VAN TIP DERGİSİ Medical Journal

CLINICAL RESEARCH / KLİNİK ÇALIŞMA

Van Tıp Derg 30(1):72-77,2023
DOI: 10.5505/vtd.2023.42223

Cervical Meningomyelocele - Single Center Experience

Servikal Meningomyelosel - Tek Merkez Deneyimi

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Abstract

Introduction: Cervical meningomyelocele (MMC) is rarely seen compared to lumbosacral and thoracolumbar meningomyelocele. There are only a few series related to cervical MMC in the literature. This study presents one of the most extensive series of cervical meningomyelocele, reviewing its clinical features, surgical management, and management strategies.

Materials and Methods: A total of 520 spina bifida patients, 25 of whom were diagnosed with cervical meningomyelocele, from January 2010 to September 2022, were included in the study.

Results: 88% (22) of the patients included in the study were newborns. The mean age was 3 days. Of the patients, 52% (13) were female and 48% (12) were male. The most common sites of cervical meningomyelocele were C4-C5, C5-C6, and C7-T1 regions with similar rates of 24%. There was a cranial anomaly in 56% (14) of the patients. The most common cranial anomalies were Chiari II with 24% (6), hydrocephalus, and Chiari type II with hydrocephalus and syringomyelia with 16%. All patients underwent surgical resection of the sac and intradural exploration.

Conclusion: Cervical meningomyelocele is structurally and clinically different from thoracolumbar and lumbosacral meningomyelocele and has more favorable outcomes after surgery. Preoperative magnetic resonance imaging and detailed patient evaluation are recommended to identify the cervical meningomyelocele's sac and spinal cord structure and additional anomalies. Surgical treatment should be done early and intradural exploration is recommended in addition to resection of the sac.

Keywords: Cervical meningomyelocele; myelomeningocele; chiari malformation; hydrocephalus; syringomyelia.

Özet

Amaç: Servikal meningomyelosel (MMS), lumbosakral ve torakalomber meningomyelosellere göre nadir görülür. Literatürde, servikal MMS ile ilgili sadece birkaç seri bulunmaktadır. Bu çalışma, en geniş servikal meningomyelosel serilerinden birini sunarak klinik özelliklerini, cerrahi tedavisini ve yönetim stratejilerini gözden geçirmektedir.

Materyal ve Yöntem: Çalışmaya Ocak 2010'dan Eylül 2022'ye kadar 25'ine servikal meningomyelosel tanısı konulan toplam 520 spina bifida hastası dahil edildi.

Bulgular: Çalışmaya alınan hastaların %88'i (22) yenidoğandı. Yaş ortalamaları 3 gündü. Hastaların %52'si (13) kadın, %48'i (12) erkekti. Servikal meningomyeloselin en sık görüldüğü bölgeler %24'lük benzer oranlarla C4-C5, C5-C6 ve C7-T1 bölgeleriydi. Hastaların %56'sında (14) kranial anomali mevcuttu. En sık görünen kranial anomaliler %24 (6) ile Chiari tip II, hidrosefali ve %16 (4) ile Chiari tip II, hidrosefali, sringomiyeli idi. Tüm hastalara kesenin cerrahi rezeksiyonu ve intradural eksplorasyon uygulandı.

Sonuç: Servikal meningomyelosel, torakolomber ve lumbosakral meningomyeloselerden yapısal ve klinik olarak farklılık gösterir ve cerrahi sonrası daha olumlu sonuçları vardır. Servikal meningomyeloselin kese ve spinal kord yapısı ile ek anomalileri tanımlamak için cerrahi öncesi manyetik rezonas görüntüleme yapılması ve hastanın detaylı değerlendirilmesi önerilir. Cerrahi tedavi erken yapılmalı ve kesenin rezeksiyonuna ek olarak intradural eksplorasyonu yapılması önerilir.

Anahtar Kelimeler: Servikal meningomyelosel; miyelomeningosel; chiari malformasyonu; hidrosefali; sringomiyeli.

