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Lumbosacral Meningocele in Adulthood

Erişkinde Lumbosakral Meningosel

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ABSTRACT

Spinal dysraphism is the incomplete fusion of the neural arch, which can be seen as an occult or open neural tube defect. Meningoceles are a form of open neural tube defect and characterized by cystic dilatation of the meninges containing cerebrospinal fluid (CSF), without the involvement of neural tissue. These lesions are often benign and typically diagnosed at birth. Neurosurgical intervention is necessary in the newborn period, since survival in advancing ages is often impossible. Therefore, meningoceles are rarely reported among spinal dysraphic lesions in adulthood. They are tethering lesions of the spinal cord, adhering to the dura and other soft tissues with fibrous elements. In addition, the caudal lesions tend to leak CSF, unlike cervical congenital midline meningoceles. Here, we present a 41 year-old female patient with a meningocele that has developed CSF leakage. The clinical course, surgical management, and follow-up period of this rarely seen adulthood meningocele are also discussed.

Key words: Adulthood, meningocele, spinal dysraphism.

ÖZET

Spinal disrafizm nöral arkın kapalı veya açık nöral tüp defekti şeklinde görülen inkomplet füzyonudur. Meningoseller açık nöral tüp defektleri olup meninkslerin içi beyin omurilik sıvısı (BOS) dolu ve nöral doku içermeyen kistik dilatasyonları olarak görülürler. Doğum sırasında tespit edilen benign lezyonlardır. Uzun yaşam süresi ile bağdaşmayacağından, meningosele yönelik cerrahi girişim hemen yenidoğan periyodunda yapılmalıdır. Meningoseller erişkin spinal disrafik lezyonlar arasında nadiren rapor edilmiştir. Duraya ve diğer yumuşak dokulara fibröz bantlarla yapışarak spinal kordu gererler. Servikal konjenital orta hat meningosellerinin aksine kaudal yerleşimli lezyonlar genellikle BOS sızıntısına neden olurlar. Lezyon yerinden BOS sızıntısıyla başvuran 41 yaşındaki kadın meningosel olgusunu rapor ediyoruz. Klinik gidişat, cerrahi yaklaşım ve ameliyat sonrası takip sürecindeki müdahaleler tartışılmıştır.

Anahtar kelimeler: Erişkin, meningosel, spinal disrafizm

INTRODUCTION

Meningoceles are a form of open neural tube defect, and characterized by the protrusion and cystic dilatation of spinal meninges, which contain cerebrospinal fluid (CSF) without the involvement of neural tissue. They compose 10% of the patients with spinal dysraphism at birth and are seen posteriorly in the cervical, thoracal, and lumbosacral regions[1].

Meningoceles protrude anteriorly and laterally, but anterolateral defects can also seen. The anterior sacral meningocele is an example of this type of lesion[2,3]. Open forms of sacral cases in adulthood have rarely been reported, since survival with a meningocele into later ages is

unusual[4]. To manage the complication of meningitis, meningocele cases should be operated on immediately after birth, if possible, in the first twenty-four hours, especially when it is perforated[1,2].

Here, we report a rare case of an adultmeningocele patient, who was not operated on in childhood, presenting as an adult with a perforated meningocele.

CASE

A 41 year-old female patient was admitted to our hospital with a complaint of - leakage from her sacral lesion, and the patient did not indicate any trauma to the lumbar region before the leakage. A sacral meningocele was diagnosed during her childhood, but the patient was not operated on because she did not accept the surgical risks. Her physical examination revealed a giant meningocele lesion in the lumbosacral region with CSF leakage. The patient had no back pain and leg pain and the neurological examination revealed no deficits. She had no bladder or stool complaints, and the patient had perianal sensation with a normal anal reflex. Cranial Computed Tomography (CT), lumbar CT, and lumbar magnetic resonance imaging (MRI) investigations were done (Figures 1b, 1c). The lumbar CT and MRI showed a spina bifida defect at the L5-S1 level, and protrusion of the meningocele sac with a normally lying conus. No hydrocephalus was seen in her cranial CT. Preoperative preparations were made, but the patient refused surgery, even tough the risks of not having surgery were explained.

One day after she was examined in the emergency department and admitted to the hospital for a perforated meningocele (Figure 1a), the patient underwent emergency surgery. After entering the perforated sac, dural dissection from the skin was made in a circular fashion. No neural tissue components were observed inside the pouch, and the dura was meticulously dissected from the rudimentary neural tissue present, then the dura was sutured. Fibrin adhesive tissue sealant was used to firm up the dural tissues and prevent leakage (Figures 2a, 2b), and no neural deficits were observed during the post-operative period. The lumbar MRI revealed that the majority of the lesion was removed, and the pouch was filled with CSF (Figure 2c). The cranial CT control that was completed during the early period showed pneumocephalus with no hydrocephalus.

