

## Case Reports

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### Mediastinal Cystic Teratoma with Right Sided Extra Renal Wilms' Tumor - A Rare Case Report

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**Abstract:**

*A teratoma of the mediastinum is an uncommon germ cell tumor. Five percent of germ cell tumors are extragonadally located and men are affected more than women. Most mediastinal teratoma produce no symptoms, but we are presenting a case which was present with respiratory distress in early childhood. Classical wilm's tumor usually found within the kidney but rarely it can occur in extra renal situations. Combination of these two is not reported until today we have searched extensively.*

*We are presenting a 26 month old boy with mediastinal cystic teratoma with right sided extra renal wilms' tumor which may be the first reported case. Combined effort of various concern departments ensures the diagnosis and management of such a rare case.*

**Key words:** Mediastinal Teratoma, Extra Renal Wilms' Tumor, Children

**Introduction:**

A **teratoma** is an encapsulated tumor with tissue or organ components resembling normal derivatives of more than one germ layer. The tissues of a teratoma, although normal in themselves, may be quite different from surrounding tissues and may be highly disparate; teratomas have been reported to contain hair, teeth, bone and, very rarely, more complex organs or processes such as eyes, torso, and hands, feet, or other limbs. Usually, a teratoma contains no organs but rather one or more tissues normally found in organs such as the brain, thyroid, liver, and lung. Sometimes, the teratoma has within its capsule one or more fluid-filled cysts; when a large cyst occurs, there is a potential for the teratoma to produce a structure within the cyst that resembles a fetus. Teratomas are usually benign, although several forms of malignant teratoma are known and some of these

are common forms of teratoma. A mature teratoma is typically benign.<sup>1-2</sup>

A teratoma of the mediastinum is an uncommon germ cell tumor. Five percent of germ cell tumors are extragonadally located and men are affected more than women. Most mediastinal teratomas produce no symptoms and they are more commonly associated with compression of adjacent structures, predominantly those of the respiratory system. Another signal is bleeding or rupture of the tumor into the bronchial tree, pleura, or pericardium.<sup>3</sup>

Wilms' tumor, also known as nephroblastoma is an embryonal malignancy of the kidney. It is the most common primary malignant renal tumor of childhood.<sup>1</sup> Most cases are sporadic, although 1-2% of patients have a familial predisposition to wilms' tumor which are inherited in an autosomal dominant manner with variable penetrance. The classical location for Wilms' tumour is the kidney. However, cases with typical morphologic features of Wilms' tumour have been recorded in extrarenal sites, including the retroperitoneum, sacrococcygeal region, testis, uterus (sometimes presenting as a cervical polyp), inguinal canal and mediastinum.<sup>4</sup>

Occurrence of extrarenal Wilms' tumour (ERWT) is very exceptional and the diagnosis is almost always

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made after surgical intervention. The exact mechanism whereby a WT occurs in extrarenal tissues is not known. Even if the histological characteristics are the same as in intrarenal WT, a retroperitoneal teratoma should be investigated for a possible admixture of WT cells. Literature search suggests a similar course for ERWT as that of the renal counterpart, therefore, similar staging and treatment protocols can be followed. There is a slight male preponderance with an M: F ratio of 3:2. The age at presentation usually ranges from two months to ten years, although exceptionally young (an eight day-old child) and old (77 year-old female) cases have been reported.<sup>5-7</sup>

### Case Summery

A 26 months old presented with fever, cough, chest pain & respiratory distress for 8 months. His fever was low grade, intermittent, not associated with chills and rigor or night sweat. However he had no H/O hemoptysis and contact with TB patient. Respiratory distress was not relieved by any medication. Pain is so severe he used to scream.

With these complains, in last 8 months, he got admitted 9 times in 6 different hospitals and investigated with CXR 16 times, USG of Abdomen 4 times, CT Scan of Chest twice and treated with many antibiotic & anti-asthmatic drugs and he was undergone minor chest surgery (ICT) 5 times and major surgery once (mini thoracotomy & open drainage). On examination he was irritable, dyspnoeic, mildly anaemic screamed with pain with features suggestive of left sided pleural effusion with scar marks & a sinus with purulent discharge on anterolateral chest wall on left side. His right kidney was palpable, ballotable & non-tender. Liver & spleen were not palpable.

