

Idiopathic Retroperitoneal Fibrosis Mimicking a Pelvic Tumor: a Case of Pericystitis Plastica

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Retroperitoneal fibrosis was first described in 1905 by Albarran, a French urologist, who performed ureterolysis for ureteral compression produced by the disease. However, this disease became an established clinical entity by Ormond's account in the English literature in 1948. Pericystitis plastica has been used to define an extremely rare type of Idiopathic retroperitoneal fibrosis (IRF) constricting the bladder. In this study, we discussed the recovery of 29-year-old woman with pericystitis plastica who was misdiagnosed as pelvic malignancy or a chronic/subacute pelvic inflammation at the first evaluation.

Key Words: Idiopathic retroperitoneal fibrosis, bladder, pericystitis plastica, pelvic tumor

INTRODUCTION

Idiopathic retroperitoneal fibrosis (Ormond disease) is a nonspecific, nonpurative inflammation of fibroadipose tissue covering the retroperitoneal structures with mostly unknown etiology. However the disease can be originated from unusual locations including pelvis and mediastaneum. The term of pericystitis plastica (or plastic pericystitis) has been used to define an extremely rare type of the IRF constricting the bladder.¹ In this case we have discussed an IRF of the bladder in term of fulminant pericystitis plastica misdiagnosed as malign pelvic tumor or a chronic infection at the first evaluation.

CASE REPORT

A 29-year-old married thin habitus woman with 4 children, presented with nonspecific abdominal pain and urgency for a few months. Medical history was insignificant. Clinical findings were normal except anterior firmness in the rectal examination and no weight loss and fever were reported. Gynecological examination and Pap smear tests were normal. Urinalysis and complete blood count were in normal limits, urine culture, tuberculous tests and tumor markers were negative. Erythrocyte sedimentation rate was found as 43 mL/h. While color Doppler Ultrasound revealed no pathology, pelvic Ultrasound showed the thickness of bladder wall and polypoid in the bladder mucosa on the ipsilateral side. Excretory urography was normal apart from a mild right hydronephrosis. An abdominal contrast enhanced computed tomography (CT) confirmed the crescent-shaped irregular bladder wall thickness about 1.5 cm (Fig. 1A) at the right side and an infiltrative mass extending to the adjacent structures and the iliac crest. The rectoscopy, colonoscopy and rectal biopsy did not indicate any pathology. The cystoscopy revealed a normal bladder capacity, and no noticeable tumor mass except disseminated bullose edema like polypoid on the right side of the bladder mucosa. The resection and fine needle biopsies of the bladder wall were interpreted as subacute nonspecific inflammatory reaction.

Due to the confused diagnosis, surgical pelvic exploration was inevitable performed. Laparotomy revealed an appearance frozen-like pelvis in which the external mass on the bladder wall had

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Fig. 1. (A) A preoperative CT reveals the right sided crescent-shaped bladder wall thickening. (B) 2 months postoperatively, CT images; the mass covers all the bladder wall as in the term of pericystitis plastica despite of the 6 weeks empirical antibiotherapy. (C) After three months steroid treatment, the tumor around the bladder had completely disappeared at CT images.

been extended to the periostium of right iliac crest, ovary and to the rectum but no evidence of fibrosis in the upper retroperitoneum. An abscess formation about 1cm in diameter insight of the mass between the right side of the bladder and rectum was also observed. Cultures of this purulent material and the tissue were negative. A resectable parts of the mass excised and distal right ureter also dissected. Histological examination demonstrated infiltration of lymphocytes that forming follicles, eosinophils and plasma cells, but predominantly fibrous tissue consisting of collagen fibrils and fibroblasts invading into the adjunctive adipose tissue with occasional germinal centers (Fig. 2). Patient was discharged with empirical ciprofloxacin and indomethazin therapy for 6 weeks.

Two months later at the control visit the patient had complaints such as nocturia, frequency and urgency. Control CT pointed out the progression of the primary lesion restricting the bladder as in the term of pericystitis plastica and bilateral hydroureteronephrosis (Fig. 1B). Urodynamic

studies showed decreased compliance a little. Internal double-J stents were placed bilaterally and prednisolone treatment was initiated with a dose of 32 mgr/d. due to the diagnosis confirmed as Ormond's disease. After two months with this steroid protocol, the tumor had completely disappeared (Fig. 1C) and thereupon steroid treatment was gradually terminated.

DISCUSSION

Classic IRF may also extend to the pelvis and sometimes mimic a pelvic tumor. A very rare case of disseminated malignant IRF to all systems in a postmortem study was also reported.² To our knowledge, three case reports of pericystitis plastica (or plastic pericystitis) have been published up to 1979 in the literature.^{1,3,4}

In our study radiological evidences mostly pointed out a malignant pelvic diseases like a sarcoma originated from bladder wall but this was not confirmed by the transurethral deep

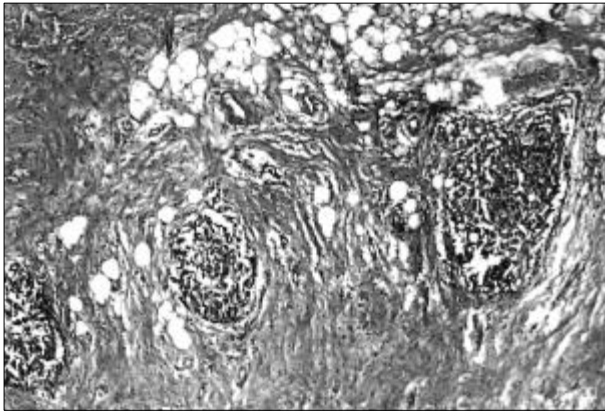


Fig. 2. Histological examination demonstrated infiltration of lymphocytes, eosinophils and plasma cells, but predominantly fibrous tissue consisting of collagen fibrils and fibroblasts invading into the adjunctive adipose tissue with occasional germinal centers. Reduced from $\times 100$.

resection, aspiration and excisional biopsies. A pelvic laparotomy was preferred for the reasons as to get a satisfactory amount of mass for an exact pathologic diagnosis and the tissue cultures, to determine tumor origin, to reduce the mass and to release right distal ureter. Although the pathologic report was pointed out IRF, due to observation of a small abscess formation insight the primary tumor mass in the laparotomy, we got into the difficulty in differentiation IRF from chronic or a subacute pelvic infection that could not shown in the cultures. Thus we decided to give an empirical antibiotics and anti-inflammatory therapy for 6 weeks for a satisfactory cure of either these pathologies. Though antibiotherapy was also a treatment option for IRF besides steroids, it seemed ineffective in our case because 2 months later, at the follow-up, right sided crescent-shaped lesion progressing into a pericystitis plastica surrounding all the bladder at CT images. Then this insufficient therapy was immediately

replaced by 32 mgr/d prednisolone treatment that is commonly a successful protocol for RF in the literature.⁵

Pelvic lipomatosis, another rare proliferative process involving the mature fat of the pelvic organs, possibly cause bilateral hydronephrosis, may be strongly confused with pericystitis plastica.⁶ Unlike in our case, this situation has been seen almost exclusively in males mostly overweight in the third to sixth decades of life and also no dominant fat tissue has been demonstrated in histopathological examination of our case as well. Moreover, specific and nonspecific infections of the bladder and the pelvis were not proved by the cultures and microscopic examinations. To conclude, this is a rare case that the clinical, radiologic, and the pathological evidences suggested a right-sided pelvic retroperitoneal fibrosis progressing into a fulminant pericystitis plastica, mimicking a pelvic malignancy or a chronic infection at the first evaluation.

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