

Inflammatory Myofibroblastic Tumor of Pelvic Cavity in a 17- Year- Old Girl with Previous History of Wilms' Tumor: A Case Report

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Background: Existence of Synchronous or metachronous tumors in one patient is a rare phenomenon. However, some genetically controlled processes can lead to multiple tumors in one patient. Inflammatory myofibroblastic tumor is a rare neoplasm that usually happens in the lung and the usual location for extra pulmonary presentation is kidney.

Case Report: In following article we report a 17- year old girl presented with hypo gastric abdominal pain fever and malaise which CT scan showed a huge mass in the pelvic cavity. The patient previously underwent left nephrectomy due to Wilms' tumor in childhood. The pathology report of the resected pelvic mass showed the spindle cell tumor and inflammatory myofibroblastic neoplasm.

Conclusion: Despite improvement in diagnostic procedures, the precise diagnosis of myofibroblastic tumor remains highly dependent on surgery and following pathologic assessments.

Keywords: Inflammatory myofibroblastic tumor, Wilms' tumor, Spindle cell tumor

Background

Wilms' tumor is a childhood neoplasm that usually is presented with a painless mass in the abdomen. The prognosis of the disease is good with 90% survival estimated at five years and most of patients are treated completely without any future complications (1). However, one of the worries about this neoplasm is the metastasis to other organs with pulmonary system and liver being the common sites involved with the metastasis (2).

Garden M et al. reported coincidence of aggressive inflammatory pseudo-tumor (IPT) of abdomen in a 15 year-old boy 9 years after therapy for Wilms' tumor (3). However, Co-incidence of Wilms' tumor with other tumors is a rare phenomenon.

Inflammatory myofibroblastic tumor is a rare disease of the kidneys that can be seen at any age. Although the most common site of tumor is lung, but it can also be seen in extra pulmonary sites such as the genitourinary system like bladder and infrequently in kidneys (4).

Here, we introduce a case which was involved with both of these neoplasms: Wilms' tumor in childhood and inflammatory myofibroblastic tumor in teenage.

Case presentation

We report a 17- year- old girl case of Wilms' tumor that presented with a huge pelvic mass several years after treatment.

The patient underwent left nephrectomy at the age 3 due to Wilms' tumor followed by adjuvant chemoradiotherapy.

During the follow up period, the patient showed no tumor related complications until the age 17

when she developed abdominal pain, especially in hypogastric area and had a huge palpable pelvic mass associated with malaise and periodic fever. Abdominal ultra-sonography revealed a solid mass in the pelvic cavity with diameter 140*75 mm. Abdominopelvic MRI also showed 158*86 mm heterogeneous pelvic mass, with irregular wall enhancement with a pressure effect on right ureter (figure 1).

Colonoscopy was normal. However an external pressure effect was observed, which suspected to arise due to a mass in the pelvic cavity.

The ultrasound guided core needle biopsy of the mass showed spindle cell proliferation without any cytological atypia. Immunohistochemistry study on biopsy specimen was positive for SMA, while other tumor markers such as CD117, S100, Desmin, Inhibin, ER, B-hCG and CA125 were negative (figure 2, A & B).

The tumor was resected by lower midline incision without damage to abdominal or pelvic organs. The tumor had just loose adhesion to the uterus and other abdominopelvic organs (figure 3). The pathology of the resected mass was in favor of benign spindle cell neoplasm that infiltrated by inflammatory cells and confirmed CNB report. Pathologic assessment diagnosed as an inflammatory myofibroblastic tumor.

