

# Primary Ewing's Sarcoma of the Kidney: A Case Report

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## ABSTRACT

*Primary adult Ewing's sarcoma is a rare entity. They most commonly occur in children and young adults. 6% of them are localized extraosseously. We present a case of a 51 year old patient with primary renal Ewing's sarcoma and multiple metastases in liver and iliac bone. Patients with metastatic disease are usually treated with aggressive chemotherapy and have a poor outcome. Our patient underwent complete surgical excision of tumour, and was treated with aggressive chemotherapy, respectively. Two and half years after presentation he is well, without any symptoms.*

**Key words:** Ewing's sarcoma, complete surgical excision

## Introduction

Ewing's family of tumours includes Ewing's sarcoma of bone, extraosseous Ewing's sarcoma, and primitive neuro-ectodermal tumour (PNET). It primarily arises in bones, most commonly in children and young adults. They are extraosseous in approximately 6% of cases. The most common sites of extraosseous Ewing's sarcomas are the trunk, extremity, head and neck, and retroperitoneum<sup>1,2</sup>. Ewing's sarcoma of primary renal origin is a rare entity in the adult population and often has an aggressive course<sup>3,4</sup>. According to the WHO, 2004 (Tumours of the urinary system and male genital organs) 35 cases of renal PNET/EWS were recorded. The mean age of presentation was 27 years with predilection for males (21 males, 14 females). The principles of management of extraskeletal Ewing's tumour have been extrapolated from the experience of treating osseous Ewing's sarcoma or from high grade sarcomas of the same site and stage, though some studies have suggested that extraosseous Ewing's sarcoma may have poorer prognosis<sup>2,6</sup>. Literature regarding primary renal Ewing's sarcoma in adults is limited to case reports or short case series. No definite

recommendations regarding optimal treatment have been defined<sup>5-8</sup>.

A single institution experience of the clinical presentation, management and outcome of this rare tumour is presented here.

## Clinical History

A 51 year old man was admitted to our hospital with severe and persistent pain in the right leg over the last two month. On physical examination impossibility for plantar and dorsal flexion of the foot was detected. His past medical history was almost unremarkable except for cerebral meningioma which was surgically resected 6 years ago with two recurrences after 2 and 3 years. Ultrasound examination and abdominal computer tomography (CT) revealed expansive tumour of the right kidney extending to and compressing psoas muscle. In the second segment of the left liver lobe, metastatic tumour mass was also found. Right wing of the iliac bone was infiltrated with expansive tumour mass.

## Materials and Methods

A fine needle aspiration biopsy (FNAB) of the iliacal tumour mass was performed, stained with May-Grunwald-Giemsa (MGG) and revealed poorly differentiated malignant cells, with some cytoplasm displaying discrete vacuolization and nuclei with coarse chromatin structure (Figure 1). Most of them had degenerative changes. A lot of naked oval and ovoid malignant nuclei were also seen. Immunocytochemical analysis revealed weak and nonspecific positivity for EMA and vimentin. It was diagnosed as metastatic epithelial tumour, of the possible renal origin.

Radical tumourectomy was performed with complete resection of the right kidney, right liver lobe (2. and 3. segment) and complete resection of the iliacal tumour mass (Figure 2 and 3). Intraoperative exploration of the liver detected 2 more metastases in the 4b. and 5. segment. On a cross section of the right kidney which

measured 14:8:4 cm, in the upper pole, a circumscribed, yellow-greyish tumour with areas of haemorrhage, measuring 5:5.5:3.7 cm was found. In the middle part of the kidney another tumour with identical gross characteristics as the previously described one, was also found. It measured 2:2: 1.5 cm. Both tumours neither invaded pelvis nor blood vessels. Microscopic examination of the excised specimen showed sheets, islands and lobules of uniform cells with round nuclei, dispersed chromatin pattern and prominent nucleoli (Figure 4). The immunohistochemical analysis revealed vimentin, CD99 (Figure 5) and NSE (Figure 6) positivity. Three metastases measuring 8 cm, 8 and 7 mm were found in the resected liver segments. Iliacal tumour mass measured 14:10:3 cm. All of them showed identical histologic picture. The clinical presentation, histological pattern, cytological characteristics and the cellular immunophenotype addressed the diagnosis towards Ewing's sarcoma (by F. Knežević). Ewing's sarcoma has been confirmed using reverse tran-

