

CASE REPORT

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Findings of MRI, CT scan and ultrasound in Marion's disease in a young woman: a case report

Shima Ghafourian Noroozi¹ and Mohsen Ghazi Soltani^{1*}

Abstract

Background Primary bladder neck obstruction (PBNO) is a rare condition of the lower urinary tract in young and middle-aged patients. PBNO is a urological condition affecting both sexes in which the bladder neck fails to open adequately during voiding, resulting in obstruction of urinary flow in the absence of anatomic obstruction, such as benign prostatic hypertrophy in men or genitourinary prolapse in women. PBNO may present voiding symptoms (decreased force of stream, hesitancy, intermittent stream, incomplete emptying) or irritative symptoms (frequency, urgency, urge incontinence, nocturia) or a combination of both.

Case presentation A 31-year-old female without medical history presented to our department with a 4–5 months history of a sense of incomplete emptying bladder and suprapubic discomfort. In Abdominal and Pelvic CT images of her CT scan, evidence of homogenous concentric hypertrophy of the urethra with mild enhancement in the post-contrast image was seen. Also, pelvic MRI shows isointense on T1-weighted and isointense to mildly hyperintense on T2-weighted images (to skeletal muscle signal) of the concentric hypertrophy muscular layer of the urethra. The post-contrast image demonstrates moderate enhancement in the hypertrophied urethra. A month later, while the patient had not received any treatment, a pelvic ultrasound showed concentric hypertrophy hypoechoic muscular layer of the urethra.

Discussion The true prevalence of PBNO is unknown but Farrar et al. reported a 2% prevalence of primary bladder neck obstruction. In 1933, Marion described PBNO as caused by fibrous narrowing of the bladder neck and detrusor hyperplasia. Also, Marion described two types of disease (congenital and acquired) and treatment methods. In this study was shown isointense on T1-weighted and isointense to mildly hyperintense on T2-weighted images of concentric hypertrophy muscular layer of urethra with moderate enhancement on the post-contrast image. A doughnut sign was also seen in our axial slices (particularly in T2-weighted images), so this sign seems to be a diagnostic finding in MRI. In our study, the diffusion sequence and ADC map showed no obvious restriction.

Conclusions Although PBNO has been described for decades, there is still a controversy about diagnosis and treatment. In patients suspected of PBNO, imaging methods, especially MRI, can help confirm the diagnosis or rule out other causes of bladder neck obstruction. In future studies, imaging modalities can be one of the diagnostic criteria for PBNO.

Keywords Marion's disease, PBNO, Spiral computed tomography, MRI, Ultrasound

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Background

Primary bladder neck obstruction (PBNO) is a rare condition of the lower urinary tract in young and middle-aged patients. Marion G., a French urologist and surgeon, was the first to describe the primary hypertrophy of the bladder neck muscle in 1933. [1] He described it as "the dysuric disturbance similar to those giving rise to hypertrophy of the prostate, disturbance caused by alteration of the bladder neck without any visible lesions of the neck and which cannot be attributed to nerve injuries"[1].

PBNO is a urological condition affecting both sexes in which the bladder neck fails to open adequately during voiding, resulting in obstruction of urinary flow in the absence of anatomic obstruction, such as benign prostatic hypertrophy in men or genitourinary prolapse in women. [2] PBNO is commonly described in men [3–7], but there are fewer studies of this condition in women [8–10]. In 1975, Farrar et al. reported PBNO for the first time in women based on poor urodynamic flow characteristics in the absence of any clear anatomical obstruction [8]. PBNO may present voiding symptoms (decreased force of stream, hesitancy, intermittent stream, incomplete emptying) or irritative symptoms (frequency, urgency, urge incontinence, nocturia) or a combination of both [11, 12]. Here, we present a case of Marion's disease in a young woman with bladder neck obstruction.

Case presentation

A 31-year-old female with no previous illness presented to our department with a 4–5 month history of a sense of incomplete emptying bladder and suprapubic discomfort. In the patient's medical history, a prostate-like lesion was reported under the bladder. The patient has no family or psycho-social history. Also, no significant physical examination or clinical findings were found.