Introduction

Spina bifida aperta mostly consists in the lower parts of the spine and is seldom in the cervical spine. It is a congenital spinal anomaly in the posterior midline of the cervical spine. This subgroup, defined as cervical meningomyelocele (MMC), constitutes only 3-8% of all spina bifida aperta cases (1, 2). Cervical MMC differs clinically from the more common lumbosacral MMC. Fewer neurological defects at presentation and a better clinical response following surgery make them a different entity when compared to other forms of spinal dysraphism such as common thoracolumbar or lumbosacral which have a higher prevalence (3). Diagnosis and treatment plans should be made by defining specific clinical traits. Thus, an inappropriate surgical technique may cause delayed neurological deterioration (4,5). There are not many publications about these lesions in the literature. The majority of the publications

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Received: 04.11.2022, Accepted: 23.12.2022

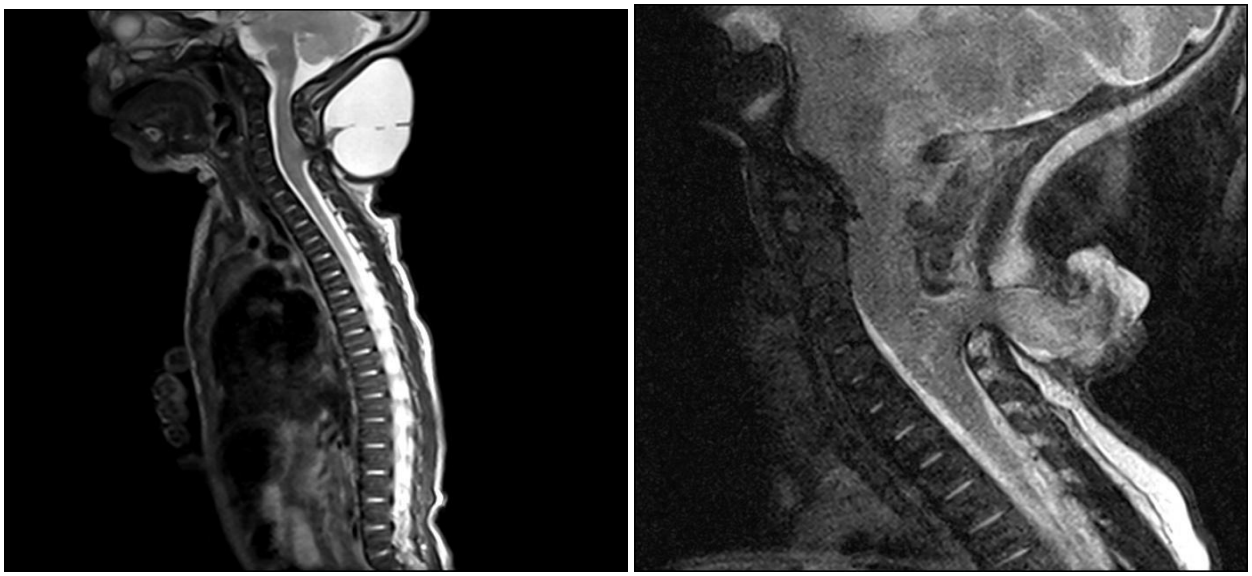

Figure 1, 2: Sagittal MRI of Cervical Meningomyelocele

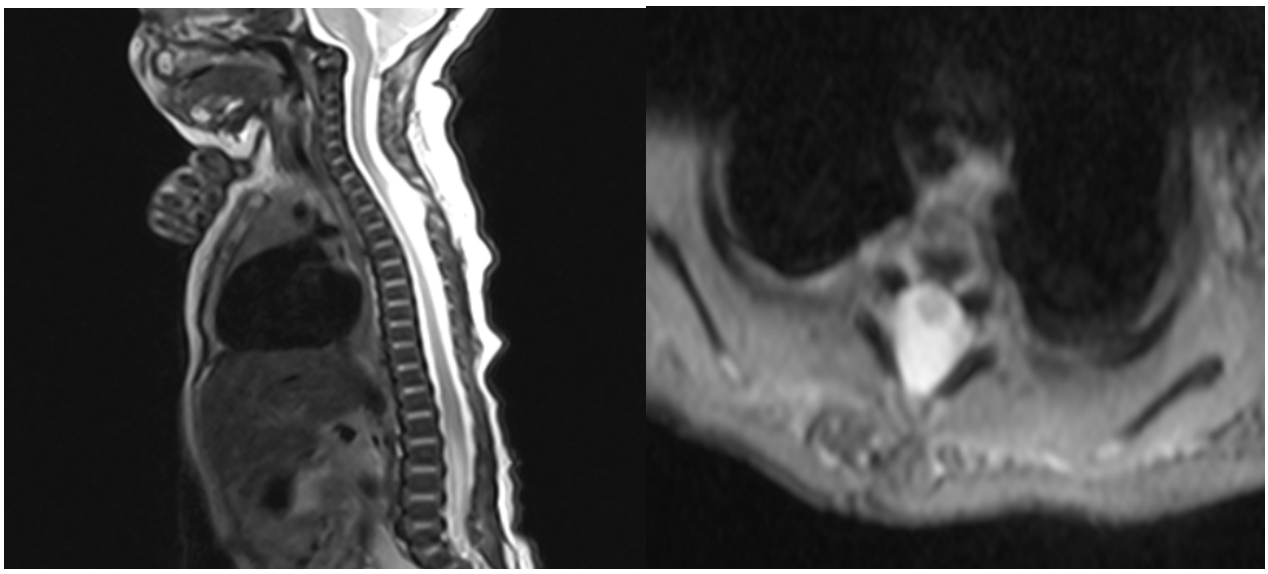

Figure 3: Postoperative Cervical Meningomyelocele, sagittal and axial MRI

investigating these lesions are limited to case series (2,3,6). In this study, we present 25 surgically treated cases of cervical MMC and aimed to discuss their clinical treatment and management strategies comparatively with the literature.

Material and Method

Patients: A total of 520 patients diagnosed with congenital spina bifida at the University hospital from January 2010 to September 2022 were included in this retrospective study. Of these 520 patients, 4.8% (25) of them were diagnosed with cervical MMC. The clinical data of the 25 patients, including their demographic profile, clinical traits, associated neurological malformations, radiological findings, surgical treatment strategies, retrospective results, and follow-ups were obtained and analyzed. The median age ranged from 8.92±18.45 (median 3) days. Thirteen of the patients were female and 12 were male. The mean age of the patients was 3 days, and 52% (13) of the patients were female, while 48% (12) were male. Neurological and radiological examinations, including magnetic resonance imaging (MRI), were performed for all patients (Figure 1,2,3). All 25 patients displayed a soft mass in the cervical region in the midline of the back; the mass had a

relatively wide dome covered with a full-thickness skin layer at the base and a thin purplish skin on top. Cerebrospinal fluid (CSF) leakage was not detected. Most of the lesions were over 10 cm in diameter. Following diagnosis, all patients had surgery using standard-of-care micro neurosurgery techniques. Ethics committee approval of this study was received from the Non-Invasive Research Ethics Committee with the date 14.10.2022 and reference number 2022/10-26.

