Case 14697

Eurorad

Para-bladder mass in symptomatic 16-year-old female patient

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Section: Uroradiology & genital male imaging
Area of Interest: Abdomen
Procedure: Surgery
Imaging Technique: MR
Imaging Technique: MR-Diffusion/Perfusion
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 16 years, female

Clinical History:

A 16-year-old female patient presented at the emergency room for pain in the left iliac fossa, exacerbate by effort, without dysuria. The clinical examination revealed pain in the left iliac fossa and arterial hypertension. Urinary stick was negative. FAST ultrasound revealed left lithiasis. During cystoscopy, a parietal protuberance was noted.
Imaging Findings:

MRI revealed urinary bladder characterized by an expansive solid lesion, with irregular borders, which involved the left posterior-lateral wall, partially exophytic and protruding into the lumen, with major diameters: 4.4 x 3 centimetres. The lesion showed intermediate and inhomogeneous MR signal on T2- and T1-weighted images and was heavily hyperintense in DWI with restricted diffusion at ADC. Neither necrotic, haemorrhagic nor fat-content areas appeared. In the post-contrastographic images the mass appeared markedly hyperintense at the arterial, venous, and equilibrium phase. The left ureteral ostium was not dissociable, but the urography sequences didn't show hydroureteronephrosis. The lesion appeared in close contiguity with the left lateral vaginal fornix, without clear cleavage fat.

MRI was performed with the following sequences: T1 and T2 FSE, with and without fat saturation, DWI (b=0, 800), LAVA pre- and post-injection of GBCA.
Discussion:

Primary paraganglioma (or extra-adrenal pheochromocytoma) of the bladder is a very rare tumour, with less than 250 cases described in literature [1]. It tends to affect females, with a very wide mean range of age (11-84 years) [2]. This is one of the few cases described in patients less than 18 years of age, and one of the biggest, considering the case series published by Henderson and colleagues [1].
The classical clinical presentation of bladder paragangliomas includes: hypertension, gross haematuria, and postmicturition syndrome [1]. Our case presented with low abdominal pain, ureteral lithiasis and hypertension.
In the MRI evaluation of bladder masses, the most important elements that must be considered are: the location in relation to the wall layers, the MRI signal, and the vascularization.
The bladder tumours can originate from each of the wall layer: epithelium, lamina propria, and adventitia serosa. They can be classified as epithelial and non-epithelial [3].
Among the epithelial ones, the great majority are of urothelial origin; other less common are squamous cell carcinoma and adenocarcinoma. Epithelial tumours arise as intraluminal filling defects, irregular morphology, while

those mesenchymal present themselves as intramural lesions in the smooth and regular margins.
The mesenchymal tumours originate from non-epithelial layers of the bladder wall and they represent 5% of bladder tumours. The paraganglioma is a mesenchymal tumour and the differential diagnosis should be made with other mesenchymal tumours, most commonly encountered in the bladder [4]: especially rhabdomyosarcoma and neurofibroma, less common leiomyosarcoma. Rhabdomyosarcoma is the most common bladder cancer in childhood with greater occurrence in males. It manifests as polypoid and heterogeneous mass at the base of the bladder. The morphology and locally aggressiveness help to distinguish it from paraganglioma.
The neurofibroma is rare and may occur as isolated form or associated with neurofibromatosis type I. It originates from intramural nerve plexus near the bladder trigone. Even angioma must be considered in the differential diagnosis in paediatric age. The angioma manifests as a small single lesion, hypervascular, more commonly in the postero-lateral walls. Benign or malignant tumours to originate from adjacent organs (prostate, uterus, ovary) can mimic bladder tumours [5].
As described in literature, the pre-operative diagnosis of these tumours is very difficult, especially in the absence of typical symptoms. The mesenchymal origin can be suspected in hypervascular submucosal masses covered by a layer of intact urothelium, in association with appropriate clinical assessment.
Differential Diagnosis List: Primary paraganglioma of the urinary bladder., Rabdomyosarcoma, Leiomyosarcoma, Neurofibroma, Angioma, Ab extrinseco compression

Final Diagnosis: Primary paraganglioma of the urinary bladder.