She underwent dynamic pelvic magnetic resonance imaging (MRI) with contrast in our hospital. A spiral Abdominal and Pelvic CT scan with contrast was performed for the patient in another center. One month later for this study and follow-up, a pelvic ultrasound was performed with a semi-full bladder. Patient preparation for bladder ultrasound (using a curved transducer with frequency of 5 to 2 MHz) was that the bladder should be full. One hour before the study, the patient was given 500 to 1000 ml (2 to 4 cups) of water until the bladder was full completely but due to the presence of symptoms, the patient could not fill the bladder, so the ultrasound was performed with semi-filled bladder. MRI with 1.5 T magnetic field equipment was performed. The MRI

multiplanar sequences we took included: T1-weighted without and with SPIR, T2-weighted without fat suppression pre-contrast sequences, axial diffusion-weighted images (DWI), exponential map axial apparent diffusion coefficient (exponential ADC) map and T1-weighted fat suppressed images after the administration of intravenous gadolinium.

In abdominopelvic images of her CT scan, evidence of homogenous concentric hypertrophy of the urethra with mild enhancement in the post-contrast image was seen (Fig. 1).

Also, pelvic MRI shows isointense on T1-weighted and isointense to mildly hyperintense on T2-weighted Images (to skeletal muscle signal) of the concentric hypertrophy muscular layer of the urethra (Fig. 2). The post-contrast image demonstrates moderate enhancement in the hypertrophied urethra (Fig. 2). The diffusion sequence and exponential ADC map showed no obvious restriction in the hypertrophied muscle layer (Fig. 3).

A month later, while the patient had not received any treatment, a pelvic ultrasound showed concentric hypertrophy hypoechoic muscular layer of the urethra (Fig. 4). The patient was referred for further work-up to the urologist.

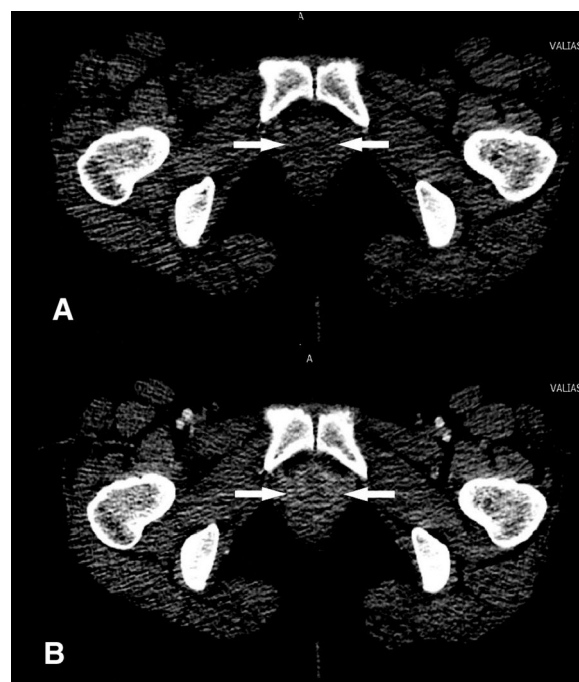
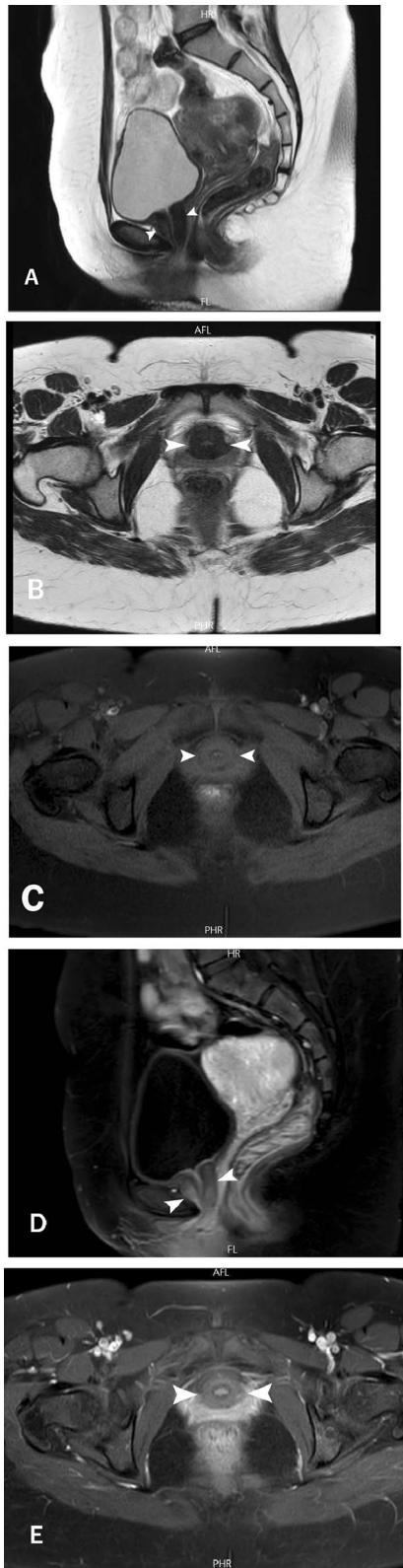


