Case Report

**Malignant Solitary Fibrous Tumor of the Kidney: Report of the First Case Managed with Interferon**

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Solitary fibrous tumors of the kidney are extremely rare tumors with unpredictable behavior. We describe a case of a patient with a solitary fibrous tumor of kidney with malignant findings with distant metastasis and nephrectomy managed with subcutaneous interferon achieving 23 months of progression-free survival. To date there is no prospective evaluation of any specific modality of treatment, but the surgical management and long-term followup are the only ones so far recommended strategies in the management of these patients. Studies are awaited with more patients to evaluate the different strategies of systemic therapy reported so far to allow adding survival benefit.

1. **Introduction**

Solitary fibrous tumors are rare mesenchymal neoplasms, considered a variant of hemangiopericytomas usually originates in the pleura; however, there have been reports of extrapleural origin (abdomen, retroperitoneum, upper extremities, orbit, cervical, meninges, mediastinum, parotid, nasal cavity, neck, etc.) [1–4]. The location is even more rare urogenital, and according to the literature, only 49 cases of solitary fibrous tumor of the kidney have been reported in the literature [5]. The origin of the majority of cases formed in the kidney is found in the renal capsule tissue or connective tissue interstitial peripelvis. Most cases presented with suspected renal cell neoplasms; however, morphologically, solitary fibrous tumors are characterized by the proliferation of spindle cells with little pattern in their architecture, and the final diagnosis was made with immunohistochemical findings that show staining for CD34 [6–9].

2. **Case Report**

A 49 year old woman with no history of importance who enters the emergency room by 2-months of dyspnea at rest, associated with pleuritic chest pain on right chest, dry cough, no fever. The chest radiograph showed a right pleural effusion and multiple nodular lesions on the pleura, so we decided to perform a CT chest and abdomen in which evidenced free right pleural effusion occupying 80% of the right chest, at least two pleural masses with solid density that capture the contrast, multiple pulmonary nodules with soft tissue density in both lungs and left kidney mass. Carried left nephrectomy with suspected metastatic renal carcinoma, shows renal mass plus liver metastases which were resected. The analysis of the tumor presented as a first option versus solitary fibrous tumor angiomyolipoma. Immunohistochemical studies that showed cell reactivity with CD34, CD99, BCL-2, and vimentin are negative for HMB-45, AMS, CD68, cytokeratin cocktail, and S100, and the Ki67 is not assessable (Figure 1). This profile supports the histological diagnosis of solitary fibrous tumor with origin in the kidney.

We decided to start treatment with interferon a2b subcutaneous dose, extrapolating the cases of patients with solitary fibrous tumor of the pleura [12]. The dose was adjusted for flu symptoms and she is with stable disease at 23 months follow-up.
3. Discussion

Solitary fibrous tumors are extremely rare tumors, arising mostly at the level of the pleura, and cases arising from the urogenital region are even more rare, with 49 cases reported so far in the literature. The histogenesis of this entity is still unknown, but recent studies suggest a primitive mesenchymal cells or level perivascular [10, 11].

The differential diagnosis of these cases includes sarcomatoid variant of renal carcinoma, angiomylipoma, fibromas, and fibrosarcomas. Table 1 presents the cases so far published, reporting the primary source, and histologic variant outcomes.

These results show a relatively rare entity, with peak presentation in the fifth decade of life, arising mostly in the renal parenchyma unilaterally (Table 2).

About 14% of patients have aggressive behavior where common management strategy is nephrectomy with complete resection of the lesions. The pathological findings that have been correlated with aggressive behavior are pleomorphism, increased cellularity and mitotic activity (>4 mitosis/10 high-power fields), necrosis, hemorrhage, and atypical sites (parietal pleura, lung parenchyma) [13]. However, even the clinical behavior can not accurately predict the histopathological findings, as some cases with results suggesting benign disease may show aggressive behavior and vice versa, so it is necessary that these patients have an indefinite period of observation [14–19].

In relation to the management of this condition, there is as yet no prospective evaluation of any specific treatment modality; however, case reports and retrospective case series suggest that complete surgical resection and long-term followup are generally most recommended strategies. In cases such as the present, which present with metastatic disease, there is no clearly defined systemic therapy. Metastasectomy is thought to improve progression-free survival, but in many cases like this, this strategy is not feasible.

In case reports hemangiopericytomas, entity closely related to solitary fibrous tumors, has achieved stable disease with the use of interferon with or without thalidomide. So far, this is the first case reported in the literature in which benefit is demonstrated with the use of interferon in a patient with a malignant variant renal solitary fibrous tumor with metastatic disease, achieving stable disease for about 20 months.