His CXR suggested left sided plural effusion, Hydropneumothorax with pleural thickening, collapse of left lung and CT scan suggested large empyema thoracis. USG of abdomen revealed well defined hypoechoic mass 3.9X3.4cm in mid & lower zone of right kidney (Nephroblastoma Rt). USG guided FNAC from renal mass represented blastemal cell of nephroblastoma.

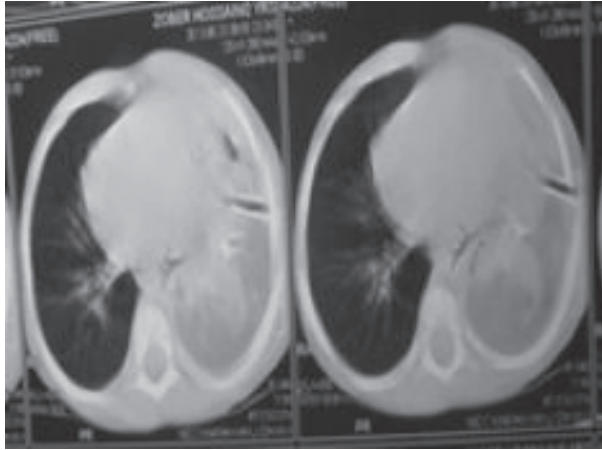
Neo-adjuvant chemotherapy started but not improved. With an intension to palliation, under high risk bond consent, mini thoracotomy was done under G/A. On exploration there was a huge mass occupying whole hemithorax which was both cystic and solid and composed of hair, tooth, and many soft tissue structures. Pus was evacuated and there was no metastasis. Histopathology of soft tissue mass revealed multiloculated cystic mass showing skin, bones, cartilage and sebaceous material. Microscopically cyst wall lined by stratified squamous epithelium with areas containing hair shaft, respiratory epithelium & sebaceous material. No granuloma or malignancy seen and diagnosed as benign cystic teratoma. The child got relieved from respiratory distress and made ready for excision of renal mass. The mass was totally removed with capsule which was situated on the lateral border of the right kidney. Frozen section report was positive for malignant cell. Histopathology of tumor mass microscopic appearance: Sections from submitted tissue show Extra Renal Wilms' tumor. The slide was reviewed by a board of pathologist and confirmed that it was a case of extra renal Wilms' Tumor. Immuno-histochemistry also done in Pathology Dept. of BSMMU. The pt treated with chemotherapy containing Vincristine, Actinomycin-D and Doxorubicin. Now he is on regular follow up for last 50(fifty) months. He is growing well in good health without any adverse event.