Fig. 1 Pelvic axial CT scan without **A** and with contrast **B** shows homogenous concentric hypertrophy of the urethra with mild enhancement (arrows)



◀ **Fig. 2** Sagittal **A** and axial **B** non-fat suppression T2-weighted images and axial **C** fat suppression pre-contrast T1-weighted images of pelvic MRI shows isointense on T1-weighted and isointense to mildly hyperintense on T2-weighted Images of the concentric hypertrophy muscular layer of the urethra (arrowheads). Sagittal **D** and axial **E** planes contrast-enhanced fat suppression T1-weighted Images demonstrate moderate enhancement in the hypertrophied urethra (arrowheads)

Discussion

The true prevalence of PBNO is unknown but Farrar et al. reported a 2% prevalence of primary bladder neck obstruction based on a large series of 169 women who underwent urodynamic studies [8]. The clear pathophysiology of PBNO is unknown, also. In 1933, Marion described PBNO as caused by fibrous narrowing of the bladder neck and detrusor hyperplasia. Also, Marion described two types of disease (congenital and acquired) and treatment methods [1]. Also later studies, several pathophysiology's have been described such as: accumulation of abnormal amounts of non-muscular connective tissue in hypertrophic smooth muscle and inflammatory changes [13], incompetency bladder neck opening [3], functional bladder neck disorder [14], and increased sympathetic contractile system in the urethra [15]. Currently, the diagnosis of PBNO is by VUDS (video urodynamic study) characterized by relative high-pressure, low-flow voiding with radiographic evidence of obstruction at the bladder neck and no evidence of distal obstruction [16–19].

MRI and ultrasound findings of Marion's disease are not well described. In our study was shown isointense on T1-weighted and isointense to mildly hyperintense on T2-weighted Images (to skeletal muscle signal) of concentric hypertrophy muscular layer of urethra with moderate enhancement on the post-contrast image. In 2012, Ernesto Lima et al. presented isointense T1-weighted and slightly hyperintense T2-weighted images of concentric muscle hypertrophy of the entire urethra with characteristic finding bulb of a sphygmomanometer on the sagittal and coronal, and a doughnut sign on the axial planes from Marion's disease [20]. A doughnut sign was also seen in our axial slices (particularly in T2-weighted images), so this sign seems to be a diagnostic finding in MRI. In our study, the diffusion sequence and ADC map showed no obvious restriction, while Ernesto Lima et al. observed Diffusion restriction in the hypertrophied muscle layer on the ADC map [20].

