MiT translocation renal cell carcinomas: two subgroups of tumours with translocations involving 6p21 [t (6; 11)] and Xp11.2 [t (X; 1 or X or 17)]

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Abstract

Introduction: MiT translocation renal cell carcinomas (TRCC) predominantly occur in younger patients with only 25% of patients being over 40 years. TRCC contains two main subgroups with translocations involving 6p21 or Xp11.2. Herein we present 10 cases.

Materials: Eight cases were treated at main author’s institution (identified among 1653 (0.48%) cases of kidney tumours in adults). Two cases were retrieved from the Pilsen (CZ) Tumour Registry.

Results: Six cases were type Xp11.2 and four 6p21; 7 female, 3 male patients; Xp11.2 4:2, 6p21 3:1. The mean age 49 years (range: 21–80), 5 patients (50%) over 40 years. The mean age of the group with Xp11.2 TRCCs was 55 (median 51) and 6p21 41 (32) years. One female with a 6p21 tumour (24 years) underwent nephrectomy at 4 months of pregnancy. Stage (UICC, 7th ed. 2009) was 5xI, 3xIII, 2xIV. The mean size of tumour was 80 (40–165) mm. The mean follow-up was 33.2 (1–92) months. In patients with 6p21 tumours, one (25%) died after 3 months due to widely metastatic disease. In patients with Xp11.2 tumours, 3 (50%) succumbed due to metastatic disease (range 1–8 months). Three patients with Xp11.2 are alive at 7, 52 and 92 months of follow-up, were diagnosed at early stage (T1a).

Conclusion: TRCCs were more common in females. Patient with 6p21 tumours were younger than those with Xp11.2. Both types have definitive malignant potential Type Xp11.2 seems to be a more aggressive neoplasm than 6p21. The case with metastatic 6p21 tumour is the 4th case described in the English literature.

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 about 25% affecting patients over 40 year of age (see Table 1). TRCC contains two main subgroups: Srigley et al. 2013 Tumours with translocation 6p21 [t (6;11)] have characteristic histopathological features and immunohistochemical properties and have been labelled “rosette forming HMB45 positive renal tumour” in addition to “TFEB RCC”. Hora et al. 2008 The second subgroup is composed of tumours with translocations involving Xp11.2 [t (X; 1 or X or 17)]. TRCC Xp11.2 is included in the 2004 WHO renal tumours classification already. In the ISUP (International Society of Urological Pathology) Vancouver classification of renal neoplasia Srigley et al. 2013 these tumours have been added as a new subgroup of RCC: “MiT family TRCC” with two subgroup – Xp11 TRCC and t (6;11) RCC.

Papers dealing with TRCCs have been published mostly by pathologists and geneticists. It is difficult and time consuming to get clinically relevant data useful for daily urological practice. We present 10 cases of TRCC collected from the whole Czech Republic (10 million of inhabitants) focusing on data important from to point of view of practicing urologists.

Material and methods

During 2001 to 2012, 1653 kidney tumours were surgically treated at the urological department of the main...
| Author      | Year  | No of patients | Age | Sex | Size | Staging (at presentation/maximal by follow-up) | Surgery | Oncological therapy | Survival |
|-------------|-------|----------------|-----|-----|------|-----------------------------------------------|---------|---------------------|----------|
| Argani      | 2007  | 28             | 28  | 0   | 37.2 | T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | UK      | in 1 case, imuno-   | 6.3 malignant vs bening course |
| Rais-Bahrami| 2007  | 1              | 6   | NA  | 0    | T1-3N1M0                                      | 0/1     | TKI, progression, death in 8 ms         |
| LaGrande    | 2007  | 1              | 63  | NA  | 1    | T4NXM0, TXN2M0, TXNXM1                         | 2 ys without sign of recurrence |
| Franzini    | 2007  | 1              | 79  | NA  | 1    |                                                | none    | metastases in 1 m |
| Hora        | 2008  | 2              | 57  | 34-80 | 1   | T2N0M0                                        | none    | one died mfor       |
| Camparo     | 2008  | 3423           | 29  | 2   | 24.6 | T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | 4/27    | FU 29.5 ms, 21 NoF, 5 died, 3 metastases, 2 lost for FU |
| Hora        | 2009  | 15000          | 0   | 3   | 28.3 | T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | 2/1     | no recurrence        |
| Suárez-Vilela| 2009 | 1              | 22.0| 2   | 0    | T4NXM0, TXN2M0, TXNXM1                         | 0/1     | no recurrence        |
| Koie        | 2009  | 1              | 28.0| NA  | 0    | T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | 0/1     | excision of local recurrence, cytokins |
| Armah       | 2009  | 1              | 26.0| NA  | 0    | T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | 1/0     | died in 24 ms        |
| Kuroda      | 2010  | 1              | 73.0| 1   | 100.0%| T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | 1/0     | pregnant, 27 ms without recurrence |
| Jing        | 2010  | 1              | 12.0| NA  | 0    | T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | 1/0     | Unknown              |
| Choueiri    | 2010  | 15             | 41.0| 18-65 | UK  | UK                                            | 0/12    | VEGF-targeted therapy |
| Ishihara    | 2011  | 1              | 45.0| NA  | 1    | T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | 0/1     | Ch,T, RT, temsirolimus    |
| Liu         | 2011  | 1              | 27.0| NA  | 0    | T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | 0/1     | metastasectomy, gemcitabin, sunitinib |
| Nelius      | 2011  | 1              | 19.0| NA  | 0    | T1N0M0, T2N0M0, T3N0M0, T1-3N1M0               | 1/0     | Ch,T, sorafenib, temsirolimus, bavacizumab, RT |

