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# 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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**Abstract:** 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(IMT) is a rare neoplasm that usually happens in the lung and the usual location for extra pulmonary presentation is kidney. 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. 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 usual- ly is presented with a painless mass in the abdo- men. 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 or- gans with pulmonary system and liver being the common sites involved with the metastasis (2).

Garden M et al. reported coincidence of aggres- sive 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.

IMT 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 tu- mor in teenage.

## Case presentation

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

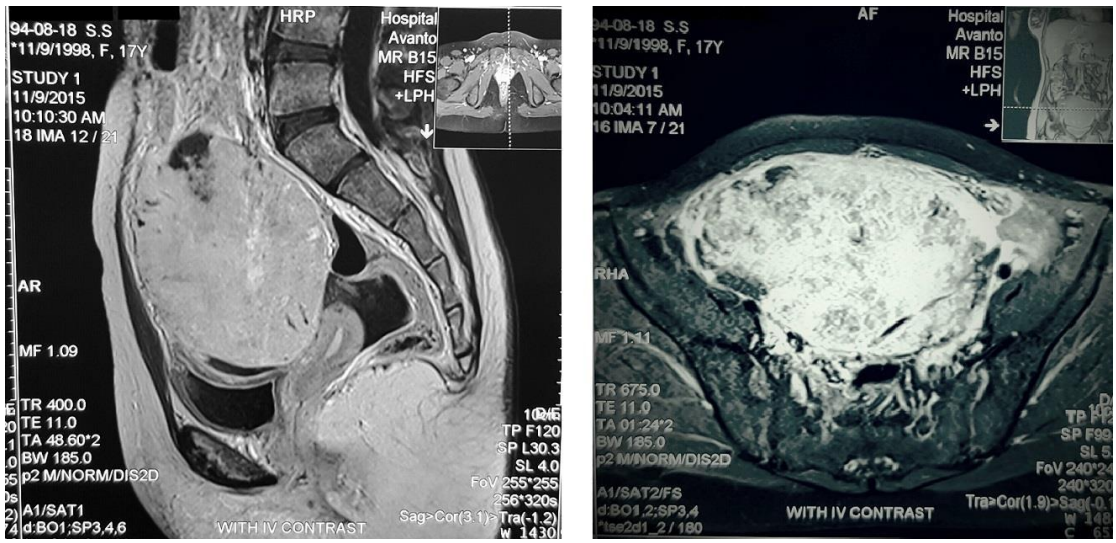
The patient underwent left nephrectomy at the age 3 due to Wilms' tumor followed by adjuvant chemoradiotherapy.

During the follow up period, the patient showed no tumor related complications until the age 17 when she developed abdominal pain, especially in hypogastic 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. Ab- dominopelvic MRI also showed 158*86 mm hetero- geneous pelvic mass, with irregular wall enhance- ment 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.

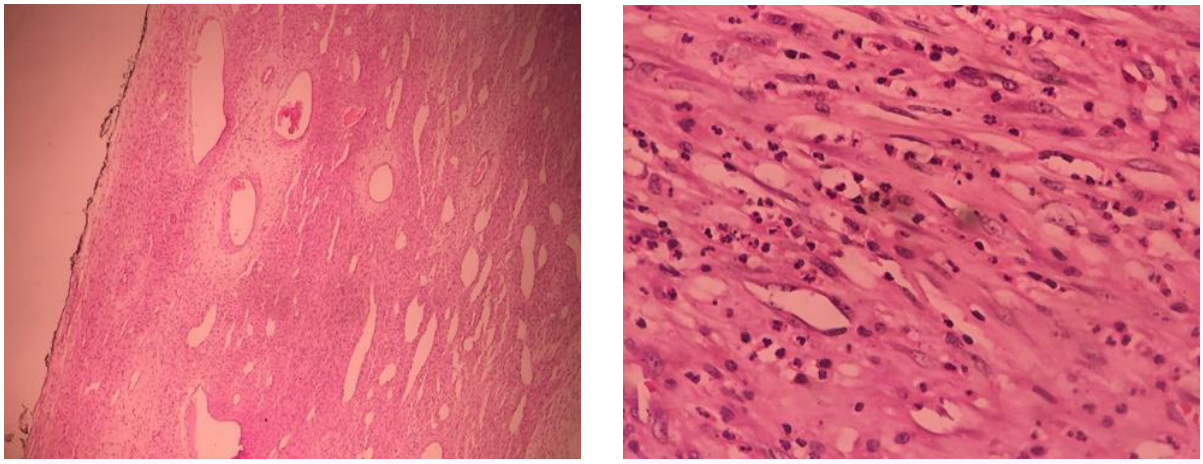
The ultrasound guided core needle biopsy of the mass showed spindle cell proliferation without any cytological atypia. Imunohistochemistry study on biopsy specimen was positive for SMA, while other tumor markers such as CD117, S100, Desmin, In- hibin, ER, B-hCG and CA125were negative (figure 2, A & B).

