Management of urologic complications of desmoid tumors: Three case reports and literature review

Prise en charge des complications urologiques des tumeurs desmodièes : A propos de trois cas


ABSTRACT
We report three cases of abdominal fibromatosis that caused hydronephrosis by extrinsic ureteral compression in young women with a fruitlessly surgical resection. This ureteral compression was treated by endoscopic placing of double pigtail stents or percutaneous nephrostomy associated with tamoxifen with, apparently, stabilization of the disease. All three patients were regularly monitored with change of the ureteral stents.

Keywords
Desmoid tumour, Complication, urology.

Introduction
Mesenteric or retroperitoneal desmoid tumors are rare fibromatoses, more common in Gardner's syndrome [1]. Mostly asymptomatic, they can, however, be revealed by intestinal, vascular and urologic complications.

We report three cases of ureteral compression by abdominal desmoid tumors whose treatment was different from one case to another.

Patients and Methods
This work involved a retrospective study 2005-2015 of three cases of urological complications of desmoid tumors in urology "B" department IBN SINA University hospital-RABAT.

Our objective is to illustrate urological complications of desmoid tumors while exposing epidemiological, clinical and therapeutic aspects of these tumors.

There were three patients whose respective age was 26, 28, 30 years. Two patients, one of which was monitored for type I diabetes had been operated on for mesenteric desmoid tumor. The third had a history of familial adenomatous polyposis. She underwent a resection of the mass (Figure 1) extended to the right colon and a portion of the last intestine of about 40 cm.

Clinical signs were dominated by abdominal pain, low back pain accompanied by vomiting with slightly impaired renal function in one patient. All patients underwent an ultrasound and an abdominopelvic CT scan that showed respectively bilateral hydroureter without extrinsic compression of the ureter secondary to retro peritoneal fibrosis in two patients, a large peritoneal mass of about 25 cm long axis with tissue density (Figure 2), the left kidney pushed forward and to the right, small intestines plated at back with bilateral pyelocalyceal dilatation in the third patient.

Results
Two patients received palliative treatment by placement of bilateral JJ stents for the first patient and to the left side for the second that were changed every six months and annually, with normal renal function. The evolution was marked 2 years later in the second patient by stenosis of the right ureter with pyelocalyceal dilatation (Figure 3) which was treated with an ileal ureteroplasty. A year later the patient died from complications of her diabetes. After a failed attempt of placement of ureteral double-J stent in the third patient, bilateral nephrostomy was performed. In view to unresectable tumor, surgical biopsy was done with the histological result in favor of desmoid tumor (figure4) and she was treated with tamoxifen for 1 year though proved ineffective and stopped following the occurrence of side effects.

Discussion
Desmoid tumors are rare since they represent less than 0.03% of fibrous tumors [1] and predominate in women. Our subjects were all female and presented surgical history: familial adenomatous polyposis and aggressive fibromatosis. Among our three patients, two showed sporadic desmoid tumors, while for the third; diagnosis was done 3 years after right colectomy for familial adenomatous polyposis (FAP) which is comparable to literature data that reports an average of 2 to 3 years after surgery [3].

Clinical signs and radiology were dominated by tumor syndrome associated with urinary compression symptoms in the three patients. Some authors report that urologic complications are relatively less frequent and most often in rapport with ureteral compression which may be responsible for symptoms or clinically silent.
The evolution of intra-abdominal fibromatosis is responsible for 11% of deaths among patients with familial adenomatous polyposis [1].

Certain clinical observations have suspected the hormone-dependent character of desmoid tumors: they are aggravated during pregnancy and can spontaneously regress at menopause [9].

This phenomenon has been confirmed in vitro [2]: tissue proliferation and synthesis of collagen are estrogen-dependent. Thus tamoxifen which is an anti-estrogen has been proposed to treat these tumors with satisfactory results although too few to draw valid conclusions [1, 4, 6]. Other treatments have shown encouraging results: the case of non-steroidal anti-inflammatory: Sulindac [4]. High-dose radiotherapy (55-60 Grays) has also shown its effectiveness though partial (about 50% recurrences) and very limited indication of doses often incompatible with location [5]. As for chemotherapy, it has never demonstrated its effectiveness. Indeed, complete surgical resection is often impossible in 50% of cases, especially as loco-regional recurrences are frequent (14-75%) [3].

Therapeutic strategy in our series was surgical excision of tumor in two patients, while the third had only surgical biopsy in view to the tumor volume and operative risk. Furthermore, tamoxifen was administered in one patient, but with no favourable result.

From urological aspect, two patients underwent endoscopic treatment namely ureteral endoprosthesis and percutaneous nephrostomy in the third patient.

Evolution was marked by tumor relapse in one of our patients and tumor progression in the third patient.

One patient is regularly monitored in our department: by change of ureteral endoprosthesis every year. The other two died, one from complications of her diabetes and the other by compression of the large vessels by increase in tumor volume.

**Conclusion**

Desmoid tumors are rare tumors, they occur at any age with a marked female predominance. Urological complications of desmoid tumors are relatively infrequent, most often in connection with ureteral compression. There is no consensus on the treatment of desmoid tumors but the recommended first-line treatment currently by most authors is medical, surgical treatment being reserved for complicated forms.

In urological terms, possible therapy comes down to endoscopic treatment namely double J ureteral endoprosthesis or percutaneous nephrostomy. This treatment offers above all the advantage of relieving the patient’s pain and preserve renal function.

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**References**