The patient was clinically stable throughout the one-year follow-up. However, after this, she was admitted to the hospital again with CSF leakage from the operation site, after recent lumbosacral trauma. Routine clinical and radiological investigations were conducted, and she was conservatively treated with bed-rest and medication; however, lumbar spinal drainage was placed, since there was no response to the conservative treatment. After one week of follow-up with drainage, no leakage was observed from her operation site and she was discharged. After six months, she was clinically stable, had no neurological deficits, and there was no leakage from her operation site during her hospital visits.

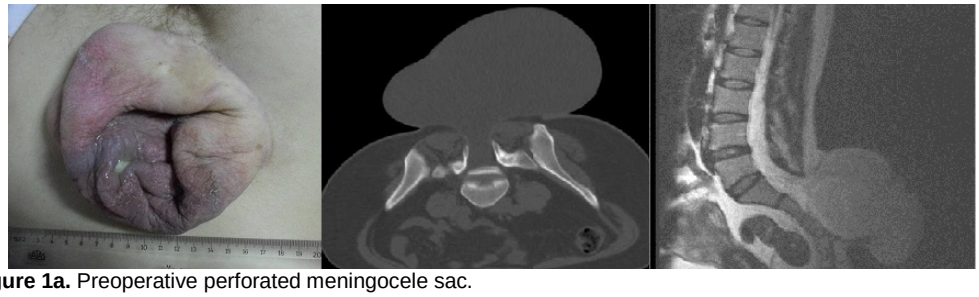

Figure 1a. Preoperative perforated meningocele sac.

Figure 1b. Preoperative lumbar CT demonstrates spina bifida defect and a large meningocele sac.

Figure 1c. Preoperative lumbar T2 weighted sagittal MRI showed protrusion of the meningocele sac.

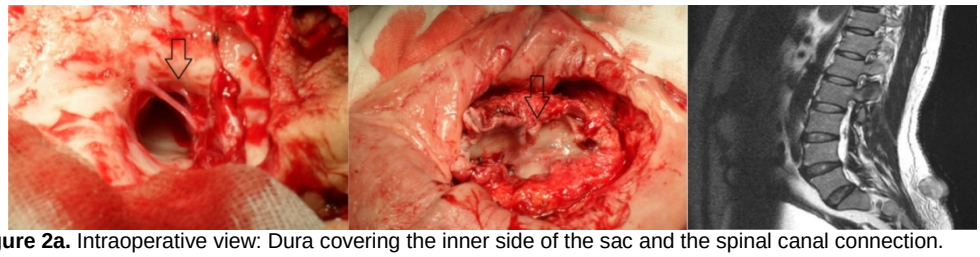

Figure 2a. Intraoperative view: Dura covering the inner side of the sac and the spinal canal connection.
Figure 2b. Dura dissected from the meningocele sac
Figure 2c. Postoperative T-2 weighted sagittal MRI

DISCUSSION

A meningocele is a congenital anomaly of neural arch fusion in association with an open neural tube defect, and characterized by protrusion of the spinal meninges which contain CSF without involvement of the neural tissue. Perforated sac lesions are especially dangerous, since infection is a great risk[1,2,4].

Most meningoceles are surgically repaired during the newborn period, or at least in childhood. The incidence of survival is low without intervention, so adult meningoceles are rarely seen. Life expectancy at birth is shorter in myelomeningocele patients, although effective treatment for hydrocephalus and intermittent catheterization for the management of the neurogenic bladder can improve the quality of life for these patients[1,5,6,7,8,9].

Adult tethered cord syndrome, occult intrasacral meningocele, cervical midline meningocele, and thoracic meningocele cases with neurofibromatosis have been reported in the literature, but posterior lumbosacral meningocele cases have rarely been reported[1,4,5,6,10,11,12,13,14]. The long-term follow-up results for adults with sacral myelomeningoceles are not as good as in children, since other neurological abnormalities such as hydromyelia, syringomyelia, tethered cord and chiari malformations, and hydrocephalus, accompany this lesion. Meningocele patients may improve after sac repair, although meningocele infections and hydrocephalus can be seen in these patients[15].

Early surgical intervention is recommended in meningocele cases. A good cosmetic result is one of the aims of surgical repair, in addition to preventing neurological deformities and infection[1,6,10,16,17]. In our case, infection was prevented by repairing the perforated meningocele sac, and a good cosmetic result was obtained by minimizing the pouch.

The MRI is a good investigation choice for evaluating the meningocele sac in the sagittal plane, and to observe the spinal cord itself, as well as the possible congenital anomalies associated with it. Ultrasonography may also be useful in both the pre and postoperative periods[2,18]. Our patient was admitted to the emergency unit with a perforated meningocele sac, and preoperative radiological examinations were done in the prone position in order to prevent further damage. In addition, urodynamic tests are valuable in evaluating urinary continence[1]. Our patient was taken to emergency surgery, so the preoperative specific tests could not be done, with the exception of the essential radiological investigations.