References:

Henderson SJ (2015) Patients with urinary bladder paragangliomas: a compiled case series from a literature review for clinical management. Urology Apr;85(4):e25-9 (PMID: 25618559)
Kouba E (2016) Neuroendocrine Tumors of the Urinary Bladder According to the 2016 World Health Organization Classification: Molecular and Clinical Characteristics. Endocr Pathol Sep;27(3):188-99 (PMID: 27334654)
Klaile Y (2016) Variant histology in bladder cancer: how it should change the management in non-muscle invasive and muscle invasive disease?. Transl Androl Urol Oct;5(5):692-701 (PMID: 5071184)
Cheng L (2012) Bladder Pathology.
Loveys FW (2015) Urinary bladder paraganglioma: AIRP best cases in radiologic-pathological correlation. RadioGraphics Sep-Oct; 35 (5): 1433-8 (PMID: 26284302)

Figure 1

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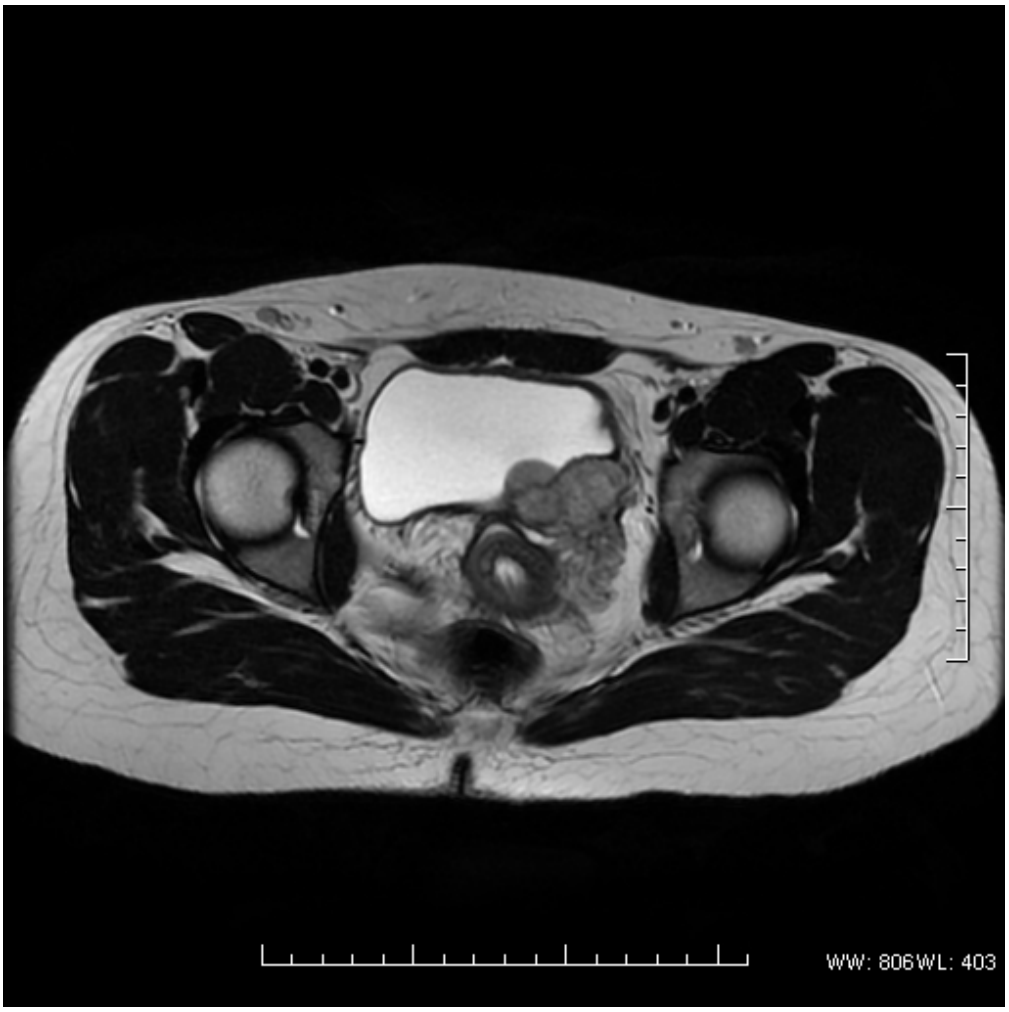

Description: Axial T2w TSE **Origin:** Department of Radiology, Ospedale Galliera, Genova