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 pa- thology of the resected mass was in favor of be- nign spindle cell neoplasm that infiltrated by in- flammatory cells and confirmed CNB report. Patho- logic assessment diagnosed as an inflammatory myofibroblastic tumor.



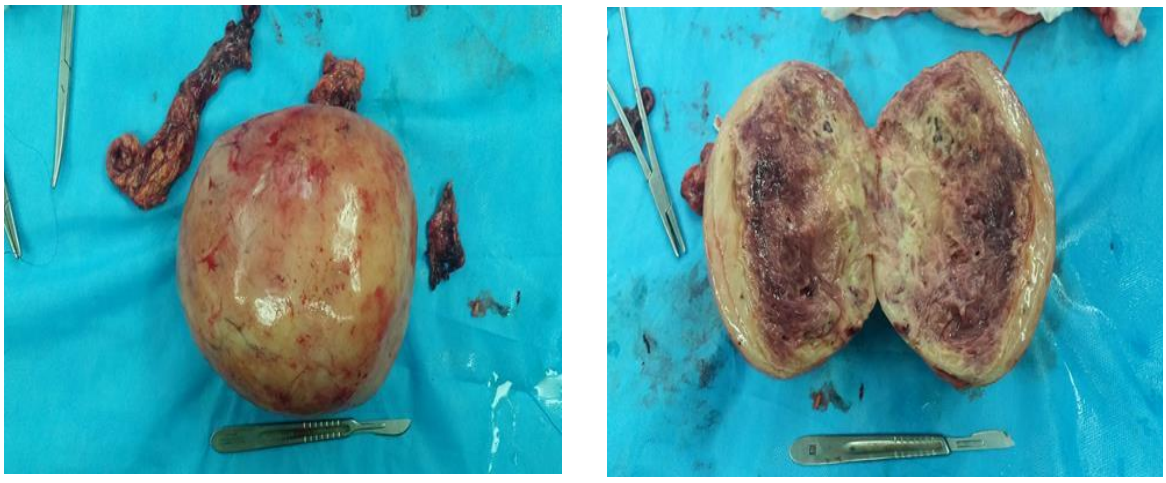

(A) (B)

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



(A) (B)

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



(A) (B)

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 methanephric blastoma, stromal and epithelial de- rivatives. 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 ethnic- ity can increase the risk of Wilms' tumor. Also the risk of cancer is higher in patients with anhydria, hemihypertrophy, undescended testis and hypo-spadiasis (8).

The syndromes which are related to Wilms' tu- mor are:

1. WAGR syndrome, which is represented by aniridia, 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 macrogolossia 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 uri- nary 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 proba- ble 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.

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 ex- pressed 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 neo- plasms 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. Validat- ing 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 oc- curs in patients with Hx Wilms' tumor, which make it necessary to check genetic cause and follow the Wilms' tumor patients for inflammatory myofibro- blastic neoplasm.

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