In meningocele cases, neurological involvement is not seen as often as in myelomeningocele lesions, but the local signs of sacral nerve root involvement are seen as pain in both legs and bladder dysfunction. Taking this in to consideration, somatosensory evoked potentials (SSEP) can be used in these patients as in myelomeningoceles[3,13].

Bed sores are usually associated with sensorial involvement in myelomeningocele patients, which are explained by the increased risk of mechanical and thermal skin damage due to reduced sensation. However, sores can also be seen in meningocele patients due to CSF leakage, so the management of this is important in both of these groups[19].

After the repair and the closure of the sac, hydrocephalus may be seen as a common complication[1,2]. Neurodeficits, CSF fistula, and hydrocephalus were not observed in the early post-operative period in our patient, although she was taken for a clinical follow-up for the development of hydrocephalus. However, after 1-year of follow-up she again had a CSF leakage from the operation site, and this was treated successfully with lumbar drainage after a conservative treatment period.

Lumbosacral meningoceles are rarely seen in adulthood. Even though no neurodeficit is observed, preoperative investigations including lumbar MRI, CT, SSPE, and urodynamic tests should be conducted. Surgical treatment should be considered to prevent infection, increase the quality of life, and obtain a good cosmetic result.

REFERENCES

1. Akay KM, Ersahin Y, Cakır Y: Tethered cord syndrome in adults. Acta Neurochir(Wien). 2000;142:1111-5.
2. Akay KM, Gonul E, Ocal E, Timurkaynak E: The initial treatment of meningocele lesions in adulthood: experiences with seven patients. Neurosurg Rev. 2003;26:162-7.
3. Brinker M, Rosenfeld S, Feiwell E, Granger S, Mitchell D, Rice J: Myelomeningocele at the sacral level. Journal of Bone and Joint Surgery. 1994;76-A:1293-1300.
4. Dahlgren R, Baron E, Vaccaro A: Pathophysiology, diagnosis, and treatment of spinal meningoceles and arachnoid cysts. Department of Orthopaedic Surgery. Faculty Papers, 2007, p. 4. Available from: http://jdc.jefferson.edu/
5. Duz B, Arslan E, Gonul E: Cervical congenital midline meningoceles in adults. Neurosurgery. 2008;63:938-44.
6. Duz B, Gocmen S, Secer Hİ, Basal S, Gonul E: Tethered cord syndrome in adulthood. J Spinal Cord Med. 2008;31:272-8.
7. Gok B, Ayberk G, Tosun H, Seckin Z: Clinical course and evaluation of meningocele lesion in adulthood: a case report. Neuroanatomy. 2005;4:52-4.
8. Konya D, Dagcınar A, Akakın A, Gercek A, Ozgen S, Pamir MN: Cervical meningocele causing symptoms in adulthood: case report and review of the literature. Journal of Spinal Disorders and Techniques. 2006;19:531-3.
9. Laffey P, Kricun ME: Sonographic recognition of postoperative meningocele. AJR. 1984;143:177-8.
10. Lee GY, Paradiso G, Tator CH, Gentili F, Massicotte EM, Fehlings MG: Surgical management of tethered cord syndrome in adults: indications, techniques, and long-term outcomes in 60 patients. J Neurosurg Spine. 2006;4:123-31.
11. Pang D, Wilberger JE: Tethered cord syndrome in adults. J Neurosurg. 1982;57:32-47.
12. Piatt JH: Treatment of myelomeningocele: a review of outcomes and continuing neurosurgical considerations among adults. J Neurosurg Pediatrics. 2010;6:515-525.
13. Plaum PE, Riemer G, Froslie K: Risk factors for pressure sores in adult patients with myelomeningocele-a questionnaire-based study. Cerebrospinal Fluid Research. 2006;3:14.
14. Pool JL: Spinal cord and local signs secondary to occult sacral meningoceles in adults. Bull N Y Acad Med. 1952;28:655-63.
15. Shih P, Halpin R, Ganju A, Liu JC, Koski TR: Management of recurrent adult tethered cord syndrome. Neurosurg Focus. 2010;29:E5.
16. Stetler WR, Park P, Sullivan S: Pathophysiology of adult tethered cord syndrome: review of the literature. Neurosurg Focus. 2010;29:E2.
17. Walcott BP, Kahle KT, Wain JC, Borges LF : Giant thoracic meningocele associated with neurofibromatosis 1. Neurology. 2011;31;76:1943.

18. Woodhouse CRJ: Myelomeningocele in young adults. BJU. 2005;95:223-30.

19. Woodhouse CRJ: Myelomeningocele: neglected aspects. Pediatr Nephrol. 2008;23:1223-31.

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