



# MiT translocation renal cell carcinoma – A subset of tumours with translocation 6p21 [t(6;11)] or Xp11.2 [t(X;1 or X or 17)]

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## Introduction

MiT translocation renal cell carcinomas (TRCC) constitute a group of recently described rare kidney tumours. These tumours predominantly occur in younger patients with only 25% affecting patients over 40 year of age. TRCC contains two main subgroups: tumours, either with translocation 6p21 [t(6;11)] or Xp11.2 [t(X;1 or X or 17)]. We present 10 cases of TRCC collected from the whole Czech Republic (10 million of inhabitants). Complete clinical, radiological and pathological data and follow-up were obtained in all cases.

## Materials & Methods

Eight cases were treated at the University Hospital in Pilsen. These cases were identified among 1653 cases of kidney tumours in adults (0.48%). Two cases treated at other hospitals were retrieved from the Pilsen Tumour registry. The pathological diagnosis of TRCCs was made on the basis of routine histopathological examination in conjunction with immunohistochemical and ultrastructural studies.

## Results

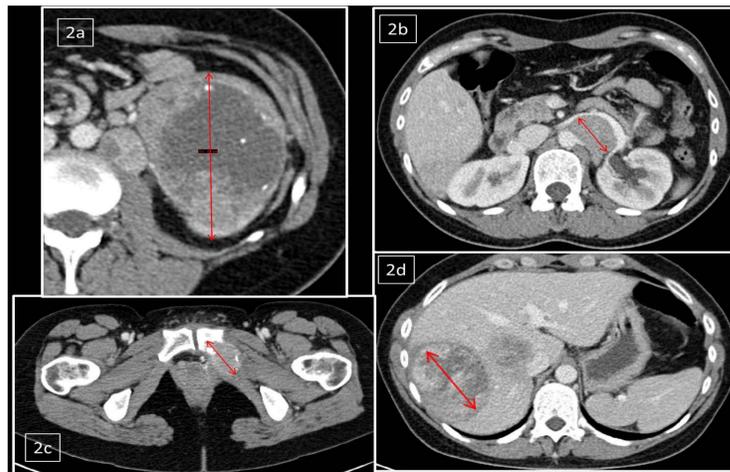
Six tumours were located on the left site and 4 on the right. Six cases were type Xp11.2 and four 6p21 TRCC. There were 7 female and 3 male patients (Xp11.2 4:2, 6p21 3:1) with a mean age of 49 years (21-80), 5 patients (50%) were over 40 years. The mean age of Xp11.2 TRCCs was 55 (median 51) and 6p21 40 (32) year. Six patients underwent radical and three partial nephrectomy and in one patient only biopsy of the tumour was performed. One female patient (24years) underwent radical nephrectomy at 4 months of pregnancy. Stage (by UICC, 7th ed. 2009) was 5x I, 3x III, 2x IV. The mean size of tumour in the greatest diameter was 80 (40-165) mm. The mean follow-up was 33.0 (1-76) months.

In patients with 6p21 TRCCs, no tumour recurrences or metastatic events were documented. In contrast, in patients with Xp11.2 tumours, 4 patients (67%) have advanced tumours – 3 succumbed due to metastatic disease (range 1 to 8 months), one with metastatic disease is stabilised on temsirolimus for 7 months. Only two patients with Xp11.2-tumours (pT1, size of tumours: 32 and 40 mm) were alive at 52 and 92 months follow-up.

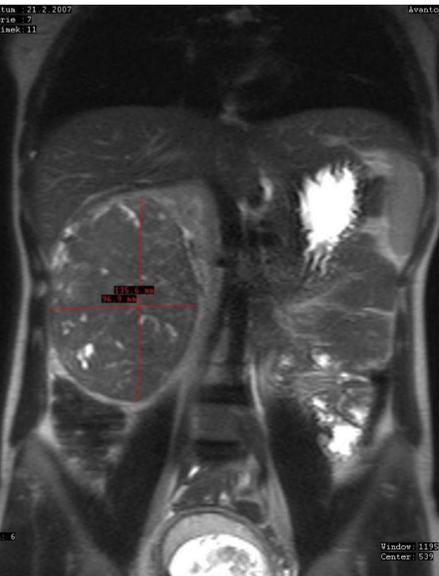
## Cases



Case 1 b, c Woman, 75 y/o, Translocation carcinoma Xp11.2, subtype ASPL-TFE3 (CT 34 mm, spheroid, 34 mm, relatively homogenous, postcontrast density (venous phase) 71 HU). Open translumbal resection was performed. A dissected specimen at operation: ochre-orange relatively homogenous



Case 2 Woman 34y/o, Translocation carcinoma Xp11.2, subtype ASPL-TFE3 of the left kidney (2a) T3aN2M1, metastases to the paraaortal lymph nodes (2b), left pubic bone (2c), liver (2d)



Case 3: Translocation carcinoma 6p21 [t(6;11)] ("Rosette Forming", HMB45 Positive). 24-year-old pregnant woman, 14th week of gravidity. CT: Tumor of the right kidney 136 mm. Open radical nephrectomy performed. without complications for the patient or the fetus. Dissected specimen at operation. Tumor was homogenous and brown. Published in Hora et al: Int Urol Nephrol, 2009, 41: 553-7

## Conclusion

In our cohort, TRCCs were more common in females. Patients with TRCC type 6p21 were younger than those with Xp11.2 TRCC. Although the two main group of TRCC are similar in etiopathogenesis, the biological behaviour is completely different. Type Xp11.2 TRCC is a very aggressive neoplasm whereas all patients with 6p21 TRCC had a benign course. From a clinical point of view, strict subclassification of TRCCs is of utmost clinical relevance.

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