

Primary Ewing Sarcoma of the Kidney with Inferior Vena Cava Invasion

Jong Kil Nam*

Department of Urology, Pusan National University, School of Medicine, Yangsan Hospital, Beomo-ri, Mulgeum-eup, Yangsan, Gyeongnam 626-770, Korea

Abstract

Ewing sarcoma is typically a skeletal based tumor of presumed neuroectodermal origin. Primary Ewing sarcoma of the kidney is exceedingly rare and is usually found in young adults. We report a case of primary Ewing sarcoma with inferior vena cava (IVC) invasion that was managed successfully with radical nephrectomy and partial replacement of IVC using bovine pericardial patch.

Keywords: Ewing sarcoma; Kidney; Inferior Vena Cava

Introduction

Ewing sarcoma is high-grade malignant tumors typically manifest in children and young adults and most common in bone [1,2]. However, extraskelatal Ewing sarcoma such as skin, soft tissue, viscera is less common in and can affect the skin, soft tissue or viscera. Few case of Ewing sarcoma arising from kidney has been previously described in the literature and is potential clinical course toward metastatic disease and death [3,4]. Most patients are young adults with a median age of 28 years (range 4 to 69) and a slight male predominance (1.5:1) [2]. We believe that surgery is the mainstay of treatment [4]. In patient with tumor thrombus or IVC invasion, aggressive surgery to remove the thrombus and invasion in the IVC is reported to be effective in prolonging survival in patients with renal cell carcinoma [5]. However, its value in treating Ewing sarcoma of the kidney is unclear because of limited data and the aggressive nature of the disease. In case of IVC invasion a complete resection is necessary and then IVC resection is main technical challenge.

We report a case of radical nephrectomy and partial replacement of IVC using bovine pericardial patch for primary Ewing sarcoma of the kidney with IVC invasion.

Case Report

A 30-year-old man complained of a one-month history of intermittent palpitation and left flank dull pain. He denied having another symptoms including gross hematuria, fever or general symptoms and looked healthy. He had no remarkable past medical history or familial history. The physical examination showed mild costovertebral angle tenderness. Urine analysis, urine culture, and routine blood chemistry results were within normal limits. Abdominal computed tomography showed that most of the left kidney had been occupied by a huge irregular tumor about 30×20 cm size (Figure 1). The tumor had heterogenous enhancement and some areas of necrosis. The left peri-renal space had strand-like infiltrations and the renal fascia had thickened. The tumor had invaded into the left renal vein and the IVC. Under the impression of renal cell carcinoma, we underwent left radical nephrectomy with IVC partial replacement of IVC using bovine pericardial patch.

The tumor was mobilized en bloc with the left kidney, adrenal gland, ureter, and regional lymph node. The IVC was clamped proximally, contralaterally and distally, and resected for infiltrated tissue of 2 cm in length. The tumor was then removed en bloc with the infiltrated IVC, the left kidney, adrenal gland, ureter. The vascular defect was therefore repaired with the bovine pericardial patch (Figure 2).

Grossly, a huge brown-white color tumor measuring 29×18 cm in dimension was seen, involving the whole kidney. The tumor had invaded the perinephric fat and the renal vein and IVC. Microscopically,

the tumor cells were composed predominantly of monomorphic, small round, hyperchromatic nuclei with scanty eosinophilic cytoplasm, and show rosetoid arrangement (Figure 3). All nodes were negative. In immunohistochemical analysis the tumor showed diffuse strong staining of CD99, a focal staining of synaptophysin and Ki-67 (Figure 4). The tumor cells were negative for the vimentin, CD56, desmin, myogenin, panCK and LCA markers. The final pathological diagnosis was a primary Ewing's sarcoma of the kidney with IVC invasion.

The patient was discharged on the 8th day after surgery. 1 month later, the patient initiated chemotherapy with vincristine, doxorubicin, and cyclophosphamide alternating with isosfamide and etoposide. The patient has completed 12 cycles of chemotherapy and has experienced no adverse event. There is no sign of recurrence in this patient two year after the operation.

Discussion

Ewing's sarcoma is the second most common skeletal tumor in

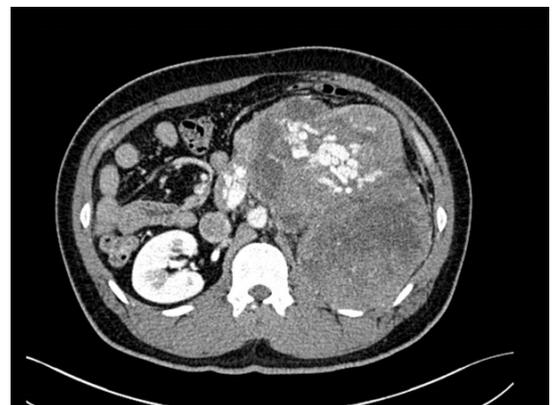


Figure 1: Abdominal computed tomography showing that most of the left kidney had been occupied by a huge irregular tumor.

