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Dissynergic Bladder and Hydronephrosis in A Case of Spinal Intramedullary Dermoid Cyst

Ebru GERÇEK[a1], Metin KAPLAN[1], Fatih Serhat EROL[1], Mehmet Faik ÖZVEREN[2]

[1]*Fırat Tıp Merkezi, Nöroşirurji, ELAZIĞ, Türkiye*

[2]*Ankara Atatürk Eğitim Hastanesi, Nöroşirurji, ANKARA, Türkiye*

ABSTRACT

Spinal dermoid cyst is a rare pathology among the intamedullary spinal cord tumors. An 11 year old female patient applied with nausea, vomiting, and urine-bowel incontinence. The personal history of the patient revealed that she had been treated at the infectious diseases, pediatrics, and psychiatry departments of various centers for reccurent urinary tract infection and enuresis nocturna. Investigations revealed grade III hydronephrosis with dissynergic bladder and spinal intramedullary mass. The content of the mass was removed. The pathological diagnosis was dermoid cyst.The urinary incontinence of the patient improved to time to time night micturations. Hydronephrosis grade III had improved to grade II on the renal ultrasonography at the postoperative 1st year. In pediatric cases with frequent need for urination, continual drops after urination and relapsing urinary tract infections, the pathologies of neurogenic origin should be taken into consideration, and spinal imaging must be performed

Key words: *Dissynergic bladder, hydronephrosis, spinal intramedullary dermoid cyst*

ÖZET

Dissinerjik Mesane ile Presente Olan Spinal Dermoid Kist Olgusu

Spinal dermoid kistler, intramedüller spinal kord tümörleri içerisinde nadir bir gruptur. 11 yaşında kız hasta bulantı, kusma ve idrar inkontinansı şikâyeti ile hastanemize başvurdu. Hastanın anamnezinden çeşitli merkezlerin; enfeksiyon hastalıkları, pediatri ve psikiyatri kliniklerinde üriner sistem enfeksiyonu ve enuresis nokturna tedavisi gördüğü öğrenildi. Yapılan incelemelerde hastada grade III hidronefroz, dissinerjik mesane ve spinal intramedüller kitle tespit edildi. Kitle içeriği boşaltıldı ve patolojik tanı dermoid kist olarak raporlandı. Zaman zaman olan gece idrar kaçırmaları dışında üriner inkontinansı düzeldi. Postoperatif birinci yılında yapılan renal ultrasonografide hidronefrozun gradeIII'den gradeII'ye gerilediği gözlendi. Çocuklarda sık idrara çıkma, işeme sonrası damla şeklinde idrar yapma ve tekrarlayan üriner sistem enfeksiyonlarında patolojinin nörojenik kökenli olabileceği düşünülerek spinal görüntüleme yapılmalıdır.

Anahtar Sözcükler: *Dissinerjik mesane, hydronefroz, spinal intramedüller dermoid kist*

Spinal dermoid cyst is a rare intramedullary spinal cord tumor. It is usually encountered in the patients of childhood period (1,2,3). Intramedullary dermoid cyts are frequently located at conus level, and the symptoms develop slowly and gradually. The patients present with back pain, sensory-motor deficits, and sphincter problems (3,4). A case of intramedullary dermoid cyst with dissynergic bladder, and neurogenic bladder mechanism in intramedullary tumors has been discussed in this report.

CASE REPORT

An 11-year-old female patient applied to the Neurosurgery Department of Firat University Medical Faculty with nausea, vomiting, and urine-bowel incontinence. The personal history of the patient revealed that she had been treated at the infectious diseases, pediatrics, and psychiatry departments of various centers for recurrent urinary tract infection and enuresis nocturna. Upon her neurological examination, urinary sphincter deficit and loss of anal sphincter tonus were detected. All other neurological findings were normal. The urine analysis showed severe urinary tract infection. On the renal ultrasonography, both kidneys were hydronephrotic (grade III) (Figure 1), and the urodynamics revealed dissynergic type neurogenic bladder. The lumbosacral graphy of the patient presented spina bifida at L4-L5 level. On contrast lumbar magnetic resonance imaging (MRI), a non-enhancing intramedullary mass of 35x15mm was detected at L1 vertebra level. The mass was hypointense on T1 weighted images and hyperintense on T2 weighted images (Figure 2). The patient underwent a total laminectomy at L1-L2-L3 level in prone position and the intradural mass was accessed. The mass was distinguished from the normal spinal tissue with a dirty white capsule and had a content of hair and semisolid dirty white keratin-like material. The contents of the mass were discharged but the capsule was left unharmed (Figure 3). In the light of the histopathology of the mass, the diagnosis was a dermoid cyst.