Surgical procedures: The patient, with all pressure points supported, was placed on the operating table under general endotracheal anesthesia in the prone position. In the surgery, a longitudinal skin incision was made on the ceiling of the mass. The skin was meticulously dissected from the incisors and descended to the base of the pouch. It was circumferentially dissected towards the fascia, where the sac was seen to narrow with a stalk and merge with the dura. It was resected up to the adjacent laminae when deemed necessary in the cases. After the upper part of the meningomyelocele, the sac was opened, and a thick fibrous band extending to the dorsal surface of the sac was seen. After the dura was spread around the peduncle, it was observed to enter the expanded cord in the posterior midline. This junction in the sac was sharply dissected and released using the microsurgical dissection technique. Stem amputation was performed at the cord level so that the fibro neural bands were completely free. In patients with neural tissue in the sac, after the fibrous bands were released, the neural tissues were left in place to avoid postoperative neurologic deficits. All patients underwent excision of the sac with standard micro-neurosurgery techniques and intradural exploration and handling of other associated anomalies.

Ethical consent: Ethics committee approval of this study was received from the Van Yuzuncu Yıl University Non-Invasive Research Ethics Committee with the date 14.10.2022 and reference number 2022/10-26.

Statistical analysis: Data were analyzed using the SPSS 22.0 statistical program. Number and percentage values were given for categorical variables. Categorical variables were evaluated with the Pearson Chi-Square test. The limit of significance was taken as $p \leq 0.05$.

Results

88% of the patients were newborns. The mean age of the patients, 48% male, and 52% female, were 8.92±18.45 (median 3) days. The most common sites of cervical meningomyelocele were C4-C5, C5-C6, and C7-T1 regions with similar rates of 24%. The cranial anomaly was present in 56% of the patients. The most common cranial anomalies were Chiari type II with 24%, hydrocephalus, and Chiari II with hydrocephalus, and syringomyelia with 16%. The pouch size is divided into two groups as below and above 10 cm diameter. The sac size was above 10 cm in diameter in 68% of the patients. Frequency distributions according to some variables are presented in Table 1.