**Note:** The table includes a review of literature about MiT TRCC since 2007, with details on the number of patients, age, sex, size, staging at presentation/maximal, surgery, oncological therapy, and survival outcomes.
| Name       | Year | ID | Age  | Gender | Follow-up | Progression | Treatment | Response | Survival | Status | Other Details |
|------------|------|----|------|--------|-----------|-------------|-----------|----------|----------|--------|---------------|
| Numakura   | 2011 | 1  | 43.0 | NA     | 100.0%    | 1 : 0       | 100       | 1        | 0/1      | : 0    | sunitinib - PR for over 3 years |
| Kato       | 2011 | 1  | 18.0 | 0      | 0.0%      | 0 : 1       | 41        | 1        | 0/1      | : 0    | lung metatases 2 yrs from surgery |
| Klatte     | 2012 | 848| 2    | 0      | NA        | 50.0%       | 1 : 1      | 100      | 0/2      | : 0    | dendritics cells and interferon-alfa-2a (CR) |
| Morii      | 2012 | 1  | 38.0 | 0      | 0.0%      | 1 : 0       | 75        | 0        | 0/1      | : 0    | immunotherapy, sunitinib, sorafenib, mTOR I |
| Rao        | 2012 | 7  | 30.6 | 0      | 0.0%      | 4 : 3       | 51        | 5        | 0/1      | : 0    | died in 16 ms |
| Inamura    | 2012 | 200| 47.0 | 2      | 66.7%     | 0 : 3       | 100       | 0        | 0/1      | : 0    | UK |
| Arnoux     | 2012 | 170| 0    | 4      | 100.0%    | 1 : 0       | 100       | 0        | 0/1      | : 0    | immunotherapy, sunitinib, sorafenib, mTOR I |
| Dang       | 2012 | 9  | 29.6 | 1      | 11.1%     | 3 : 6       | 58        | UK       | 0/1      | : 0    | ChT 4 cases, RT 2 cases, 2 excision of retroperitoneal recurrence |
| Komai      | 2013 | 443| 42.0 | 5      | 71.4%     | 3 : 4       | 83        | 3        | 0/1      | : 0    | 5 of 7 (71.4%) generalisation |
| Gaillot-Durand | 2013 | 92 | 0    | UK     | UK        | UK          | UK        | UK       | 21       | 40    | UK |
| Together   | 20255| 110| 29   | UK     | UK        | UK          | UK        | UK       | 40       | 36%   | UK |
Author. Eight of them were TRCC (0.48%). Two more cases were identified in the tumour registry of the Department of Pathology, Faculty Hospital, Pilsen, CZ. The registry includes over 16,000 cases of renal tumours, of which a significant component is international consult cases. Due to easy access to clinical data including CT and to avoid ethical problems with approving the study in different countries, only cases from CZ were included. These ten cases are presented in detail. Five cases have previously been published Hora et al. 2008; Hora et al. 2009. Extended follow-up information is provided for these cases in this paper. These cases are: two TRCC Xp.11 (in Table 2 cases 2 and 5) Hora et al. 2008 and three TFEB TRCC (in Table 2 cases 1, 3, 4) Hora et al. 2009. The morphological diagnosis was supported by immunohistochemical examination. In 1 of 4 cases of Xp11.2 TRCC, morphological and immunohistochemical results were extended by FISH analysis of TFE3 break. See Table 3. In one case, the diagnosis was verified. The other 4 cases were not analysable due to low quality of DNA. Among 4 cases of 6p21, three were morphological and immunohistochemical analysis supported by FISH and RT PCR (reverse transcription polymerase chain reaction) analysis. The presence of the translocation t (6;11) (Alpha-TFEB) was confirmed in these 3 analysed cases. In one case (percutaneous biopsy only), the diagnosis was established without molecular genetic confirmation, i.e. based exclusively on morphology and results of immunohistochemistry.