[a] Corresponding Address: Dr. Ebru GERÇEK, Fırat Tıp Merkezi, Nöroşirurji, ELAZIĞ, Türkiye
e-mail: ebrugercek79@yahoo.com

On the postoperative 10th day, the anal sphincter tonus of the patient completely recovered. In the postoperative 9th month, the patient had no complaints of urinary incontinence other than occasional bed wetting at night. On the postoperative 1st year renal ultrasonography, hydronephrosis grade III had improved to grade II.

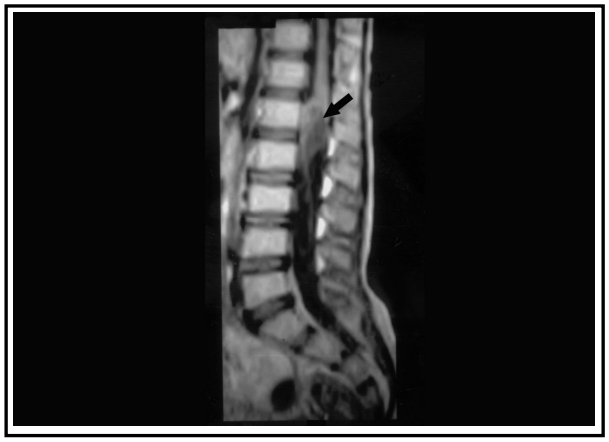

Figure 1. *Bilateral renal ultrasonography is showing an largement of both chalices (Grade III hydronephrosis)*

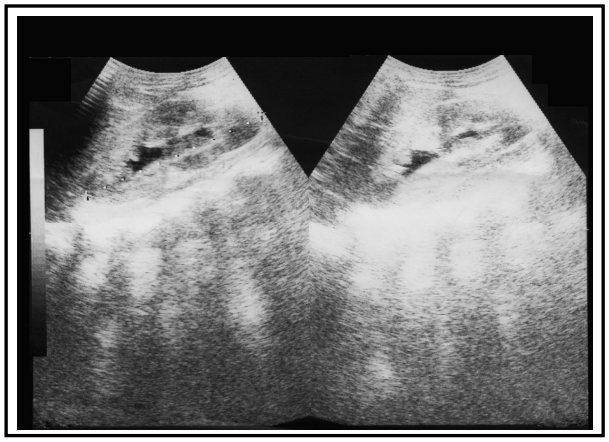

Figure 2. *There is no contrast enhancement on the T1 weighted sagittal lumbar magnetic resonance imaging (MRI) of the mass lesion at the conus medullaris level (arrow)*

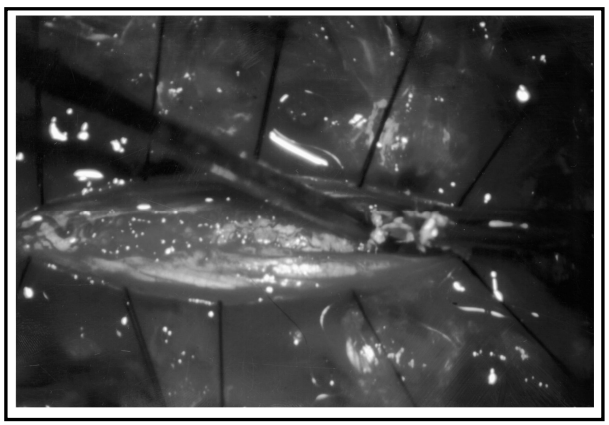

Figure 3. *The operative photography is showing the expansion of the conus medullaris (star) by the mass (arrow)*

DISCUSSION

Dermoid cysts are more frequent in childhood and comprise as little as 0.5-1% of intramedullary spinal tumors (5,6,7). Half of the spinal dermoid cyts that are intadural extramedullary tumors, while the other half is intramedullary located (8,9). Cervical or upper thoracic localization of dermoid cysts is a rare occasion, however, their conus level localization is common, as was in our case (10,11).

