LETTER / Genito-urinary

Leiomyosarcoma of the ureter: A rare case

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The majority of tumours of the ureter (approximately 95%) are primitive epithelial tumours, generally transitional cell carcinomas. Leiomyosarcoma is an extremely rare tumour since only about 20 cases have ever been described in the literature [1]. We report a case of leiomyosarcoma of the ureter detected by CT.

Observation

Mrs R., aged 57, had had abdominal pain predominantly in the right lumbar fossa for several weeks. Ultrasound exploration was therefore undertaken and revealed isolated right ureterohydronephrosis. A double J ureteral endoprosthesis was inserted to alleviate the pain. Secondly, a CT urogram was carried out and revealed a well limited and rounded retroperitoneal mass with peripheral enhancement which was in very close contact with the right ureter (Figs. 1 and 2). No other significant abnormality came to light, particularly none of the bladder. Two hypotheses were put forward: either retroperitoneal adenomegaly extrinsically compressing the ureter or an urothelial tumour developing extrinsically. A diagnostic biopsy was performed. From the anatomopathological analysis and more particularly, study of the immunohistochemical markers, it was possible to confirm the diagnosis of leiomyosarcoma of the ureter (Figs. 3 and 4). An additional PET scan confirmed a retroperitoneal right para-ureteral focus of hyperfixation, with no other focus of hyperfixation in the lymph nodes (Fig. 5). Complete surgical exeresis of the lesion was undertaken with confection of a termino-terminal ureteroureteral anastomosis. Postoperative monitoring was simple. Since the patient’s general condition permitted it, adjuvant chemotherapy was started. Early scan checks and at 9 months did not reveal any signs of complication, nor of recurrence.

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Figure 1. Abdominopelvic computed tomography on portal phase in axial section showing a right retroperitoneal mass syndrome [upper pole of the lesion (a) and middle portion (b)] in very close contact with the ureter identified by the J stent (arrow).

Figure 2. Abdominopelvic computed tomography on portal in coronal MIP reconstructions (a) and sagittal (b) focus on the right ureter in its proximal portion. Limited and rounded peritoneal mass with a peripheral enhancement, in close contact with the ureter, whose light is detected by the J stent (arrow).

Discussion

Ureteral leiomyosarcoma is an extremely rare malignant mesenchymal tumour, since only some 20 cases have been described in the literature [1]. The circumstances in which it is mainly detected involve lumbar pain associated with macroscopic haematuria [2]. Its prognosis is poor. The survival rate at 5 years is in the order of 50% [2–4]. The frequent

Figure 3. Histology, magnification 250, HE staining: dense proliferation of fasciculated spindle cells.

Figure 4. Immunohistochemistry, magnification 100, anti-actin antibody: diffuse and intense cytoplasmic staining of tumor cells.
(synchronous and metasynchronous) secondary localisations are reported (mesentery, lung, liver and lymphatic vessels) [2]. Current treatment relies on complete surgical exeresis. Adjuvant management is still highly debated but is based on chemotherapy (particularly for metastatic disease) and/or radiotherapy (for a voluminous lesion, and/or if the margins of the exeresis are affected) [1,3].

The imaging appearance of cross-sections of the leiomyosarcoma of the ureter has been very little reported in the literature owing to the small number of cases studied, and did not seem specific. In general the infiltrating lesion arose from the wall of the ureter and was seen as eccentric or circumferential parietal thickening with no specific enhancement characteristic [5]. Our case however was more of a nodular mass of the wall of the ureter developing exophytically with "crown" enhancement (a hypodense centre and highly enhanced periphery). This type of enhancement is classically described in malignant retroperitoneal tumours, the hypodensity of the centre of the lesion witnessing to necrotic modification which is all the more visible the more voluminous the tumours and/or the greater the degree of malignancy [6]. The kinetics of enhancement of leiomyosarcomas of the ureter are not specifically described in the literature, but it would seem that they are generally enhanced rather early and massively with a hypodense necrotic centre showing their aggressiveness [6]. Our tumour exhibited this type of enhancement.

However, faced with a retroperitoneal nodular mass in contact with the ureter with a CT scan appearance as unspecific as described above, it is necessary to put forward several diagnostic hypotheses.

Because of its frequency, we shall discuss urothelial carcinoma, all the more so since episodes have been reported of haematuria [7]. This carcinoma presents in most cases as circumferential parietal thickening, which is most often symmetrical, rapidly stenosing and in two thirds of the cases involves the distal ureter [7,8]. The lesions are therefore readily multifocal with associated synchronous involvement of the bladder in about 40% of cases [5,7].

The main differential diagnosis is extrinsic compression of the ureter by a tissue process and particularly by a retroperitoneal adenomegaly.

This compression may, in order of frequency, be related either to lymphoproliferative proliferation, to secondary localisation of a urogenital cancer, or to an infection (tuberculosis). In the first case, there are generally multiple, perivascular, polycyclic and confluent adenomegalies forming a uniform mass of tissue not modified by injection. As for metastatic adenopathies, the imaging appearance closely resembles the appearance of lymphomas, so that the clinical context will suggest the diagnosis more than the anatomical distribution of the lymph nodes. However, the tuberculous lymph nodes above all could have mimicked the appearance of our lesion since the adenopathies in question are much smaller than lymphomas, poorly confluent and showing necrotic hypodensity in the centre of the lesion with peripheral enhancement [6]. Analysing the contact angles could in theory differentiate a lesion extrinsic to the ureter of a compressive adenomegaly type, which would have acute contact angles, from an intrinsic lesion, starting in the wall of the ureter and developing exophytically, like the leiomyosarcoma in question, which should have obtuse contact angles. This however is still just theoretical and in practice it is often difficult to differentiate these two entities.

Other less common metastases to be mentioned found in the retroperitoneum originate mainly from melanoma and lung cancer (extra-adrenal metastasis) [6].

Retroperitoneal fibrosis presents as an infiltration "sheathing" the ureter, usually medially, and is responsible for proximal, unilateral or bilateral ureterohydronephrosis [6,7]. The clinical context is again a key element for deciding the diagnosis.

Faced with this abundance of diagnostic hypotheses, the diagnosis is most often confirmed from the
anatomopathological analysis of the lesion following percutaneous or surgical biopsy. In the case of leiomyosarcoma of the ureter, the confirmation of the diagnosis is based more particularly on analysis of the immunohistochemical markers. Indeed, this type of tumour has immunoreactivity for smooth muscle actin and for h-caldesmon, which differentiates it from a rhabdomyosarcoma (the markers of which are desmin, myoglobin and myogenin) [1,9,10].

Conclusion

Leiomyosarcoma of the ureter is a rare tumour with a poor prognosis, the CT imaging appearance of which is little reported in the literature. It is an eccentric, nodular or infiltrating lesion with no associated involvement of the bladder. The characteristics of enhancement are not sufficiently specific to make a diagnosis. This must therefore be based on the anatomopathological analysis with immunohistochemical markers. Treatment is by surgery with adjuvant management which is still to date controversial.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References