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Some authors suggest the use of antiangiogenic therapies (bevacizumab, sunitinib, pazopanib, etc.), based on the findings of high vascularity and a possible origin of pericytes at this entity [15]. The combination of bevacizumab associated with temozolomide is a potentially promising scheme for patients with solitary fibrous tumors. A series of 14 patients with solitary fibrous tumor unresectable or metastatic, were treated with temozolomide 150 mg/m2 orally on days 1–7 and days 15–21 and bevacizumab 5 mg/kg intravenously on days 8 and 22, with cycles every 28 days. In this study, 11 patients (79%) achieved partial response assessed by Choi criteria with 2 cases (14%) with stable disease. The median progression-free survival was 8.6 months [12, 19, 20].
| Case | Reference | Age | Sex | Localization | Size (cms) | Subtype | Followup (months) | Outcomes |
|------|-----------|-----|-----|--------------|------------|---------|------------------|----------|
| 1    | Fain et al. J Urol Pathol 1996; 4: 227–238 | 45  | F   | Right kidney | 6          | Benign   | 8                | Tumor free |
| 2    | Fain et al. J Urol Pathol 1996; 4: 227–238 | 46  | F   | Right kidney | 7.2        | Benign   | 33               | Tumor free |
| 3    | Fain et al. J Urol Pathol 1996; 4: 227–238 | 51  | M   | Left kidney  | 4.5        | Benign   | 2                | Tumor free |
| 4    | Gelb et al. Am J Surg Pathol 1996; 20: 1288–1295; Fukunaga and Nikaido Histopathology 1997; 30: 451–456 | 48  | F   | Right kidney | 3          | Benign   | 1                | Death from other cause Tumor free |
| 5    | Fukunaga and Nikaido Histopathology 1997; 30: 451–456 | 33  | F   | Right kidney | 3.5        | Benign   | 90               | Tumor free |
| 6    | Fukunaga and Nikaido Histopathology 1997; 30: 451–456 | 36  | F   | Left kidney  | 2          | Benign   | 12               | Tumor free |
| 7    | Hasegawa et al. [2] | 64  | M   | Kidney (laterality not reported) | 4.5 | Benign | 8                | Tumor free |
| 8    | Leroy et al. Urol Int 2000; 65: 49–52; Morimitsu et al. APMIS 2000; 108: 617–625; Yazaki et al. Int J Urol 2001; 8: 504–508; Wang et al. Am J Surg Pathol 2001; 25: 1194–1199 | 66  | F   | Right kidney | 9          | Benign   | 9                | Tumor free |
| 9    | Wang et al. Am J Surg Pathol 2001; 25: 1194–1199; Cortes-Gutierrez et al. J Urol 2001; 166: 602 | 72  | F   | Right kidney | 8          | Benign   | 10               | Tumor free |
| 10   | Wang et al. Am J Surg Pathol 2001; 25: 1194–1199 | 70  | M   | Right kidney | 6          | Benign   | N.A.             | N.A.     |
| 11   | Wang et al. Am J Surg Pathol 2001; 25: 1194–1199 | 41  | M   | Right kidney | 14         | Benign   | 48               | Tumor free |
| 12   | Wang et al. Am J Surg Pathol 2001; 25: 1194–1199 | 72  | M   | Right kidney | 13         | Benign   | 5                | Tumor free |
| 13   | Cortes-Gutierrez et al. J Urol 2001; 166: 602 | 28  | F   | Left kidney  | 15         | Benign   | 12               | Tumor free |
| Case | Reference | Age | Sex | Localization | Size (cms) | Subtype | Followup (months) | Outcomes |
|------|-----------|-----|-----|--------------|------------|---------|------------------|----------|
| 14   | Magro et al. Pathol Res Pract 2002; 198: 37–43 | 31  | F   | Right kidney | 8.6        | Benign  | 8                | Tumor free |
| 15   | Durand et al. Prog Urol 2003; 13: 491–494 | 35  | M   | Right kidney | 17         | Benign  | 6                | Tumor free |
| 16-17| Llarena Ibarguren et al. Arch Esp Urol 2003; 56: 835–840 Bugel et al. Prog Urol 2003; 13: 1397–1401 | 51  | F   | Bilateral    | 25 (left) 2 (right) | Benign  | N.A.             | N.A.      |
| 18   | Urol 2003; 13: 1397–1401 | 60  | F   | Right kidney | 11         | Benign  | 48               | Tumor free |
| 19   | Gres et al. Prog Urol 2004; 14: 65–66 | 82  | M   | Right kidney | 9          | Benign  | 13               | Tumor free |
| 20   | Yamada et al. Pathol Int 2004; 54: 914–917 Pierson et al. Mod Pathol 2005; 18: 159A | 59  | M   | Left kidney  | 6.8        | Benign  | N.A.             | N.A.      |
| 21–27| Median (52.6) range 29–79 | N.A. | N.A. | N.A. median (5.7), range 2.2–10 | Benign  | N.A. | N.A.             | N.A.      |
| 28   | Kawagoe et al. Nishinohon J Urol 2005; 67: 568–571 Johnson et al. J Comput Assist Tomogr 2005; 29: 481–483 | 83  | F   | Left kidney  | 11         | Benign  | 20               | Tumor free |
| 29   | Yamaguchi et al. Urology 2005; 65: 175 | 51  | F   | Right kidney | 11         | Benign  | N.A.             | N.A.      |
