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Case Report
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Teratoid Wilms Tumor with Tumor Embolus in Inferior Vena Cava and Right Atrium: Experience from Single Tertiary Hospital

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Abstract

Teratoid Wilms Tumor (TWT) with tumor embolus in inferior vena cava and right atrium is extremely rare. These patients have risks of cardiac arrest and need emergency treatment after diagnosis. However, pathologic diagnosis may be difficult at the initial diagnosis due to the limited tumor tissue by needle biopsy and uncommon unique pathological characteristics in TWT. This makes it hard to decide the appropriate initial treatment. We reported a three-year-old boy presented with abdominal pain and a huge mass with tumor embolus in inferior vena cava and right atrium. Pathologic diagnosis by needle biopsy for this patient was uncertain because the tumor tissue was composed of immature rhabdomyomatous elements but free of stromal, blastemal, and epithelial elements. After discussion by multidisciplinary team, two courses of chemotherapy were given according to clinically diagnosed Wilms tumor. But the tumor enlarged. Then surgical operation was immediately performed by pediatric surgeon combined with cardiothoracic surgeon. Both the primary tumor (include left kidney) and the tumor embolus were completely resected. The final pathology revealed the diagnosis as TWT. Further eight cycles of chemotherapy and local postoperative radiotherapy were given. The child has been followed up for 27 months and still with disease-free survival. The cooperation of multidisciplinary team during the whole diagnosis and treatment was an important factor for the long-term survival of this case.

Keywords: Teratoid; Wilms tumor; Tumor embolus; Multidisciplinary team

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Introduction

Teratoid Wilms Tumor (TWT) is a rare variant of Wilms Tumor (WT). It was first described by Variend [1], and then defined by Fernandes as a variant of WT, which more than 50% of the mass contains heterologous elements [2]. There were only 33 cases reported till now according to the literature [3]. The rate of invasion of WT to the Inferior Vena Cava (IVC) was approximately 5%, while the rate of invasion of WT to the right atrium was less than 1% based on available reports [4,5]. We present an extremely rare case which diagnosed as TWT with huge tumor embolus in both IVC and right atrium. Difficulty of diagnosis and poor effects to pre-operative chemotherapy were two tricky problems during the treatment.

Case Presentation

A three years old boy came to our hospital with abdominal pain and huge abdominal mass. Actually, the abdominal pain occurred three months ago, just with intermittent, slight and without other discomfort, the parents did not pay any attention to his symptom. One day, the mass was found in the left abdomen by coincidence, showing firm, stable, local tenderness. The ultrasound revealed a solid tumor associated with left kidney measuring 164 mm × 83 mm × 67 mm with mixed signal and calcification. To our surprise, filling defect was found in whole of IVC (Figure 1A) on enhanced Computed Tomography (CT). The tumor embolus even spread to the right atrium. The pathologic diagnosis of the tumor was uncertain since the biopsy tissue was composed of immature rhabdomyomatous (Figure 2A) elements but free of stromal, blastemal, and epithelial elements. No metastasis was detected. We made a clinical diagnosis as Wilms tumor based on radiographic characteristic after discussion by Multidisciplinary Team (MDT). Then two cycles of ICE (ifosfamide + vincristine + etoposide) chemotherapy were given to the patient. The tumor

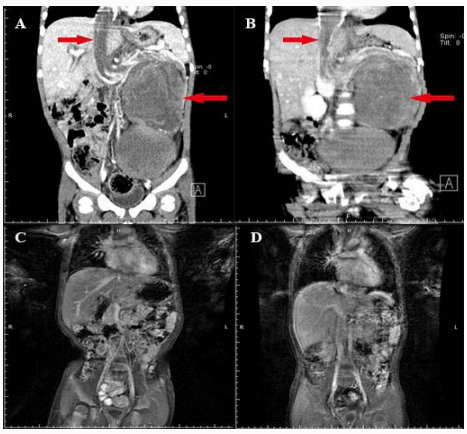

Figure 1: Imaging findings of the patient with Teratoid Wilms tumor.