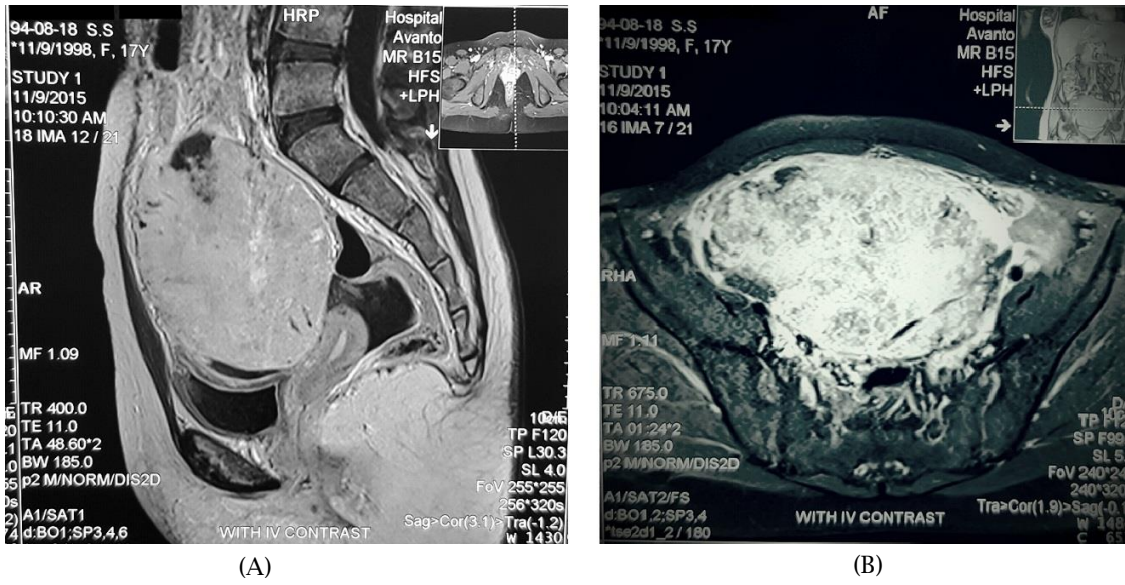


Figure 1. Pelvic MRI with contrast of patient, (A) Sagittal view (B) Axial view

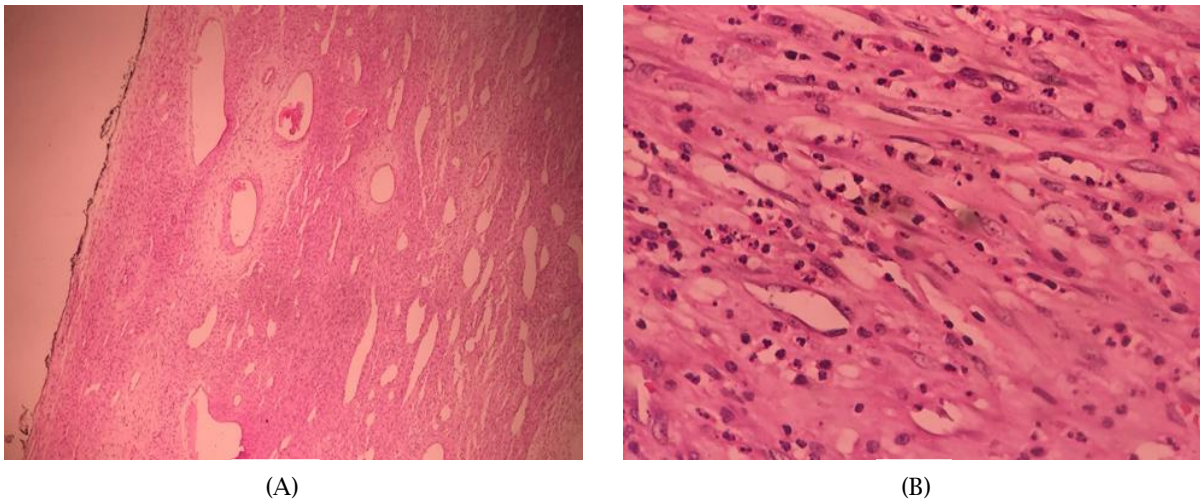


Figure 2. (A). H&E slide of core needle biopsy; (B) IHC slide of core needle biopsy.