Results
The results are summarised in the Table 2. Selected cases, see Figures 1, 2 and 3. Based on these, we can conclude that TRCCs were more common in females (70%). Patient with TRCC type 6p21 were younger than those with Xp11.2 TRCC (average 40.7 ± 25.8 vs. 54.6 ± 20.3 years, median 31.9 vs. 51.2). The biological behaviour of the two main group of MiT TRCC is probably different. Type Xp11.2 TRCC is a more aggressive neoplasm with (malignant course in 3/6 cases – 50%). In four patients with 6p21 TRCC, one (25%) died due to generalisation.

Discussion
MiT (F) (microphthalmia transcription factor) TRCCs most likely originate from the proximal tubule of the nephron. TRCCs are found predominantly in children and young adults, but are increasingly being recognized in adults. These tumours are characterized by the occurrence of recurrent chromosomal translocations, which result in disruption and fusion of either the TFE3 or TFEB genes, both members of the MiT family of basic helix-loop-helix/leucine-zipper transcription factor genes. Hence the name MiT translocation subgroup of RCCs. The MiT family (MiTF/TFE family) includes TFE3, TFEB, and also TFEC and MiTF transcription factors Srigley et al. 2013.

In clinical practice, we most commonly encounter TRCC TFE3 (divided in at least 6 subtypes, see Table 3) and TFEB type. We have identified 110 published cases of Xp11.2 TRCC in the English literature since 2007 to October 2013. See Table 1. The TFE3 gene is located on the X chromosome, locus p11.2. Translocations result in fusions of the TFE3 gene with several other genes which are listed in Table 3. Question The correct diagnosis is reached through a combination of histomorphology, immunohistochemistry and, in selected cases, molecular genetic studies. Since the translocations lead to overexpression of the TFE3 protein, immunohistochemical staining for TFE3 is widely used as a surrogate marker for the Xp11.2 translocation Klatte et al. 2012. For example, Komai et al. Komai et al. 2009 described 7 cases of TRCC Xp11.2, 4 of them were diagnosed cyto-genetically, 3 by immunohistochemistry only. Argani et al. 2007 confirmed genetically in group of 28 Xp11 TRCC only three. But Klatte et al. 2012 found in 17 TFE3 positive tumours Xp11.2 translocation in only 2 cases and they recommended making diagnosis of Xp11.2 translocation RCC only genetically. Other group verified Xp11.2 by RT-PCR 2 cases in 6 TFE3 positive Gaillot-Durand et al. 2013. On the other hand, TF3 negative tumours on immunohistochemistry can be FISH positive Rao et al. 2013. Our ten cases of TRCC were diagnosed by morphology and immunohistochemistry and only one verified genetically because the limitation of the quality of DNA. But we hope that modern reliable immunohistochemical staining can be very precise and more exact. Sensitivity and specificity of TFE3 tests are now being discussed by pathologists. Manual overnight labelling seems to be more precise than automated immunostainer with 30 min incubation Gaillot-Durand et al. 2013. However precise histological diagnosis verified by cytogenetic studies is missing in some cases and it can be source of bias of this study.

There is little information on clinically relevant data when dealing with patients with Xp11.2 TRCC. The tumours most commonly affect patient under 45 year up to 15% Kuroda et al. 2012. In some previous studies data on the radiological features (CT and MRI) of Xp11 TRCC have been presented. These have failed to identify any specific radiological features of this tumour Liu et al. 2013; Koo et al. 2013; Kato et al. 2011).