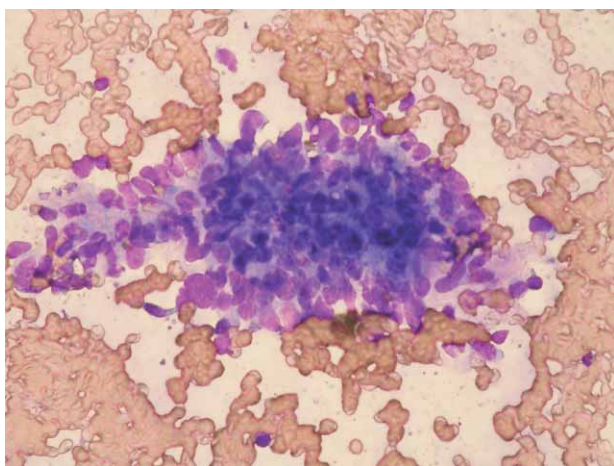


Fig. 1. Cytological analysis detected accumulations of poorly differentiated malignant cells with coarse chromatin structure in nuclei.

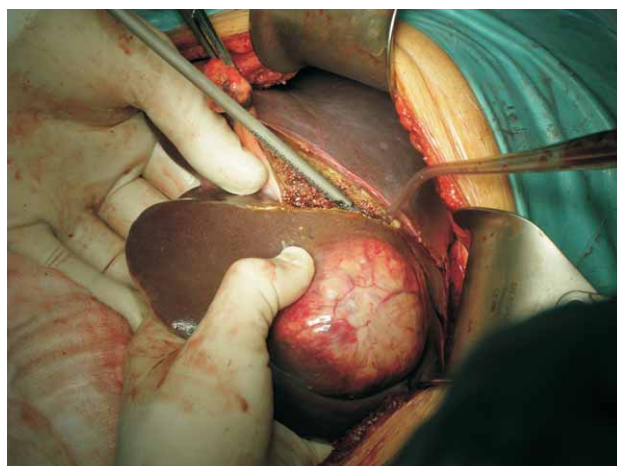


Fig. 3. Liver metastasis measuring 8 cm.



Fig. 2. Complete resection of the right kidney which measured 14:8:4 cm.

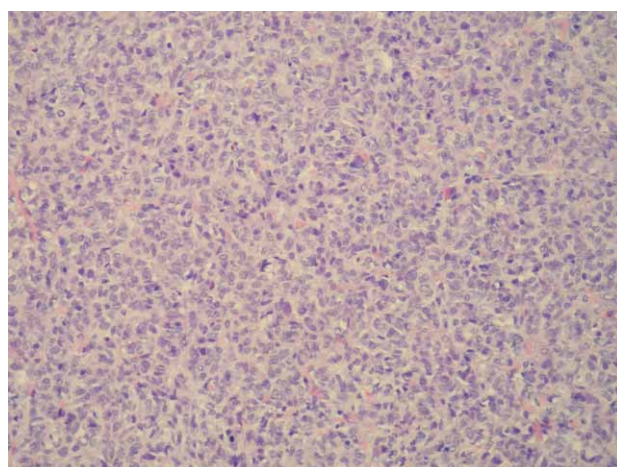


Fig. 4. The tumour is composed of uniform cells with round nuclei, dispersed chromatin pattern and prominent nucleoli. Tumour cells form sheets, islands and lobules.