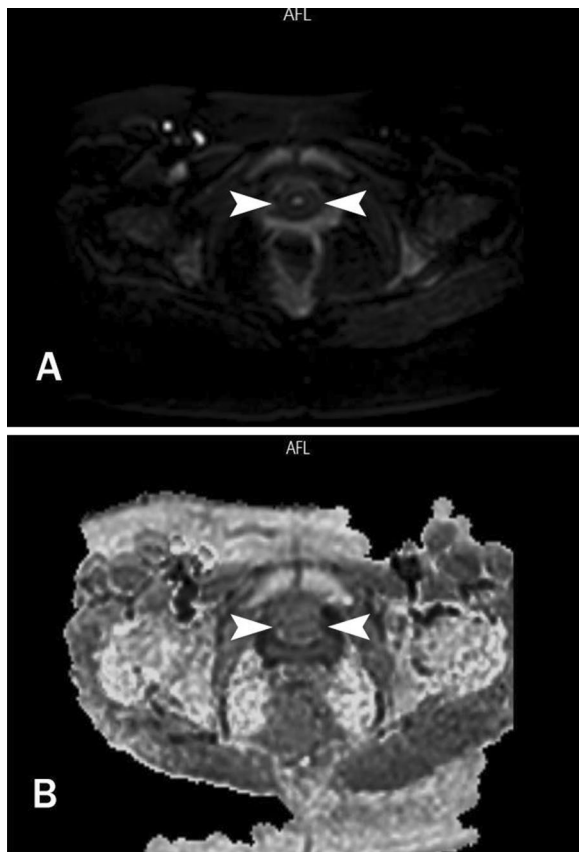


Fig. 3 The axial diffusion sequence **A** and axial exponential ADC map **B** showed no obvious restriction in the hypertrophied muscle layer (arrowheads)

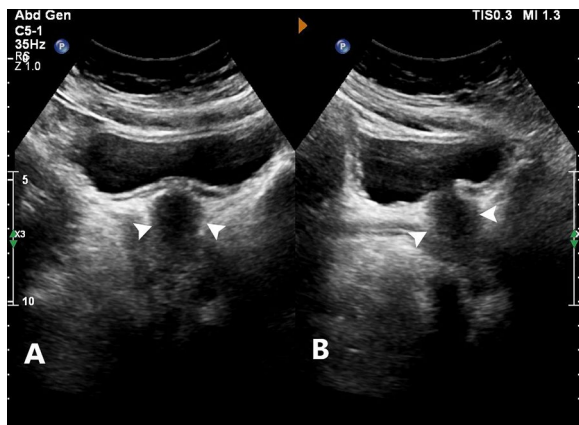


Fig. 4 Pelvic TAS Transverse **A** and sagittal **B** images show concentric hypertrophy hypoechoic muscular layer of the urethra (arrowheads)

The increasing use of dynamic pelvic MRI can provide new imaging findings for the diagnosis of PBNO. Unfortunately, one of the most important limitations of our study was the unavailability of VUDS (video urodynamic study) to confirm our diagnosis.

Conclusions

Although PBNO has been described for decades, there is still a controversy about diagnosis and treatment. Also, its findings are not well described in radiology modalities. In patients suspected of PBNO, imaging methods, especially MRI, can help confirm the diagnosis or rule out other causes of bladder neck obstruction. More studies and research are needed for a more detailed of PBNO findings in imaging modalities, especially MRI. It is important to notice and report these findings in clinical imaging. In future studies, imaging modalities can be one of the diagnostic criteria for PBNO and help its treatment.

Abbreviations

MRI	Magnetic resonance imaging
CT	Computed tomography
PBNO	Primary bladder neck obstruction
TAS	Trans-abdominal sonography
ADC	Apparent diffusion coefficient
DWI	Diffusion-weighted imaging
VUDS	Video urodynamic study

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Not applicable.

Author contributions

ShGN chose the case, performed the ultrasound and reported the CT scan and ultrasound and supervised manuscript preparation, and edited the case report. MGHs interviewed the patient, reviewed the published literature, and wrote the case report manuscript. The authors read and approved the final manuscript.

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Availability of data and materials

The datasets generated and/or analyzed during the current study are available from corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate

The methods and the reasons for the research were explained to the patient in simple and understandable language. It was explained that the information (the text and any pictures or videos) is published without attaching the name, but its complete anonymity cannot be guaranteed. It was also explained that information (the text and any pictures or videos) will be freely available on the internet and may be seen by the general public her anonymized. It was also explained that this research does not any physical, mental, or economic harm and even helps to treat the disease better and will also help patients who are similar to her in the future. It was offered to read the manuscript and inform us with informed consent. Written informed consent was obtained from the patient to use and publish her anonymized medical records. In consulting with the ethics committee of Rasoul-e-Akram Hospital approved the anonymous use of patient data.

Consent for publication

The methods and the reasons for the research were explained to the patient in simple and understandable language. It was explained that the information (the text and any pictures or videos) is published without attaching the name, but its complete anonymity cannot be guaranteed. It was also explained that information (the text and any pictures or videos) will be freely available on the

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Competing interests

The authors declare that they have no competing interests.

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