| 30   | Kohl et al. Arch Pathol Lab Med 2006; 130: 117–119 | 51  | F   | Left kidney  | 10         | Benign  | N.A.             | N.A.      |
| 31   | Koroku et al. Hinyokika Kiyo 2006; 52: 705–706 Provance / Ferrari et al. Clin Pediatr (Phila) 2006; 45: 871–873 | 85  | F   | Left kidney  | 3.5        | Benign  | N.A.             | N.A.      |
| 32   | Hinyokika Kiyo 2006; 52: 705–706 | 18  | F   | Left kidney  | 3.2        | Benign  | 15               | Tumor free |
| 33   | Provance / Ferrari et al. Clin Pediatr (Phila) 2006; 45: 871–873 | 4   | M   | Right kidney | 8          | Benign  | N.A.             | N.A.      |
| Case | Reference | Age | Sex | Localization | Size (cms) | Subtype | Followup (months) | Outcomes          |
|------|-----------|-----|-----|--------------|------------|---------|------------------|-------------------|
| 34   | Fine et al. [3] Bozkurt et al. APMIS 2007; 115: 259–262 | 76  | M   | Left kidney  | 12         | Malignant | 4                | Persistent tumor  |
| 35   | Bozkurt et al. APMIS 2007; 115: 259–262 | 51  | F   | Left kidney  | 4          | Benign    | 10               | Tumor free        |
| 36   | Znati et al. [10] Constantinidis et al. Can J Urol 2007; 14: 3583–3587 | 70  | M   | Left kidney  | 15         | Benign    | 6                | Tumor free        |
| 37   | Znati et al. [10] Constantinidis et al. Can J Urol 2007; 14: 3583–3587 | 26  | M   | Right kidney | 5          | Benign    | 6                | Tumor free        |
| 38   | Znati et al. [10] Constantinidis et al. Can J Urol 2007; 14: 3583–3587 | 44  | F   | Left kidney  | 5.8        | Benign    | 28               | Tumor free        |
| 39   | Magro et al. [11] Amano et al. Hinyokika Kiyo 2008; 54: 765–769 | 34  | F   | Left kidney  | 9          | Malignant | 15               | Tumor free        |
| 40   | Hidayatullah et al. Hinyokika Kiyo 2008; 54: 357–359 | 67  | M   | Left kidney  | 7          | Benign    | 10               | Tumor free        |
| 41   | Hidayatullah et al. Hinyokika Kiyo 2008; 54: 357–359 | 76  | F   | Right kidney | 2.2        | Benign    | 48               | Tumor free        |
| 42   | Hirano et al. [6] | 75  | M   | Left kidney  | 4.5        | Benign    | 9                | Tumor free        |
| 43   | Hirano et al. [6] | 39  | F   | Left kidney  | 2.5        | Benign    | 12               | Tumor free        |
| 44   | Hirano et al. [6] | 39  | F   | Left kidney  | 20         | Benign    | 6                | Tumor free        |
| 45   | Hirano et al. [6] | 72  | F   | Left kidney  | 19         | Malignant | N.A.             | N.A.              |
| 46   | Hsieh et al. [8] | 50  | F   | Right kidney | 9          | Malignant | 30               | Tumor free        |
| 47   | De Martino et al. [5] | 68  | F   | Left kidney  | 7          | Malignant | 5                | Death by the disease |
| 48   | Caso actual | 49  | F   | Left kidney  | 9.8        | Malignant | 23               | Stable disease    |
Table 2: Clinicopathologic features and outcomes of the 49 cases reported with solitary fibrous tumors of the kidney.

| Feature                        | Value          |
|--------------------------------|----------------|
| Median age in years (range)     | 51 (4–85)      |
| Sex                            |                |
| Male                           | 14             |
| Female                         | 28             |
| Unknown                        | 7              |
| Location                       |                |
| Left kidney                    | 23             |
| Right kidney                   | 17             |
| Bilateral                      | 1              |
| Unknown                        | 8              |
| Site                           |                |
| Kidney                         | 33             |
| Renal capsule                  | 6              |
| Peripelvis                     | 3              |
| Pelvis                         | 1              |
| Unknown                        | 6              |
| Medium size in cm (range)      | 7.6 (2–20)     |
| Histology                      |                |
| Benign                         | 42             |
| Malignant                      | 7              |
| Treatment                      |                |
| Tumor resection                | 2              |
| Nephrectomy                    | 41             |
| Unknown                        | 6              |
| Subcutaneous interferon        | 1              |
| Outcome                        |                |
| No evidence of disease         | 25             |
| Metastasis                     | 4              |
| Unknown                        | 20             |

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