Figure 2

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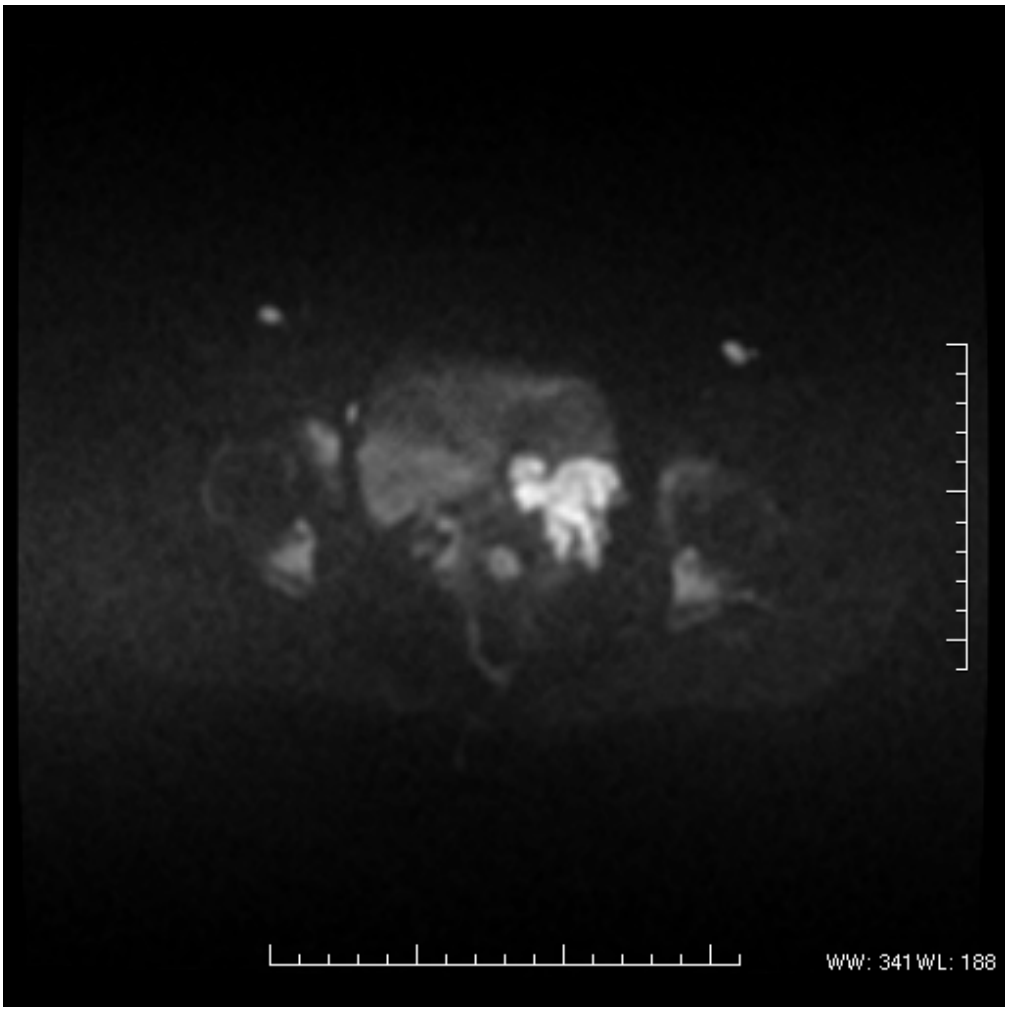

Description: Axial DWI (b = 800) **Origin:** Department of Radiology, Ospedale Galliera, Genova