Spinal dermoid cysts are congenital and they develop through heterotopic embryological remnants or through focal expansion of dermal sinus along the conus or cauda. The cysts are usually accompanied with spina bifida and hemivertebra abnormalities, (10) as was in our case.

The symptoms of spinal dermoid cysts usually develop slowly in time (6,8,9). The most common complaint is back pain increasing in severity upon vertebral movements or valsavla maneuver. Intramedullary dermoid cyst cases often suffer from sensory and motor findings accompanied by the pain. Sensory deficits may be in the form of radicular or segmental paresthesia as well as dissociated loss of sense with insensitivity to pain and heat, and intact two-point-discrimination, position and deep tension sensation. Loss of strength due to motor involvement, atrophy and fasciculation, and disordered gait may be present. However, although less frequently, sphincter deficits, scoliosis and spinal subarachnoidal hemorrhages have been reported (11,12). Like in our case, 50% of the cases with dermoid cysts at conus level also have sphincter deficits (6,8).

The determination of bladder function disorders is difficult in children. Particularly the children only with sphincter deficit primarily apply to psychiatry, urology, nephrology and infectious diseases clinics (12). Therefore, the diagnosis is delayed and the clinical picture is deteriorated by the time the diagnosis is established. Of the patients applying to psychiatry clinics for enuresis nocturna, 5% have organic etiologies (11,5). Likewise, the primary pathology of 1-3% of the patients applying to urology clinics with neurogenic bladder picture has been found to be associated with a spinal mass (6,2). Therefore, in the cases with frequent need for urination, continual drops after urination and unaccountable urinary tract infections, particularly in children, the pathologies of neurogenic origin should be taken into consideration, and spinal imaging must be performed.

Urine control is provided by the peripheral reflex arcus in the sacral segments of the spine. This arcus is controlled by cortical centers. The urination center of the spinal cord is located at S2-S4 levels of conus medullaris. Based on their localization, the lesions revealing the sphincter deficits are termed as above S2-S4 suprasacral, S2-S4 sacral, and under this level, infrasacral. For normal function of urination, intact innervation of the bladder is required. In suprasacral lesions, detrussor hyperreflexia, and in sacral and infrasacral lesions, detrussor areflexia are expected (Table 1).

The muscles of the bladder are essentially parasympathetic. Pelvic splancnic nerves with parasympathetic effect originate from the sacral cord (S2-S4). They terminate at the ganglion located on the bladder wall and internal sphincter muscle. Parasympathetic stimulus stimulates contraction of the detrusory muscle and dilatation of the internal sphincter, through which the bladder is emptied.

The sympathetic nerves of the bladder originate from the lateral horns of T12-L1-L2 segments and extend to the bladder muscles through inferior hipogastric plexus. Sympathetic nerves leads to the contraction of internal sphincter muscle; thus, urine is accumulated in the bladder.

Somatic fibres, on the other hand, are located in the anterior horn (S2-S4) of the sacral cord. These are called pudental nerves and innervate external sphincter. When urine pass through the internal sphincter, this muscle opens and remains open until the bladder is empty

The neurogenic bladder picture develops after somatic, parasympathetic, and sympathetic systems are affected by various factors. Sympathetic system is the first to be affected in the intramedullary masses compromising the bladder function. In the sympathetic system disfunctions, internal urethral sphincter will not contract adequately, thus, leading to initial findings of neurogenic bladder urine leakage after urination and stress-related incontinence (7,8). Severely affected parasympathetic system, however, disrupts the detrusory mechanism, which results in a hypotonic and areflexed bladder. When both systems are involved, the patient not only loses his/her ability to empty the bladder but also develops incontinence due to sphincter deficit. While hypotonic bladder usually does not constitute a major risk for upper urinary tract, areflexed hypertonic bladder due to parasympathetic denervation may lead to reflux associated with high intrabladder tension.

