Case Report

Myelomeningocele and Urinary Lithiasis - Case Report

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To cite this article:

Received: June 1, 2016; Accepted: August 25, 2016; Published: December 17, 2016

Abstract: Myelomeningocele is a congenital malformation of central nervous system associated with urinary tract anomalies in several cases. This case report describes a 19 years old, female patient, admitted in Santa Casa de Belo Horizonte Hospital where she was previously diagnosed with lumbar myelomeningocele associated, lately, with several kidneys and bladder dysfunctions, including bladder lithiasis, evaluated by laboratory findings, images, and specific renal depuration tests such as DTPA (diethylenetriaminepentaacetic acid labeled with technetium 99 m) renal scintigraphy. After nephrectomy and antibiotic treatment, the patient had an improvement of her clinical symptoms.

Keywords: Myelomeningocele, Myelomeningocele, Lithiasis

1. Introduction

Myelomeningocele is the most complex of all congenital malformations of central nervous system compatible with prolonged survival, involving abnormalities of spine, brain, peripheral nerves and osteo-articular system. Contrary to what one might suppose, large numbers of patients have normal IQ or close to normal.¹, ²

Urinary tract anomalies associated occurs in about 7% of patients with myelomeningocele, therefore, patients with low-pressure losses, associated with hydronephrosis, especially if unilateral, should be investigated for presence of other urinary tract obstruction causes.³, ⁴

2. Objective

The report provides a description of a patient admitted to urology service of Santa Casa de Belo Horizonte Hospital, in adulthood with serious complications of urinary tract resulting from medullary stretch and change of vesico-sphincter behavior, common in first 2 years of life, but it went unnoticed and was not treated at right time.

3. Method

Literature review on myelomeningocele was performed in Pubmed, Scielo and LiLACS with following terms: “myelomeningocele”, “mielomeningoceles”, “mielomeningocele e litíase”, and “myelomeningocele and lithiasis”. Articles of most relevance were selected and reviewed, corresponding period of years between 1986 and 2016. Description of the case was carried out and for the bibliographic citation was used Endnote X7 for Mac software. We followed all research rules according to current legislation.

4. Case Report

It is female patient, 19, with lumbar myelomeningocele at
birth, and with nine years old soon progressing to bladder dysfunction signs - wall hypertrophy, sphincter dyssynergism and vesicoureteral reflux. At 11 years old, the patient underwent bladder augmentation surgery for improvement in bladder symptoms due to low capacity and complacency bladder.

Since then, the patient lost clinical follow-up with a urologist and developed chronic renal failure predialysis, diagnosed six months ago by nephrology team at Santa Casa de Belo Horizonte Hospital.

The case was referred to urology service for clinical follow-up. During the propaedeutic investigation, she was diagnosed bladder lithiasis and left staghorn calculus, by kidneys and urinary tract sonographic.

CT examination confirmed the sonographic diagnosis of bladder lithiasis and left nephrolithiasis, plus important right pyelocalical dilation.

Voiding cystourethrography confirmed bilateral vesicoureteral reflux degree V, low capacity bladder (100ml), with diffuse walls hypertrophy, multiple bladder diverticula; with bladder augmentation positioned at bladder’s bottom, functioning and no leaks.

DTPA renal scintigraphy revealed renal uptake of 2% left and 1% right, with significant deficiency perfusion and excretion of radiopharmaceutical, setting clinical condition compatible with bilateral renal exclusion.

Pre-operative laboratory tests revealed creatinine 6.3 mg / dL; urea 98 mg / dL; Haemoglobin 11.4 g / dL and total leukocyte 10400 / mm³.

After proper anesthetic evaluation, the patient underwent left nephrectomy by classic lumbotomy and cistolitotomy by Pfannenstiel incision. The patient received antibiotic prophylaxis with cefazolin 2 g and anesthesia, was general combined with epidural. Surgical procedure lasted about 3 hours and was uneventful.

In the first DO (the day after surgery) patient developed polyuria (urine output of 3500ml / 24h) with the improvement of renal function.

The patient was discharged from the ICU in 2nd DO day and discharged on seventh DO with stable renal function (creatinine 2.8 mg / dL and urea 62 mg / dL), diuresis of 1000ml, with intermittent catheterization, antibiotic and anticholinergic.

Was scheduled patients’ follow-up, with urodynamic evaluation and new renal scintigraphy.

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The case attracted attention from urology, nephrology, and intensive care teams, whereas a patient with bilateral renal exclusion evidenced by scintigraphy, improved dramatically diuresis and creatinine levels after unilateral nephrectomy.

5. Discussion

The case reported above shows an example of a tragic outcome for urinary tract secondary to damage caused by neurological disease.

It is known that clinical management of patients with congenital myelomeningocele is essential because there is a risk of 40-60% urinary tract deterioration over a period of five years if not properly treated. Therefore, diagnosis and early treatment of these patients urinary complications reduce need for bladder augmentation surgery.

Many cases of myelomeningocele are treated during childhood, which unfortunately was not the case of our patient. Plenty of times, late diagnosis leads to complications throughout life, bringing problems to patients.

The improvements in the management of newborns with myelomeningocele have obtained an increase in survival, allowing them to get longevity, but data regarding urologic diseases during adult age are still missing.

Urinary lithiasis is common in adults with myelomeningocele, but in our work we found a patient presenting atypical clinical features, which justifies the publishing of this paper to disseminate scientific knowledge.

Many techniques are available to close a myelomeningocele, but large lesions can be particularly difficult to operate, new surgical techniques are still being tested.

Children with myelomeningocele develop a wide variety of deformities, which needs diagnosis and treatment at the right time, preferably as early as possible.

The report also shows a certain unusual in literature: patients with bilateral renal exclusion, no dialytic, progressing with the functional improvement of remnant kidney after contralateral excluded kidney nephrectomy and complex staghorn calculi inside.

6. Conclusion

We did not find in literature review any scientific reference that justifies the fact, however, experts opinions converge on the idea that there are proteins (cytokines) functioning as signaling in excluded kidney with stones which act interfering or modulating negatively on contralateral renal function through downregulation mechanisms on afferent arteriolar dilatation.

Conflict of Interest

The author declares no conflict of interest.

References


