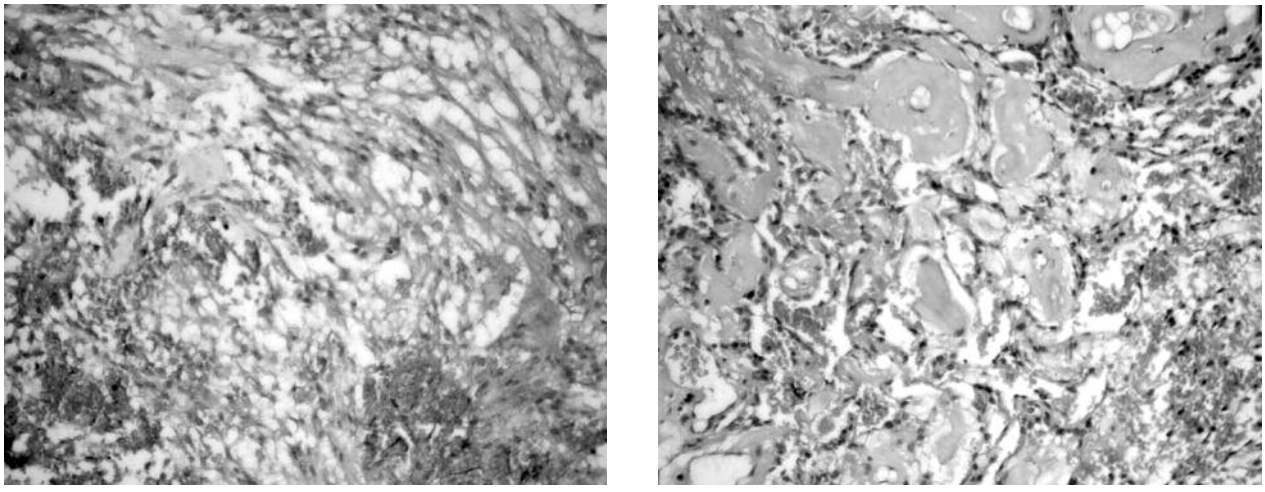

Figure-2. Histopathological sectioning of both tumors revealed that tumor tissues showed mucinous stromal changes **(a).** There were cuboidal uniform tumor cells which spread papillary structures usually as single-row on the hyalinized stroma **(b).** There was no mitosis.

There are no obvious clinical or histological features which allow a reliable prediction of the ability of MPEs for dissemination and metastasis [3]. Mitotic activity, cytologic atypia, extensive mucinous change and, hyalinization are unassociated with recurrence rates and differences in prognosis [15].

En bloc total removal exhibited a lower proportion of dissemination, the only factor likely to have a positive influence on the prognosis. Piecemeal resection, even when completely resected, or partial resection carry a higher incidence of recurrence and dissemination. Violation of the capsule during surgery may lead to CSF seeding and dissemination [12]. Postoperative radiotherapy appears to improve local control and dissemination in patients with piecemeal or subtotally resected MPE. Early postoperative MRI with regular follow-up studies enables the recognition tumor recurrence and expansion [9].

Differential diagnosis of MPE in the region of the conus medullaris should include other varieties of ependymomas, astrocytoma, and hemangioblastoma. Subependymomas may arise in the filum terminale and have imaging characteristics virtually identical to MPEs. Other lesions include paragangliomas, nerve sheath tumors, meningioma, and metastases [16]. Less common conditions include primitive neuroectodermal tumor, lipoma, dermoid cyst, cholesteatoma, lymphoma, and neuroenteric cyst [16].

In present case with double lesion, radiological findings caused a doubt in diagnosis. Two independent lesions at the two ends of the filum terminale is troublesome in terms of defining its pathogenesis [7]. In the current case, localization and radiological features of lumbar lesion were concordant with ependymoma. It is thought that sacral lesion was drop metastasis, a discrete lesion or both lesions were metastases, and cranial and whole spinal MRI with gadolinium was performed and no other lesion was found. Trauma and intratumoral hemorrhage are known to cause tumor rupture, and dissemination along the nerve roots [11], and both tumors of our case had well circumscribed and intact capsule in surgical exploration. There was no capsular rupture causing drop metastasis or dissemination to the other localizations of central nervous system along the CSF pathways. In sacral intradural exploration, the filum terminale were going into lesion and attached to tumor. Local invasion or dissemination of lumbar tumor to the sacral region throughout filum terminale was not thought because of filum terminale between the two lesions was normal. Hallacq et al. [7] reported the filum terminale was normal between the two tumors in a patient with double MPEs and interpreted this may present one end of a spectrum, with a giant tumor of the cauda equina being to other end.

Clinical and radiological characteristics of five patients include our case presented at Table-1. Although MPE was seen commonly at third and fourth decades, mean age of five patients with bifocal MPEs was 15, and four patients, out of our case, were in pediatric age group. All patients underwent total resection without radiotheraphy. Follow-up time was 5 years without MRI in one patient, between 6 and 15 months in 3 patients and unknown in one patient and there were no local recurrence or dissemination. Favorable follow up results of patients preoccupied, the two lesions were independent ependymomas more than dissemination or metastasis.

In conclusion; double independent MPEs of filum terminale are rarely reported [1]. Right interpretation and diagnosis are open to

Table - 1. Clinical and radiological characteristics of five patients with MPEs including our case presented.

	Author, year	Age/sex	Symptoms examination	Neurological	Location surgery	Extent of (month)	Follow-up
1	Gonzales, 2001	15/K	Back and leg pain	No deficit	L2-3, S2-3	Complete removal	6
2	Hallacq, 2003	13/E	Back pain	No deficit	L2-3, S2	Complete removal	60(No MRI)
3	Nakama, 2005	7/E	Back pain, dysuria	No deficit	L1-2, S2-3	Complete removal	12
4	Falco, 2008	16/E	Weakness on both legs	Paraplegia	D7-9, L5-S2	Complete removal	NA
5	Current case	24/E	Back and leg pain	No deficit	L1-2, S1-2	Complete removal	15

discussion with diagnosis of drop metastasis and two different foci of an ependymoma [2]. Cranial and whole spinal MRI with Gadolinium included sacrococcygeal region should be done in patient with MPEs [3]. En bloc total removal of multifocal MPEs gives a good prognosis [4].

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