Figure 3

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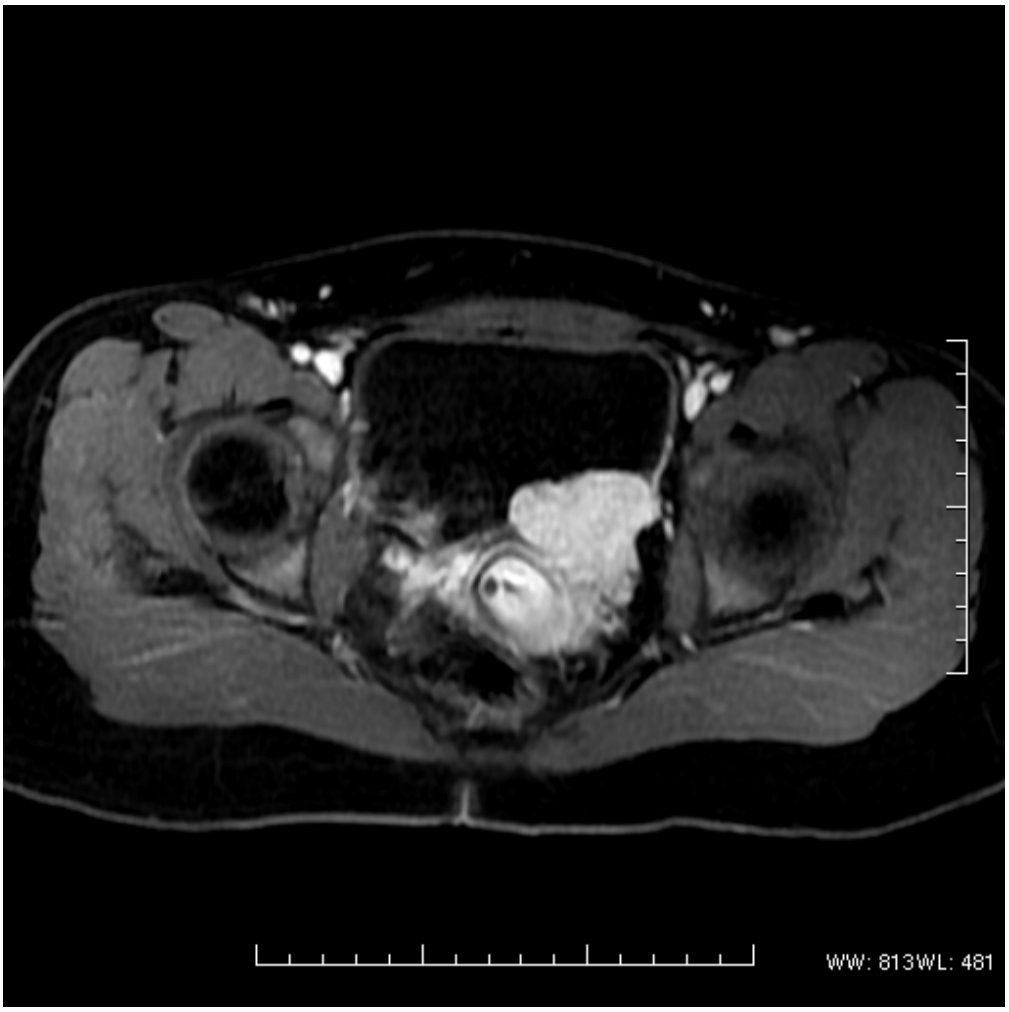

Description: Axial LAVA Post-contrast **Origin:** Department of Radiology, Ospedale Galliera, Genoa

Figure 4

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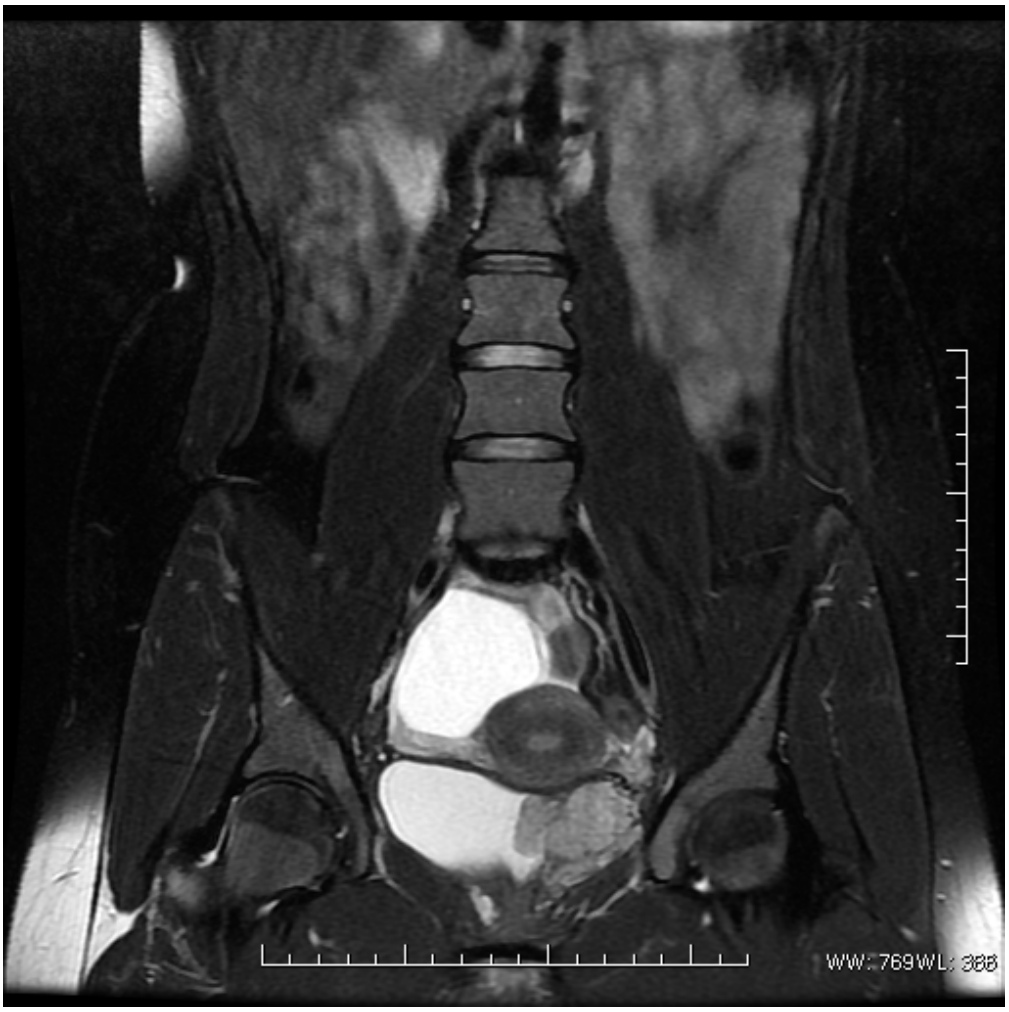