Xp11.2 RCC is a biologically aggressive neoplasm with a bad prognosis and previously published data suggest that the prognosis may be even worse in men over 50 years Kuroda et al. 2012; Arnoux et al. 2012. In our group of 6 cases, the ages were similar in patients with aggressive vs. with benign clinical course 52.4 vs. 56.8 years). The
| No | Age | Sex | Date of surgery | Side | Type of surgery | Note | AE | LAE | Size of tumour (mm) | Staging | Stage at diagnosis | Stage at the last follow-up | Grading by Fuhrmann | Follow-up in months |
|----|-----|-----|----------------|------|----------------|------|----|-----|-------------------|---------|-------------------|------------------------|-------------------|--------------------|
| 10 | 34.5 | 34.5 | F | 1/2/2013 | L | Biopsy under CT | T3aN2M1 - metastasis to liver, pubic bone, cisplatin and temsirolimus no effect | 0 | 0 | 96 | 3a | 2 | 1 | 4 | 4 | UK | 8 |
| 9  | 77.6 | 77.6 | F | 10/3/2012 | L | ONE, AE, LND | Cytoreductive NE of bulky tumour (1850 g), lung metastases | 1 | 1 | 165 | 3a | c1p0 | p1* | 3 | 4 | 3 | 3 |
| 8  | 75.3 | 75.3 | F | 21.11 (November) 2012 | R | OR | Followed like a cystic renal lesion, control MRI Bosniak IV, published formerly Hora et al. (2008) | 0 | 0 | 33 | 1a | 0 | 0 | 1 | 0 | 3 | 7 |
| 6  | 60.2 | 60.2 | F | 21.11 (November) 2012 | L | ONE | Multiple skeletal metastases, liver, lung | 0 | 0 | 40 | 3a | 1 | 1 | 4 | 4 | 3 | 8 |
| 7  | 42.3 | 42.3 | M | 21.11 (November) 2012 | L | ONE | Followed like a cystic renal lesion, control MRI Bosniak IV, published formerly Hora et al. (2008) | 0 | 0 | 32 | 1a | 0 | 0 | 1 | 0 | 3 | 64 |
| 3  | 39.3 | 39.3 | F | 23.10/2006 | R | OR | Open nephrectomy, published formerly Hora et al. (2009) | 0 | 0 | 128 | 3a | 0 | 0 | 3 | 1 | 64 |
| 4  | 24.4 | 24.4 | F | 6/11/2006 | R | OR | Open nephrectomy, published formerly Hora et al. (2009) | 0 | 0 | 10 | 3a | 0 | 0 | 0 | 1 | 52 |
| 5  | 80.3 | 80.3 | M | 28 Dec 2007 | L | ONE | Open nephrectomy, published formerly Hora et al. (2009) | 0 | 0 | 130 | 3a | 0 | 0 | 3 | 3 | 1 |
| 6  | 21.4 | 21.4 | M | 5/1/2005 | R | ONE | Open nephrectomy, published formerly Hora et al. (2009) | 0 | 0 | 40 | 3a | 0 | 0 | 1 | 0 | 1 | 92 |

Notes, abbreviations: Black windows – the patient died due to tumour. 
AE – adrenalectomy, ONE – open nephrectomy, LNE - laparoscopic NE, OR – open resection, LND lymph node dissection, UK unknown, STDEV.S – standard deviation, MIN – minimal value, MAX maximal value, NA – DNA from specimen not analysable due to low quality, ND – not done, pos. – positive, TRCC Translocation renal cell carcinoma. 
Cases 1, 9 and 10 see Figures 1, 2 and 3.
oncological therapy is reviewed in Table 1. The same oncological treatment protocols as for clear cell RCC have been used. No firm conclusions regarding the efficacy can be drawn based on this data. In our group, 2/3 of cases have a fatal course in relatively short follow-up.

The other (rare) but more commonly encountered MiT TRCC is “CC with t (6:11) translocation” (abbreviated “6p21 TRCC” or “t (6:11) TRCC”). Owing to the characteristic histopathological features and immunohistochemical properties of this tumour, it has been termed “osette-like forming, HMB45-positive renal tumour” Hora et al. 2009. An alternative designation/term that is used is “TFEB RCC”. This tumour harbours translocations involving the transcription factor EB (TFEB) and Alpha (the latter also known as MALATI). Genetically, TFEB RCC has been characterized by the fusion of the 5’ portion of Alpha, also known as MALATI (Genbank accession number AF203815), an intronless gene mapped at 11q12, with TFEB at 6p21 Inamura et al. 2012; Rao et al. 2012. TFEB RCC is extremely rare, with fewer than 30 cases reported to date Hora et al. 2009; Inamura et al. 2012; Rao et al. 2012; Camparo et al. 2008; Suarez-Vilela et al. 2011. First 11 cases were reviewed by Hora et al. 2009, cases published since 2007 are summarised in Table 1. The morphology of TFEB TRCC is distinctive and the diagnosis can be established based on a combination of histopathological examination in conjunction with immunohistochemistry. Role of molecular genetic studies is not as crucial as in Xp11.2 TRCC Hora et al. 2009. The malignant potential is low. Only three cases