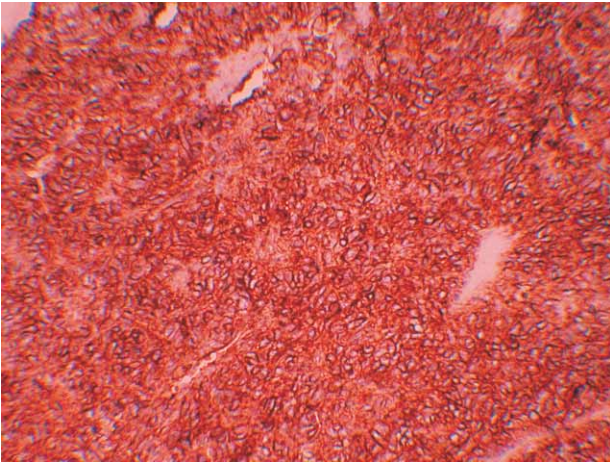


Fig. 5. Immunohistochemical evaluation revealed CD99 positivity.

scription PCR analysis where tumour specific translocation t(11;22) and fusion gene EWS/ FLI-1 was found.

The patient received 6 cycles of chemotherapy in the last two years according to the VAIA protocol which included Uromitexan (4600 mg i.v. u 1000 ml fiz.ot/24h), Vinkristin (2 mg i.v.), Holoxan (3650 mg i.v. u 1000 ml fiz ot/24h), Kytril (1 amp i.v.) Daktinomycin (0.9 mg i.v.). The patient is well, without any symptoms two and half years after the operation.

## Discussion

Ewing's sarcoma (EWS)/ peripheral primitive neuroektodermal tumour (PNET) and small round cell tumours occurring in bone and soft tissues characterised as

a group by the presence of the typical translocation t(11;22) (q24;q12) and its variants<sup>9</sup>. A total of approximately 65 primary renal cases have been reported to date<sup>10</sup>. In practice, all Ewing's family tumours are treated in the same fashion using a combination of local therapy and aggressive combination chemotherapy. There is no absolute protocol or treatment for Ewing/PNET owing to its rarity. The prognosis is poor with a 5 year disease free survival rate of about 50% despite multimodal therapy.

A retrospective study was conducted at the Royal Marsden Hospital from January 1993-December 2007<sup>11</sup>. The study included 7 patients (range 25–57 years, both sexes) 4 of them with nonmetastatic disease and 3 with metastatic disease at presentation. They were all treated with aggressive chemotherapy. The median follow-up was 36 month. None of the patients with metastatic disease at presentation had a durable response to chemotherapy and all had a short survival. They all died in the interval of 1 to 5 years after the tumour was diagnosed. The oldest patient (50 years old) had the shortest survival. It should be pointed out that none of the patients with metastatic disease underwent radical nephrectomy. On contrary, all of the patients with localised disease underwent radical nephrectomy and received chemotherapy. They had much better survival. Our patient, in spite of metastatic disease, underwent radical nephrectomy with complete excision of all the metastases, received aggressive chemotherapy and is two and half years after the presentation well and without any signs of recurrence. Some reports suggest that long-term survival of patients with renal Ewing's sarcoma could be due to complete surgery, especially radical nephrectomy<sup>12</sup>. Our experience confirms this hypothesis.

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## **PRIMARNI EWINGOV SARKOM BUBREGA: PRIKAZ SLUČAJA**

### **S A Ž E T A K**

Primarni Ewingov sarkom odraslih osoba je vrlo rijedak. Najčešće se pojavlju u dječjoj i mlađoj adolescentnoj dobi. 6% ih je smješteno izvan kostiju. Prikazujemo slučaj 51 godišnjeg bolesnika sa primarnim Ewingovim sarkomom bubrega i sa više metastaza u jetri i ilijačnoj kosti. Pacijenti sa metastatskom bolesti se obično tretiraju agresivnom kemoterapijom i imaju lošu prognozu. Našem je pacijentu kompletno kirurški odstranjen tumor, a tek nakon toga je bio liječen agresivnom kemoterapijom. 2 i pol godine nakon početka liječenja, pacijent je dobro, bez ikakvih simptoma.