Description: Coronal T2w TSE **Origin:** Department of Radiology, Ospedale Galliera, Genova

Figure 5

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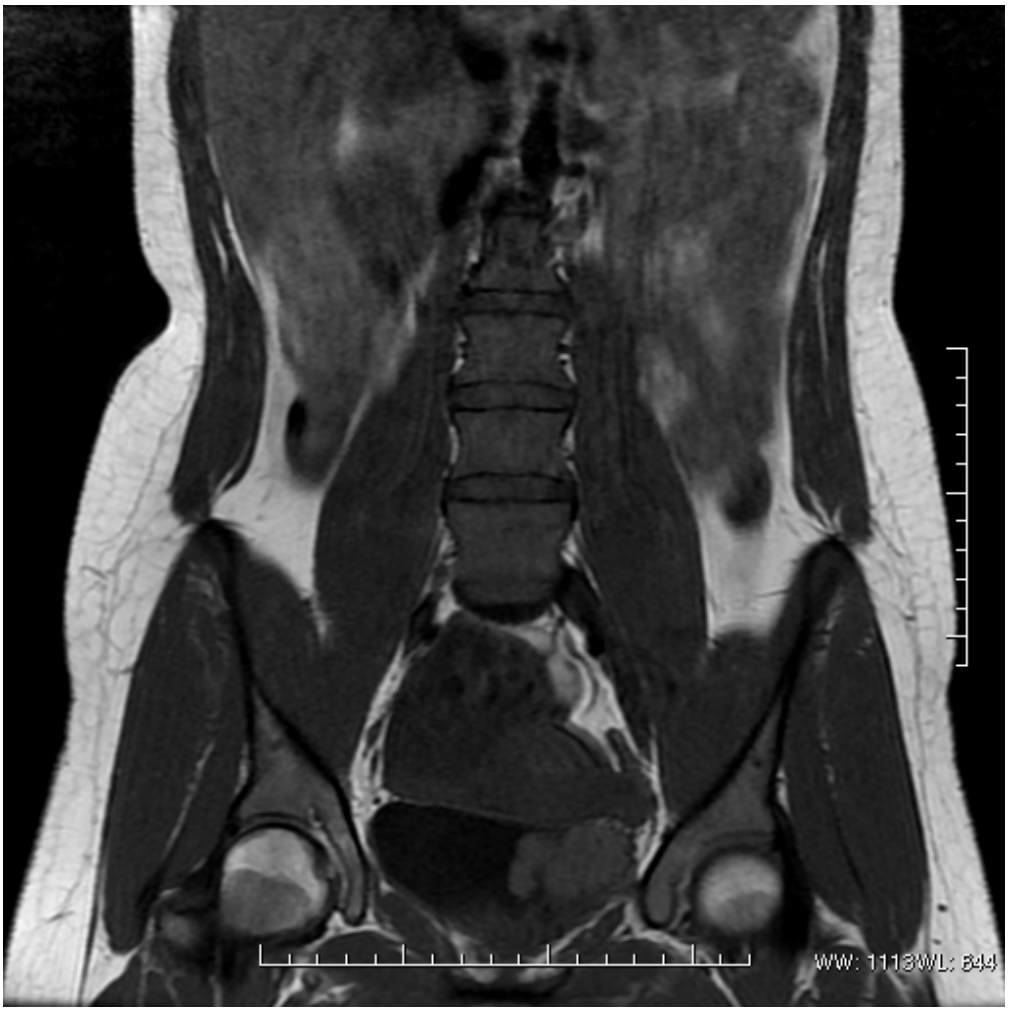

Description: Coronal T1w TSE **Origin:** Department of Radiology, Ospedale Galliera, Genova