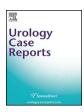
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Oncology

Primary desmoid tumor in renal transplant graft site: First case report



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Introduction

Desmoid tumors are benign neoplasms that make up a subgroup of fibroblastic tumors, characterized for their aggressive local invasion and rare metastization. They originate most commonly in the extremities, intraperitoneal cavity and abdominal and thoracic walls, 75–85% of which are sporadic cases, and the rest related to familial adenomatous polyposis (FAP). They have a correlation with recent pregnancy and previous trauma, occurring two to three times more often in women. Symptomatology may not exist, or be related to local tumor growth, such as pain, palpable mass and intestinal obstruction. Although there are two reports in the literature of onset of primary desmoid tumor after transplantation of solid viscera, 3 we conclude that this is the first case originated in renal graft site.

Case report

We report the case of a 67-year-old female patient, hypertensive and insulin dependent diabetic, with chronic kidney disease diagnosed three years ago, when she underwent renal replacement therapy (hemodialysis) and started nephrological outpatient follow-up aiming renal transplantation. She underwent laparoscopic cholecystectomy ten months before transplantation, without intra or postoperative complications. Renal transplantation of the deceased donor's right kidney was performed, a procedure considered of low immunological risk due to induction with basiliximab, maintaining stable renal function after the use of prednisone, tacrolimus and mycophenolate. The procedure lasted 120 minutes, with two arteries and a 23-min anastomosis time, with adequate perfusion. Post-transplantation ultrasound showed transplanted kidney in the left iliac fossa with smooth contours and preserved cortico-medullary differentiation. One year after transplantation, a new ultrasound of control of the abdomen showed an exophytic nodule next to the middle third of the transplanted kidney, of homogeneous echogenicity, measuring approximately $2.9 \times 2.0 \times 2.8$ cm. Doppler echography was performed four months after that ultrasound scan,

which revealed a hyposonic nodule of 5.4 cm in the middle third of the transplanted kidney, with arterial vessels with a high resistance index inside the lesion compatible with neoplasia. In the same period, a computed tomography scan of the abdomen without contrast was performed. The report showed a well delimited nodular lesion located on the anterior third of the medial third of the renal graft, determining impression on the abdominal wall muscles, measuring $5.8 \times 5.7 \times 4.6$ cm, compromising the entire thickness of the parenchyma (Fig. 1), of etiology to be clarified, considering the possibility of neoplastic lesion due to the significant increase in dimensions and flow at the Doppler; lymphoproliferative disease was questioned. After investigation by imaging tests, the patient was referred to the renal transplantation outpatient clinic of the urology service, and hospital admission was indicated due to probable grafting of renal neoplasia in mind. The surgical team, based on the tomographic hypothesis of lymphoproliferative lesion - whose treatment would save the renal graft - opted for a biopsy with Tru-Cut® in the operation room and sent the material for transoperative examination by frozen-sections. The pathologist's impression consisted of fusocellular lesion without malignancy characteristics, but with the possibility of an inadequate sample. Consequently, an incisional biopsy of the lesion was also performed with the same prior impression. This was followed by resection of the lesion, free macroscopic margins and preservation of the renal graft. The lesion imprinted on the graft, however, it did not invade it (Fig. 2). The definitive anatomopathological examination defined the diagnosis of fusocellular mesenchymal neoplasia, interspersed by collagenated connective tissue, infiltrating fibrous tissue and perirenal skeletal muscle, with 2 mitoses present in 10 fields of increase (Fig. 3). The immunohistochemical panel favored the diagnosis of perirenal soft tissue fibromatosis (CD34, CD99, alpha SMA, desmin, AE1 + AE3 negative and beta-catenin positive). It was then chosen a follow-up as outpatient with ultrasound control every three months.

Discussion

Desmoid tumors mainly occur in the age group of 30-40 years, ¹

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Fig. 1. CT scan showing a well-limited nodular lesion of 5.8cm on the anterior face of renal graft.

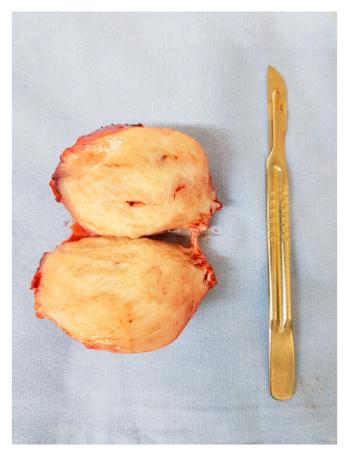


Fig. 2. Final macroscopic aspect of desmoid tumor originated in renal graft site.

composing 0.03% of all neoplasias.⁴ Diagnosis is generally suspected after imaging studies (computed tomography and magnetic resonance), but due to its radiological similarity to other soft tissue sarcomas, core needle biopsy should be used when feasible.¹ From the molecular point of view, stabilization of beta-catenin occurs, either through mutations in the CTNNB1 gene (in sporadic form), or in the APC gene (FAP-associated). In both cases, this protein exerts a proliferative effect through the activation of transcription factors.¹ According to the study by Carlson and Fletcher,⁵ nuclear immunopositivity to beta-catenin was detected in 80% of cases of sporadic desmoid fibromatosis and in 67% of tumors in patients with familial adenomatous polyposis. The authors concluded that nuclear staining for b-catenin is "supportive, but not definitive, of the diagnosis of desmoid fibromatosis", because it can be positive in others fibromatosis, such as

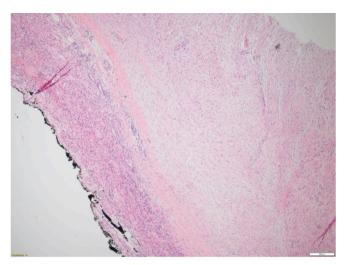


Fig. 3. Microscopic aspect of desmoid tumor originated in renal graft site.

superficial, as well as be negative in this condition. The histological study demonstrates collagenous dense material with interspersed spindle cells and fibroblasts, which may have important mitotic activity. In our case, there was invasion of fibroadipose tissue and perirenal skeletal muscle, consistent with the local aggressiveness of desmoid tumors. In these three cases involving desmoid tumor appearance after solid organ transplantation, in both renal transplants were used basiliximab, in two, tacrolimus, and all patients received corticosteroids in a particular moment of their evolution. There is an increase of up to 10-fold in the risk of neoplasia arising in transplant patients submitted to immunosuppressive therapy, since the immune system also plays a key role in cancer surveillance and recognition. Malignant neoplasms occur in 10-27% after ten years of immunosuppression. There are several guidelines for treatment, such as the European Society for Medical Oncology and National Comprehensive Cancer Network documents, but, in summary, treatment should generally be surgical with negative margins and with preservation of function.

Conclusion

This original report describes the occurrence of a desmoid tumor in the site of renal transplantation, being evaluated as a differential diagnosis, although rare, of primary renal neoplasm of the graft. The understanding of the possibility of this tumor occurrence, simulating primary renal neoplasia (rapid growth and locoregional aggressiveness), would avoid, as in this case, unnecessary graft removal, sparing patients from evolution to dialytic chronic renal disease again.

Conflicts of interest

We have no financial support or conflicts of interest to disclose.

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