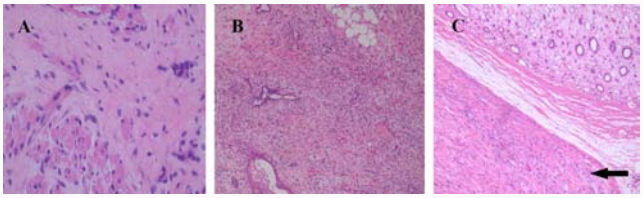

Figure 2: Pathological characteristics of the patient with Teratoid Wilms tumor.

even became larger after chemotherapy (Figure 1B). So surgical intervention was immediately performed by pediatric surgeon combined with cardiothoracic surgeon. Both the primary tumor (include left kidney) and the tumor embolus were completely resected (Figure 1C). The tumor size was 200 mm × 100 mm × 90 mm, when the tumor embolus showed trab-like structure with the size of 100 mm long and 20 mm to 25 mm in diameter. The final pathology revealed the diagnosis as TWT. A few epithelial elements were found in the tumor tissue (Figure 2). Immunohistochemical staining for WT1 was negative. Local lymph nodes were free of tumor. However, blood vessels were invaded by tumor tissues under light microscope. Post-operative monitoring of circulation system and respiratory system, anti-infective therapy and nutrition support were given to the patient in the pediatric intensive care. Local radiotherapy followed closely after operation. Then further eight cycles chemotherapy were given based on the protocol of WTSG-5-DD4A. The patient was free of disease during the following-up for 27 months (Figure 1D). Cardiac and kidney function were at normal range during the outpatient service. A) CT scan of abdomen (before initial diagnosis) showing a large mass (79 mm × 88 mm × 155 mm) associated with left kidney and tumor embolus in IVC and right atrium (arrow heads). B) CT scan of abdomen (after two cycles of chemotherapy) showing the mass (arrow heads) enlarged (81 mm × 93 mm × 162 mm). C) Magnetic Resonance Imaging (MRI) scan of abdomen (one month after operation) showing no residual tumor and tumor embolus. D) MRI scan of abdomen (two years after initial diagnosis) revealing no recurrent disease. A) High magnification photomicrograph revealing immature rhabdomyomatous of the tumor (H&E; x400). B) Low magnification photomicrograph revealing fibrous tissue, immature rhabdomyomatous, adipose tissue and mixed epithelial including renal tubular epithelial cell and squamous epithelial cell (H&E; x100). C) Another area showing tumor tissue (arrow heads) and normal renal tissue.

Discussion

Abdominal pain was the only initial symptom for this patient, similar to other patients with WT [6]. However, the tumor embolus in IVC and right atrium would never be considered only when it was found by imaging studies. Fortunately, the child did not suffer from cardiac arrest. But for other patients with WT, cardiac arrest may be the only initial symptom [4], which needed to attract pediatrician attention. The mesenchymal contents may consist of 50% to 90% in the TWT [3]. Needle biopsy of the tumor may be uncertain due to the limited tumor tissue and uncommon unique pathological characteristics [7,8]. This explained why the initial pathologic diagnosis was difficult in our case. To prevent from cardiac arrest in our patient, chemotherapy was firstly considered to reduce tumor volume after tissue biopsy. However, the tumor enlarged. In fact, several cases of TWT showed quite insensitive to chemotherapy or radiotherapy due to the well-differentiated elements (>50%) in tumors according to the literatures [3,9]. It also deciphered the poor response to chemotherapy at the beginning in our case. In view of this, TWT should be considered once the immature rhabdomyomatous contents were the only component of biopsy tissue in renal tumors. The surgical operation was of vital importance to the outcome of this patient. Although there was no recognized optimal treatment plan for TWT due to its rarity, surgical resection was recommended which took precedence of chemotherapy or radiotherapy. While it was hard to make this decision, especially the pathological diagnosis was uncertain and tumor bulk was too huge. But surgical intervention should have priority once the chemotherapy or radiotherapy is not effective. Finally, MDT played an important role in the whole diagnosis and treatment for this patient. The imaging assessment of primary tumor and tumor embolus made contribution to the success of operation. Also, postoperative monitoring in pediatric intensive care unit greatly reduced the risk of post-operational acute pulmonary embolism.

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