*Corresponding author: Jong Kil Nam, Department of Urology, Pusan National University, School of Medicine, Yangsan Hospital, Beomo-ri, Mulgeum-eup, Yangsan, Gyeongnam 626-770, Korea, Tel: 82-55-360-2134; E-mail: toughkil76@naver.com

Received March 24, 2014; Accepted December 30, 2014; Published January 03, 2015

Citation: Nam JK (2015) Primary Ewing Sarcoma of the Kidney with Inferior Vena Cava Invasion. Med Surg Urol 3: 146. doi:10.4172/2168-9857.1000146

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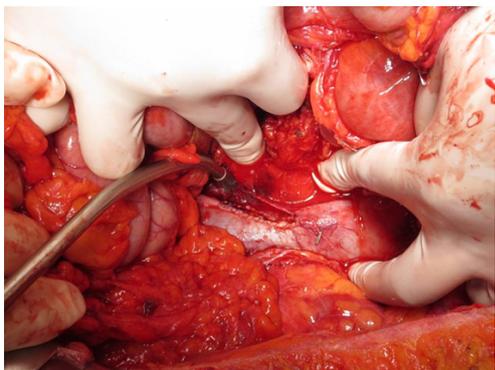


Figure 2: Intraoperative findings after caval reconstruction with a bovine pericardial patch.

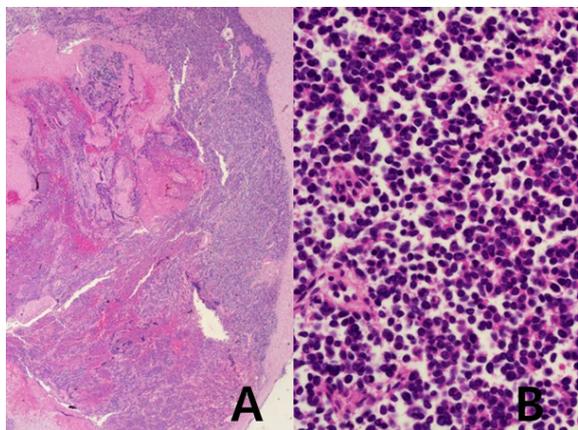


Figure 3: (A) The resected inferior vena cava showed invasive sarcoma (H&E, x40). (B) The tumor cells have round and hyperchromatic nuclei and show rosetoid arrangement (H&E, x400).

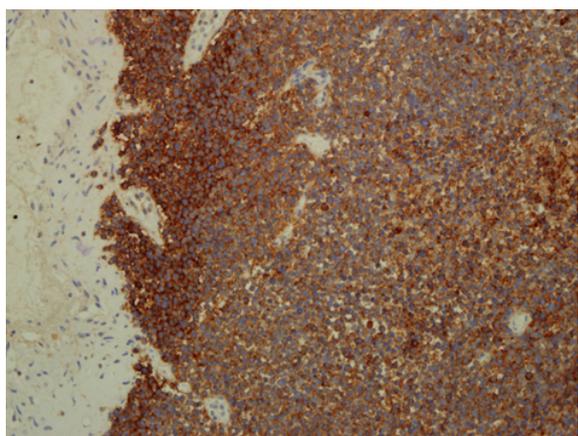


Figure 4: The tumor cells showing strong membranous immunoreactivity for CD99 (x200).

children and young adults [1]. However, primary Ewing's sarcoma of the kidney is rare, with few cases reported in the literature and no available treatment strategies [2-4]. Also, much less frequently reported is venous involvement by Ewing's sarcoma of the kidney, and it is exceedingly rare to invade the wall of the IVC [4].

The imaging appearance of Ewing's sarcoma of the kidney is nonspecific, and these tumors should be in the differential diagnosis when a large retroperitoneal mass with aggressive features is encountered [6]. The major differential consideration for a perinephric soft tissue mass includes Wilms tumor, neuroblastoma, lymphoma, rhabdomyosarcoma, renal cell carcinoma, transitional cell carcinoma and metastatic tumors [6].

In some literature, long-term disease free survival can be achieved with chemotherapy alone [7]. The role of cytoreductive nephrectomy is not defined in this patient population [7]. However, we think that surgical resection followed by chemotherapy and local radiation therapy offers the best chance for long-term survival, similar to renal cell carcinoma [5]. Parham et al. reported 79 cases of primary Ewing's sarcoma of the kidney. However, it is uncertain whether all cases were indeed primary Ewing's sarcoma of the kidney, and only small cases provided follow-up results [8]. Thyavihally et al. reported a single institution experience of 16 patients with primary Ewing's sarcoma of the kidney, which were treated with radical nephrectomy in operable cases and chemotherapy in all patients [9]. Overall median survival was 40 months with a 3-year survival of 60% and 5-year survival of 42%. A 5-year disease free survival rate of 36% was observed in patients with no lymph node or distant metastasis [9].

Ewing's sarcomas are sensitive to chemotherapy [7,10]. The use of chemotherapy has greatly improved survival for patients with localized tumor, from 10% to 70-80% [10]. However, it has less effective for patients with metastases [10]. A combination of neoadjuvant chemotherapy with local control, followed by adjuvant chemotherapy, is implemented. Local control is achieved with surgical resection and/or radiation therapy. In our cases, under the impression of renal cell carcinoma, we underwent left radical nephrectomy with IVC reconstruction. In this case, that tumor invaded the caval wall, medial venotomy carries a higher risk of narrowing after caval wall excision and renal vein repair, and then we underwent caval wall reconstruction using a bovine pericardial patch.

Despite attempts at multimodal therapies, prognosis of metastatic cases remains poor. However, we think that aggressive surgical treatment followed by chemotherapy and local radiation therapy offers the best chance for long-term survival. Long-term follow up will be necessary to verify prognosis of aggressive therapies. In this report the feasibility and safety of the aggressive surgery is documented.

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