Table 1: Patient Characteristics

	n	%
Age		
Newborn	22	88
Infant	3	12
Sex		
Female	13	52
Male	12	48
MMC Region		
C2-C3	3	12.0
C3-C4	2	8.0
C4-C5	6	24.0
C5-C6	6	24.0
C6-C7	2	8.0
C7-T1	6	24.0
Sac Diameter		
<10 centimeters	8	32
>10 centimeters	17	68
Concurrent Anomaly		
No	11	44
Yes	14	56
Cranial Anomaly		
None	10	40.0
Chiari, hydrocephalus	6	24.0
Chiari, hydrocephalus, syringomyelia	4	16.0
Chiari	2	8.0
Chiari, corpus callosum dysgenesis	2	8.0
Diastematomyelia	1	4.0

Table 2: Concurrent Anomaly Regarding Sac Size

	Concurrent Anomaly		P**
	None n (%)*	Present n (%)*	
Sac Diameter			
<10 centimeters	6(75)	2(25)	0.034
>10 centimeters	5(29.4)	12(70.6)	

* Row percentage is given. ** Pearson Chi-Square test

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All patients underwent sac excision. Pre- and postoperative examinations of all patients were normal, except for one patient with preoperative lower extremity paralysis. Patients were followed up regularly from 1 to 12 years and showed no neurological impairment. In the postoperative course of all patients, no visible changes in their neurological status, and postoperative complications such as cerebrospinal fluid leakage and infection were observed. There was no mortality during the follow-up. The presence of anomaly by sac size is presented in Table 2. There is a statistically significant difference between sac size and the presence of anomaly ($p<0.05$). The number of anomalies was significantly high in patients with a sac size greater than 10 cm in diameter.

Discussion

Meningomyelocele, a subgroup of spina bifida aperta, most commonly occurs in the lumbar and sacral regions of the spine. Cervical MMC are a less common but important subtype of myelomeningoceles. It is relatively rare and has a much lower incidence than the thoracolumbar and lumbosacral types (6-8). Cervical MMC embryology and clinical features differ from the more widespread thoracolumbar and lumbosacral variants (2). Various theories are being considered for the origin of clinical presentations of cervical meningomyelocele 3,7,9,10,11It was originally clarified on the basis of the regular closing theory. Accordingly, the neural tube closing stops in the mid-cervical region and spreads both rostrally and caudally in a Zipper-like fashion, with the cranial and caudal neuropores closing last. Van Allen improved the multisite theory to clarify the formation of neural tube defects. Moreover, cervical MMC has a few distinctive characteristics that distinguish it from thoracolumbar MMC. Among these, unspoiled neurological functions, full-thickness skin covering, and related to Chiari II malformation are lesser common. The content is not a mislaid neural plate, but a neuroglial stem that originates from the dorsal surface of the cervical cord and produces tethering. These characteristics are better elucidated by the Limited dorsal myeloschisis theory (LDM) of Pang and Dias (10). Accordingly, neurulation proceeds smoothly and the primary configuration of the neural tube is maintained, outside of a subtle shift in the dorsal midline. There is never really a separation between the cutaneous ectoderm and the neuroectoderm. Even though myofascial tissues evolve the dorsal median stem of central nervous system tissue, it continues the original link between the almost closed neural tube and the still lightly spaced cutaneous ectoderm. Apart from an extension around this stem, the meninges also develop. As CSF is formed, it slowly becomes trapped in the dural fistula and slowly widens the dome, building myelomeningocele (6). In our study, 25 cervical MMC cases were diagnosed out of 520 congenital spina bifida anomaly cases in our hospital between January 2010-September 2022. Cervical MMC incidence was found to be 4.8% (25/520) and it is lower than the literature (1,2,3,6). The general clinical presentation of cervical MMC presents with subcutaneous swelling in the posterior cervical region without significant neurological impairment (3,8,12,). In our case series, except for one patient, the others had no neurological deficits. Surgery for the lumbosacral and thoracolumbar forms of MMC is aimed at preventing mechanical injury and infection to the neural placode, and to some degree, preventing the progress of the neurological deficits offered in an important proportion of children. Moreover, in cervical MMC, surgery is done primarily for cosmetic reasons and the protection of neurological deficits by releasing all attached elements. Untreated children may develop neurological deficits later in life. (6,13). The fact that none of our patients evolved sensory symptoms or deficits and had normal intestine and bladder functions during surgery was similar to the literature (1). This indicates that although congenital lesions originate from the dorsal cervical cord, dorsal column function may remain intact. As noted in the literature, cervical MMC may have a high tendency to be related to other spinal abnormalities such as hydrocephalus, syringomyelia, diastematomyelia, tethered cord, Chiari malformation, Klippel-Feil syndrome, thoracic hemivertebrae compared to caudal myelomeningoceles (14, 15). In our series, 14 patients had accompanying anomalies. These were Chiari II malformation, hydrocephalus, syringomyelia, corpus callosum dysgenesis, and one diastematomyelia from spinal anomalies, from most to least. In addition, a statistically significant difference between sac size and the presence of anomaly was observed ($p<0.05$). In our series, an increase was found in the amount of anomaly when the size of the swabs increased. Cervical MMC may be associated with cranial and spinal anomalies. MRI (magnetic resonance imaging) is required both to show fine anatomic relationships at the lesion level and to detect other spinal cord abnormalities. (15-17). Therefore, cranial and whole spinal MRI should be performed in patients diagnosed with cervical MMC. A lack of