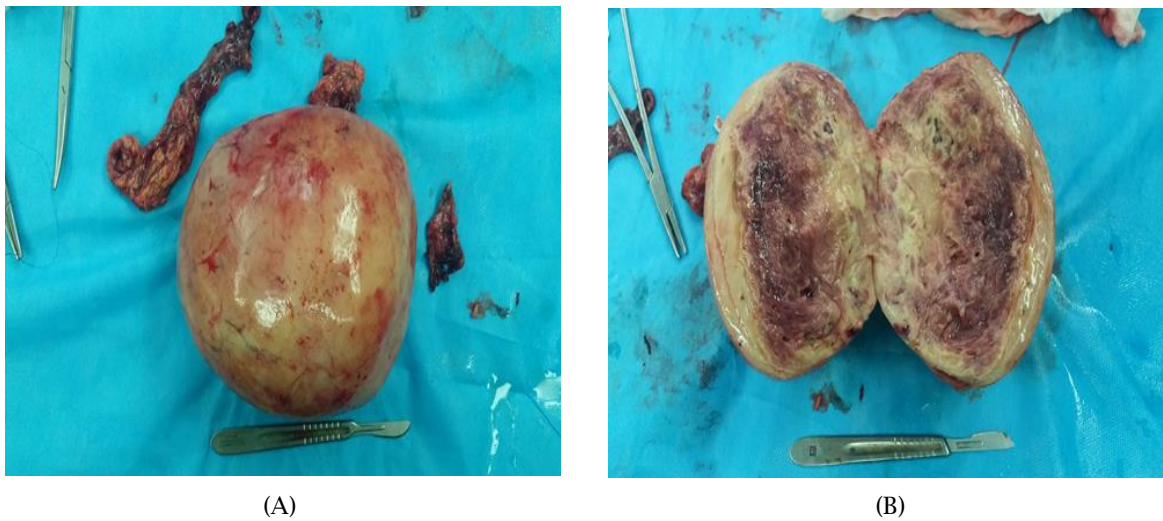


Figure 3 (A) gross morphology of resected pelvic mass; (B) gross appearance pelvic mass

Discussion

Wilms' tumor or nephroblastoma is a rare tumor of children aged 3- 5 years old and usually affects one kidney. This tumor is the most common cause of kidney neoplasm in children. The tumor rarely affects both kidneys (5).

The association of this tumor with other types of tumor is not clearly understood. The cause of cancer is possibly DNA mutations, which lead to uncontrolled proliferation of cells; as shown in many studies, WT1 gene mutations are associated with Wilms' tumor (6). This malignant cancer consists of metanephric blastoma, stromal and epithelial derivatives. Presence of abortive tubules and glomeruli that are surrounded by spindle cell stroma are the commonly known histological characteristic of this malignancy (7). The mesenchymal component may include cells showing rhabdomyoid differentiation as rhabdomyosarcomatosis Wilms' tumor (7).

Some specific conditions such as having a family history of Wilms' tumor or having the black ethnicity can increase the risk of Wilms' tumor. Also the risk of cancer is higher in patients with anhydria, hemihypertrophy, undescended testis and hypoadiadasis (8).

The syndromes which are related to Wilms' tumor are:

1. WAGR syndrome, which is represented by anidria, abnormalities of the genitalia and urinary system, mental retardation and Wilms' tumor
2. Deny Drash syndrome which is associated with kidney disease, psudohermaphroditism and Wilms' tumor.
3. Beckwith-Wiedemann syndrome: the signs are macroglossia and enlarged internal organs with Wilms' tumor (8).

Recent studies have shown relationship between Wilms tumor and other carcinomas. A study by Lange et al. showed that female survivors of Wilms' tumor had 9.1 fold increased risk of breast cancer between ages 40-45 (9).

Berdugo et al. presented the two cases of spindle cell epithelium of the vagina that expressed WT1 protein in the extracted cells. These patients had benign tumors that were treated with excision of tumor mass and morphology of the cells showed spindle cell tumor (10).

Another case report by Yao presented a 10 year old boy with metanephric adenofibroma. This cancer is a rare cancer that involves kidney and urinary tract and is known as the well differentiated form of the Wilms' tumor. The expression of WT-1 in this cancer is also proved (11).

As discussed earlier, Wilms' tumor has a probable relationship with other cancers such as breast or rhabdomyosarcoma. However the presence of this cancer with a myofibroblastic tumor is so rare. The precise and definite diagnosis of the origin of these two neoplasms is by checking WT-1 protein status.

Inflammatory myofibroblastic tumor (IMT), which is also called the inflammatory pseudo-tumor, is a rare disease of kidneys in childhood that mostly occurs in the lungs. This cancer consists of spindle cells with variable matrix surrounding those cells.

Three different histological types for IMT have been known: inflammatory infiltration, spindle cell proliferation and hypo cellular fibrous pattern. Genetic markers that are suggested to be expressed in IMT are anaplastic-lympho-markers (ALK) and WT-1 which are useful to confirm the diagnosis (12).

In this report we presented a case with two neoplasms metachronously found in one patient with both neoplasms possibly having the same origin. Overall, we suggest evaluating the tumor markers and expressed proteins in resected mass tissue to define the source of this new pelvic mass. Validating tumor markers such as WT-1 can guide us to comprehend the relationship between Wilms' tumor and inflammatory myofibroblastic neoplasms.

Conclusion

Inflammatory myofibroblastic neoplasm can occurs in patients with Hx Wilms' tumor, which make it necessary to check genetic cause and follow the Wilms' tumor patients for inflammatory myofibroblastic neoplasm.

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