EDITORIAL COMMENT BY…

Commentary to “‘Urinary disorders of Wolfram syndrome. Clinical and urodynamic analysis from 6 observations’” from: C. Leroux, M. Grasland, N. Turmel, F. Le Breton, C. Chesnel, C. Hentzen, G. Amarenco; report of six cases in two wolfram syndrome siblings

Commentaire à « Les troubles vesico-sphinctériens du syndrome de Wolfram. Analyse clinique et urodynamique à partir de 6 observations » de C. Leroux, M. Grasland, N. Turmel, F. Le Breton, C. Chesnel, C. Hentzen, G. Amarenco ; à propos de 6 cas dans deux fratries atteintes du syndrome de Wolfram

I. Boualaoui*, I. Ziani, O. Bellouki, H. El Sayegh, L. Benslimane, Y. Nouini

Department of urology A, Mohammed V university in Rabat, Ibn Sina university hospital, Rabat, Morocco

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To the editors,

We read with a great interest the article entitled “‘Urinary disorders of Wolfram syndrome. Clinical and urodynamic analysis from 6 observations’” from: C. Leroux, M. Grasland, N. Turmel, F. Le Breton, C. Chesnel, C. Hentzen, G. Amarenco, published in November 2019. Their study showed that urinary dysfunction in WOLFRAM syndrome is mainly characterized by overactive bladder and urinary retention. We would like to contribute some points of their paper.

The authors discussed the main lower urinary tract disorders in WOLFRAM syndrome. All their patients suffered from overactive bladder and urodynamic findings showed detrusor overactivity in 50% of their patients [1]. In our department, we also treated 6 cases of WOLFRAM syndrome in two...
Moroccan families, from 2000 to 2019, during ten years of minimal follow-up. The age of diagnosed cases at the first urologic consultation is $17 \pm 5$ years. Parent’s propinquity, bilateral optic nerve atrophy and sensorineural hearing loss were noticed among all these cases. Three patients were diagnosed with diabetes insipidus. Like Leroux et al., five of our patients (83%) suffered from dysuria and a clinical overactive bladder, however, we have not noticed any detrusor overactivity in the non-invasive urodynamic test (NIUD). The only patient who has no NIUD is eighteen years old. She has been treated at the age of four by a surgical vescostomy to manage a urinary retention. Due to social and economical issues, no medical consultation has been done since the paediatric surgery. The limited medical access in some far areas, explains that the most common revealing mode of the diagnosis is the complications of renal failure. It was the case of four of our patients (66%) (Table 1).

In addition, to the therapeutic issues discussed in the study, there are two additional challenging factors which complicate the treatment of lower urinary tract manifestations in our wolfram syndrome patients. Firstly, a perfect control of urinary output may have a significant impact on limiting bladder damage. Indeed, bladder dysfunction has been not only attributed to the autonomic and degenerative damage affecting the central and peripheral nerve system but also to polyuria stretch injury due to the high urinary output of diabetes mellitus and diabetes insipidus. Furthermore, the use of desmopressin has a double interest, on the one hand, it reduces the urinary output and helps in the resolution of incontinence. On the other hand, it reduces the progression of bladder dysfunction and megacystis [2]. Secondly, the upper urinary tract distension seen in 50% of Leroux et al. study and 100% of our patients, may persist after the treatment of diabetes insipidus by desmopressin and lower urinary tract symptoms by clean intermittent self-catheterization, anticholinergics or intra-detrusor injection of botulinum toxin [1]. This fact suggests that there is an own upper urinary tract damage when the neurodegenerative process reaches ureteric and pelvic innervation [3].

<table>
<thead>
<tr>
<th>Cases</th>
<th>Age at the diagnosis/gender</th>
<th>Lower urinary tract disorders</th>
<th>NIUD</th>
<th>Upper urinary tract disorders</th>
<th>Renal function after 10 years follow-up DFG (mL/min)</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case 1</td>
<td>12/man</td>
<td>OAB + dysuria</td>
<td>Normal</td>
<td>Minimal pelvic distension</td>
<td>60</td>
<td>Desmopressin</td>
</tr>
<tr>
<td>Case 2</td>
<td>14/man</td>
<td>OAB + dysuria</td>
<td>Normal</td>
<td>Bilateral ureteropelvic distension</td>
<td>60</td>
<td>Clean intermittent self-catheterization Desmopressin</td>
</tr>
<tr>
<td>Case 3</td>
<td>16/man</td>
<td>OAB</td>
<td>Normal</td>
<td>Bilateral ureteropelvic distension by vesicoureteral reflux</td>
<td>30</td>
<td>CIC Surgical urinary diversion after failed CIC and bilateral ureteral reimplantation Desmopressin</td>
</tr>
<tr>
<td>Case 4</td>
<td>18/woman</td>
<td>Urinary retention treated at the age of four by surgical vescostomy</td>
<td>Not available</td>
<td>Minimal pelvic distension</td>
<td>80</td>
<td>Surgical closure of vescostomy Anticholinergics CIC Desmopressin</td>
</tr>
<tr>
<td>Case 5</td>
<td>20/woman</td>
<td>OAB</td>
<td>Normal</td>
<td>Bilateral pelvic distension</td>
<td>35</td>
<td>Anticholinergics CIC Desmopressin</td>
</tr>
<tr>
<td>Case 6</td>
<td>22/woman</td>
<td>OAB + urge incontinence</td>
<td>Normal</td>
<td>Minimal pelvic distension</td>
<td>70</td>
<td>Anticholinergics CIC</td>
</tr>
</tbody>
</table>

OAB: overactive bladder; CIC: clean intermittent self-catheterization; NIUD: non-invasive urodynamic test.
Urological manifestations might not be obvious at the time of diagnosis unless the first urological work up. Careful urological follow-up and close monitoring are the key points enabling to avoid consequent disorders.

Disclosure of interest

The authors declare that they have no competing interest.

References