Table 3 Types of translocation renal cell carcinoma
Xp11.2 Hora et al. (2008)

| Translocation       | Fusion of genes                     |
|---------------------|-------------------------------------|
| t (X;1) (p11.2; q21)| PRCC and TFE3                       |
| t (X;1) (p11.2; p34)| PSF and TFE3                       |
| t (X;17) (p11.2; q25)| ASPL (known as RCC17 or ASPSCR1 as well) and TFE3 |
| t (X;17) (p11.2; q23)| CLTC (Clathrine) and TFE3          |
| inv (X) (p11.2; q12)| NonO (p54cre) and TFE3             |
| T (X;3) (p11.2; q23)| Unknown                             |

Figure 1 Dissected specimen at operation: woman, 75-year-old, a tumour of the right kidney. In table case No. 5. Tumour was on CT spheroid, 34 mm in maximal diameter, relatively homogenous, native density 13–26HU, postcontrast density (venous phase) 15–43 HU. R was performed. A dissected specimen at operation: ochre-orange relatively homogenous spheroid tumour very different from clear renal cell carcinoma, maybe a little similar to any papillary RCC. Histological diagnosis: Translocation carcinoma Xp11.2, subtype ASPL-TFE3, verified genetically.
Figure 2 Woman 34-year-old, tumour of the left kidney (a) T3aN2M1, metastases to the paraaortal lymph nodes (b), left pubic bone (c), liver (d). Biopsy Translocation carcinoma Xp11.2. In Table 2 case No. 10.

Figure 3 Postcontrast CT: woman 78-year-old, left kidney tumour T3aN0M1 (metastases to lung), maximal diameter of tumour 172 mm. In Table 2, case No. 9. She underwent cytoreductive nephrectomy, specimen 1850 g. Histology TRCC 6p21. She died in 3 months. The forth case of aggressive TRCC 6p21 described in literature.
with aggressive behaviour have been published (approximately 10%) Inamura et al. 2012; Pecciarini et al. 2007; Ishihara et al. 2011. We add one more case with aggressive clinical course.

**Conclusion**

TRCCs were more common in females. Patient with 6p21 were younger than those with Xp11.2. Both types have definitive malignant potential, type Xp11.2 TRCC seem to be more aggressive neoplasm. Metastatic 6p21 is 4th case described in literature. From a clinical point of view, subclassification of TRCCs is of utmost clinical relevance.

Citations of articles used in the table: Argani et al. 2007, Rais-Bahrami et al. 2007, LaGrange et al. 2007, Franzini et al. 2007, Hora et al. 2008, Camparo et al. 2008, Hora et al. 2009, Suarez-Vilela et al. 2011, Koie et al. 2009, Armah et al. 2009, Kuroda et al. 2010, Choueiri et al. 2010, Ishihara et al. 2011, Liu et al. 2011, Nelius et al. 2011, Numakura et al. 2011, Kato et al. 2011, Klatte et al. 2012, Morit et al. 2012, Rao et al. 2012, Inamura et al. 2012, Arnoux et al. 2012, Komai et al. 2009, Gaillot-Durand et al. 2013.

**Abbreviations**

ChT: Chemotherapy; m (s): month(-s); ImTOR: Inhibitors (temsirolimus evelorilimus); NED: No evidence of disease; NoR: Nor recurrence or metastasis; PR: Partial response; RT: Radiotherapy; TKI: Tyrosine kinase inhibitors; Y (s): Year (-s).

**Competing interests**

The authors declare that they have no conflict of interests.

**Authors' contributions**

MH: main author of text. MH, UT, TI: collecting of clinical data. FJ, CZ: collecting of radiological data. WM, MM, PP, KN, HO: pathological reviewers of cases. MH, OH: authors of idea of article. PF: proofing of English text. All authors read and approved the final manuscript.

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