DISCUSSION:

Wilms' tumor, named after German surgeon Carl Max Wilhelm Wilms, is said to be possibly derived from primitive metanephric blastema[1]. Wilms' tumor, also known as nephroblastoma, is the most common pediatric renal cancer[2].

Wilms' tumor is very rare in adults with an incidence of about 0.2 per million per year and represents less than 1% of all diagnosed renal tumours[3,4]. There are approximately 300 documented cases of adult Wilms' tumor in the literature to date[5]. Because of its anatomical position, venous drainage and lymphatic drainage, it is not uncommon for distant metastasis to the lungs and liver. There is no significant histological and radiological difference between adult and childhood Wilms' tumor[2]. In fact, they share the classical triphasic histopathological features such as blastemal, epithelial, and stromal features under the microscope[2,6].

Monophasic tumors pose a significant diagnostic challenge. The index case demonstrated an epithelial predominant histology, thus included papillary RCC, Metanephric like *ALK*-rearranged RCC and metanephric adenoma as the major differentials. Negativity for KRT7, AMACR and ALK excluded the first two possibilities. Metanephric adenoma was the closest differential considering the immunoprofile of PAX8+, WT1+, BRAF+, CD57+, however, the atypia, mitotically active nature of the lesion alongwith the clinical profile of liver metastases favored a malignant pathology. CD56 and BRAF positivity supported the diagnosis of Adult Wilms' Tumor (epithelial predominant), possibly arising from a metanephric adenoma.

An analysis by Wobker et al demonstrated that *BRAF V600E* mutations were not entirely restricted to typical metanephric adenoma, as they may be seen in metanephric adenoma showing mitotic activity along with a subset of epithelial-predominant WTs in adults[7]. Argani et al summarized that a significant subset of adult Wilms' tumor (specifically those of epithelial type with differentiated areas) harbor targetable *BRAF V600E* mutations and appear to arise from metanephric adenomas as a consequence of additional acquired genetic alterations[8]. Clinical outcome for adults Wilms' tumor is inferior when compared with children, although better results are reported when treated within pediatric trials[9].