understanding of the anatomical details of lesions and related abnormalities can lead to treatment failure and poor prognosis. In our cases, MRI was also taken before and after surgery. In our patients, cervical MMC localization was more common at C4-C5, C5-C6, and C7-T1 levels in MRI. In conclusion, MRI is indispensable to determine the cervical MMC level and to see the anatomical structure, and all spinal and cranial MRI is recommended (16,17). As stated in the literature, cervical MMC should be intervened early before any neurological deficit starts in patients (7,10, 18). In our clinic, as soon as cervical MMC patients were diagnosed, they were supported by the necessary imaging and surgery. Except for three of our patients, all of our patients underwent surgery during the neonatal period. Surgical treatment should insert a bi-level laminectomy, intradural exploration to remove the band of tissue connecting the spinal cord, and resectioning of the related protruding sac. A microsurgical technique should be utilized to detect any spinal cord tethering or any related spinal dysgraphic abnormality (7,10,19). An insufficient surgical technique in untreated cervical cord tethering may result in delayed neurologic deterioration. In all of our cases, intradural exploration was performed to remove the tissue bands connecting the cord, and neurologic deterioration was not observed in any of the patients after sac resection. Therefore, cervical MMC surgery should be aimed not only for cosmetic reasons but primarily at the prophylactic release of all connected cord elements and avoiding functional loss. The prognosis of the cervical MMC patient is quite favorable in neurological and systemic aspects when compared with patients with thoracolumbar and lumbosacral meningomyelocele. However, delayed neurological deterioration may occur in growing children because of tethering. Therefore, it is important to follow up with the patients regularly after surgery.

Study limitations: The low number of cases and the absence of photographs or videos during the operation are the shortcomings of our study, since we could not reach the data of the old cases.

Conclusion

Cervical meningomyelocele is structurally and clinically distinct from lumbosacral and thoracolumbar meningocele and has a more favorable course. Preoperative MRI is recommended to identify sac and spinal cord structure and additional anomalies in patients with cervical MMC who have intact neurological function. Surgical treatment should be performed early and besides resection of the sac, adequate laminotomy, intradural exploration, and excision of all ligature bands and septa are suggested. The postoperative outcome of cervical MMC lesions is satisfactory. Because postoperative neurologic deficits are not usually seen or are very rare. Surgery helps prevent the development of neurological deterioration that may later develop.

Ethical consent: Ethics committee approval of this study was received from the Van Yuzuncu Yıl University Non-Invasive Research Ethics Committee with the date 14.10.2022 and reference number 2022/10-26.

Conflict of interest statement: The authors have no conflict of interest for this study.

Financial disclosure: No financial support has been received for this study.

Author contributions: Concept (MEA, OA), Design (MEA), Data Collection and/or Processing (MEA, OA), Analysis and/or Interpretation (MEA, OA)

Acknowledgment: For sharing the data of his patients with us, thanks to Prof. Dr. Mehmet Arslan

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