The most severe reflux, which could lead to hydroneprosis, occurs in detrussory-sphincter dissynergies arising from incompatibility between the detrussory muscle and sphincter (6,7). Normally the efferent stimuli arriving at the external sphincter via pudental nerve and the stimuli arriving at the detrusor through pelvic nerves of parasympathetic system inhibit each other with the help of collateral fibres, but do not activate simultaneously (10). However, as a consequence of the dissynergie developing due to sacral spinal lesions, detrussory muscle contractions significantly increase the intrabladder tension against uncontrolled and irregular contractions of the external sphincter. This creates a reflux, most likely to result in hydrophrosis (11,12). In our case, grade III hydrophrosis was due to the dissynergy between the pelvic-pudental nerves associated with the spinal mass.

In the diagnosis of intramedullary dermoid cyts, radiological evaluations have an important role. Direct radiographies may reveal vertebral corpus destruction, enlargement of interpedicular distuces, and vertebra anomalies. Lomber CT may show increased spinal diameter. However, the primary method of diagnosis is contrasted MRI.(13)Contrasted on MRI, the characteristic appearance of a dermoid cyst is hypointense on T1 and hyperintense on T2 with no contrast enhancement, as was in our case.

In differential diagnosis of spinal dermoid cysts, all spinal cord tumors should be suspected. They should particularly be distinguished from intramedullary located epidermoid cysts, ependimoma, astrocytoma, melanoma, lymphoma and metastases (5,7). Furthermore, their differential diagnosis from demyelinized diseases, inflammatory myelites, paraneoplastic myelopathies presenting with sensory-motor and sphincter deficits should be well-established.

There are two goals in the treatment of spinal dermoid cyts: the treatment aiming at the sphincter deficit and the treatment of the mass. The former involves the use of pharmacological medications, which are anticolinergics, beta-adrenergic antagonists, alpha-adrenergic antagonists, for sphincter deficit. These agents show their effects by increasing the bladder capacity and decreasing the contraction force of the bladder at the same time (11). The studies on the treatment of the mass indicated that the best approach was the total excision of the mass at an early period (10,11). When the diagnosis is established at an earlier time, and the mass is totally excised, neurological and sphincter deficits have improved (4,5). Total mass excision is possible for extramedullary dermoid cysts; however, in intramedullary dermoid cysts, the capsule adheres to the cord, as was in our case; thus, total excision may inflict additional neurological deficits. Long-term case analyses have shown that total excision of the mass including the capsule could lead to fibrous adhesions on the spinal cord (10,11). Therefore, in the surgical interventions of spinal dermoid cysts, subtotal excision of the capsule is preferred. Literature reveals only one case of spinal dermoid cyst, which recurred 17 years later and surgically intervention was required (13,14). The cyst of our case has not recurred within the four-year-follow up period.

Table 1. *The reasons of neurogenic bladder and their clinical signs.*

Disturbance	Pathological entitiy	Anatomic site of lesion	Bladder tone	Bladder capacity	Complications
Cortical uninhibition	Brain atrophy, tumor, strokes, cerebral arteriosklerozis	Frontal lobe	Normal	Normal	Uncontorolled micturition
Spinal reflex bladder (neurogenic or automatic bladder)	Spinal cord trauma,tumor, multiple sclerosis	Spinal cord above S1	Spastic	Small	Infection(Hydronephrosis if dissynergic bladder develops)
Denervated autonomous bladder	Conus lesions, cauda equina lesions, lesions in pelvis	Sacral bladder center (S2 to S4) and its afferent and efferent connection to bladder	Flaccid	Very large	Infection(Hydronephrosis if dissynergic bladder develops)
Dissynergic bladder	All spinal cord	Dissynergic of pelvic-pudental nerves	Spastic-flaccid	Small-large	Infection, hydronephrosis

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