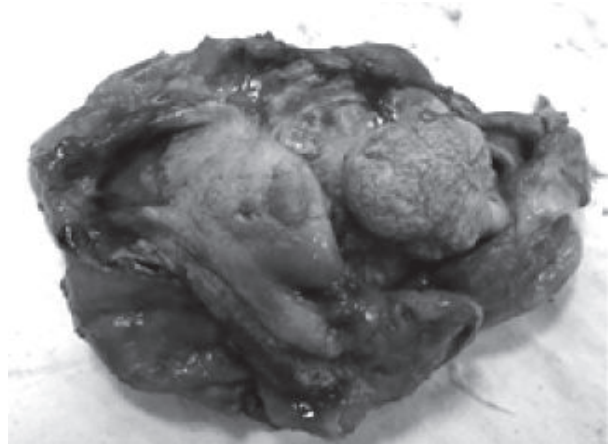
Fig.-1. Chest X-ray before Operation



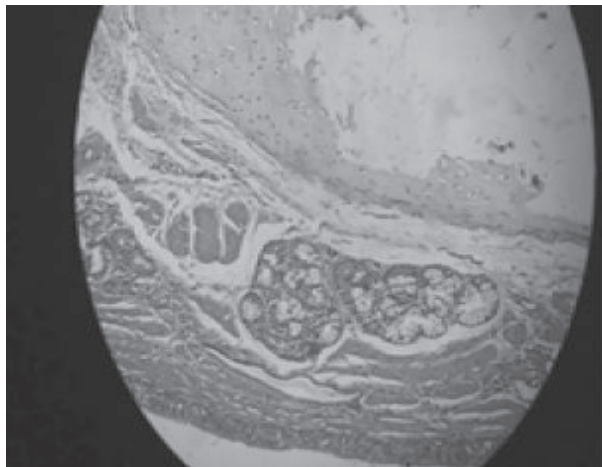
Fig.-2: Chest X-ray after Operation



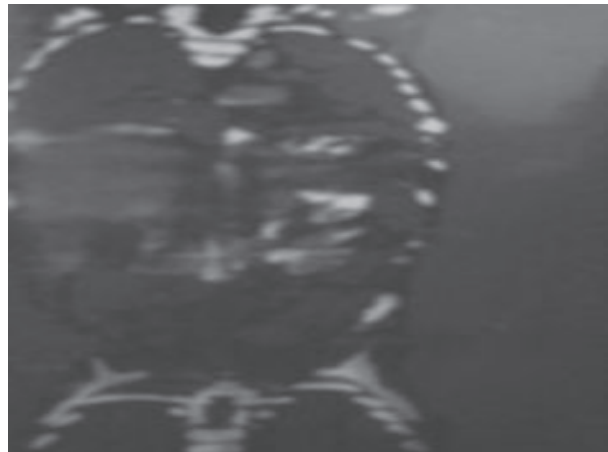
**Fig.-3:** CT Scan of chest



**Fig.-4:** Mediastinal mass(Cystic Teratoma)



**Fig.-5:** Histology of Mediastnal Teratoma



**Fig.-6:** CT Abdomen

### Discussion

Mediastinal teratomas are the most common extra-gonadal germ cell tumours. They account for approximately 15% of anterior mediastinal masses in adults and approximately 25% of anterior mediastinal masses in children. The age of presentation is wide, but typical presentation in adults is in the third or fourth decade (20s and 30s). In children, the majority of patients are asymptomatic, with the mediastinal mass discovered incidentally when the thorax is imaged for another reason. Masses that become symptomatic can do so in a variety of ways : mass effect: respiratory distress (infants), respiratory failure, superior mediastinal syndrome, neck mass, Horner syndrome, if ruptures may presents with chest pain, haemoptysis, cardiac tamponade, pleural effusion.<sup>8-9</sup>

The vast majority of mediastinal teratomas are located in the anterior mediastinum (80%), with most of the remainder involving multiple compartments (13 - 15%). Isolated posterior or middle mediastinal location is uncommon (2-8%). Appearances on chest radiography are usually indistinguishable from many of the other causes of an anterior mediastinal mass. Calcification may be visible. CT is the mainstay of diagnosis. The appearance will depend on the type of teratoma, and whether or not a cystic teratoma has ruptured. They are all however usually located in the anterior mediastinum. **Mature teratomas** have been associated with Klinefelter syndrome, **Immature teratomas** can be associated with non-lymphocytic leukaemia and pleomorphic undifferentiated sarcoma.<sup>8-9</sup>

The finding of extrarenal Wilms' tumours outside the course of germ cell migration, such as those arising

in the inguinal area, would suggest that they may not necessarily be teratoid but may have originated from misplaced primitive nephrogenic blastema. The extrarenal location of teratoid Wilms' tumour has led to the debate on whether the origin is embryonic or neoplastic, but most authors believe the origin of this tumour to be the embryonic remnants of mesonephric tissue.<sup>4</sup>

As per our search association of benign cystic teratoma with extra renal Wilms' tumor has not yet been reported. So it may be the first case identified as a rare association of Benign Cystic Teratoma with extra renal wilms' tumor.

### Conclusion

In conclusion it may be stated that association of benign cystic teratoma of mediastinum with extrarenal Wilms' tumor is very much rare. Wilms' tumor with respiratory distress may be anything other than metastasis. Apparently looking pleural effusion is not always pleural effusion, there may be double pathology. So, the clinician's awareness of this rare association may prevent misdiagnosis and delay in appropriate treatment.

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