

# Inflammatory Malignant Fibrous Histiocytoma of the Retroperitoneum

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## ABSTRACT

*The authors present a case of inflammatory malignant fibrous histiocytoma located in the left retroperitoneum. The tumor was resected enblock with kidney and suprarenal gland. During the resection the system of retractors called the pillars of Kocman was used which allowed wide exposure of the abdominal cavity. The tumor measured 23x17x10cm with the left kidney and suprarenal incorporated. The tumor was centrally pseudocystic made of xanthomatous cells, foamy cells and rare giant cells with storiform formations and infiltrated with neutrophils. Immunohistochemically, the tumor cells were vimentin and CD 68 positive and CD 20, CD3, EMA, S-100, HMB 45, CD 34 and CD 1a negative. Neutrophils were CD 15 positive.*

**Key words:** large sarcomas, surgery, retroperitoneal tumors

## Introduction

Soft tissue sarcomas are a heterogeneous group of rare tumors that account for approximately 1% of human cancers<sup>1,2</sup>. These tumors occur most commonly in the extremity (50%) and second most commonly in the peritoneum (20% of the cases). Prognosis for retroperitoneal sarcomas is considerably worse than for extremity tumors<sup>3</sup>. Many patients will die because of either metastatic disease or locally recurrent disease<sup>2</sup>. Primary sarcomas of the retroperitoneum are rare neoplasms. The most common are leiomyosarcoma, liposarcoma, and fibrosarcoma. The other sarcomas reported at the site are malignant fibrous histiocytoma, hemangiopericytoma, osteosarcoma, rhabdomyosarcoma, angiosarcoma, malignant peripheral nerve sheath tumor and synovial sarcoma.

Retroperitoneal position, size of the tumor and invasion of the local structures makes surgical resection difficult or impossible. Current chemotherapy for retroperitoneal sarcomas is not effective, and radiation is limited by toxicity to adjacent intrabdominal structures<sup>4</sup>. Therefore complete surgical resection remains the most effective modality for selected primary and recurrent disease<sup>5</sup>.

## Clinical History

A 79-year-old woman was examined because of erosive gastritis and microcytic anaemia. The patient complained of weakness and diffuse abdominal pain. Physical examination showed large tumor mass palpable in the left upper abdomen. Abdominal ultrasound showed displaced left kidney with large solid partially cystic tumor mass. Abdominal computed tomography (CT) revealed a large tumor, measured 14 cm in the largest diameter that displaced the kidney. It showed neither infiltration of the renal artery and vein nor infiltration of the abdominal aorta. The other kidney, liver and pancreas were without changes.

## Material and Methods

The radical tumorectomy was performed with enblock resection of the kidney and suprarenal gland. The spleen, pancreas, colon, small bowel and aorta showed no signs of infiltration. After laparotomic incision, wide exposure was secured with system of retractors originally designed by dr. Kocman. It consists of two S shaped pillars, a bar and two hooks (Figure 1). The two S shaped pillars are connected with a bar in order to achieve a sta-

ble triangular form. The hooks are wide and deep enough to hold and elevate the ribs. Additionally a third hook may be inserted in the middle and attached to a bar. The



Fig. 1. The pillars of Kocman.



Fig. 2. The retracting system.

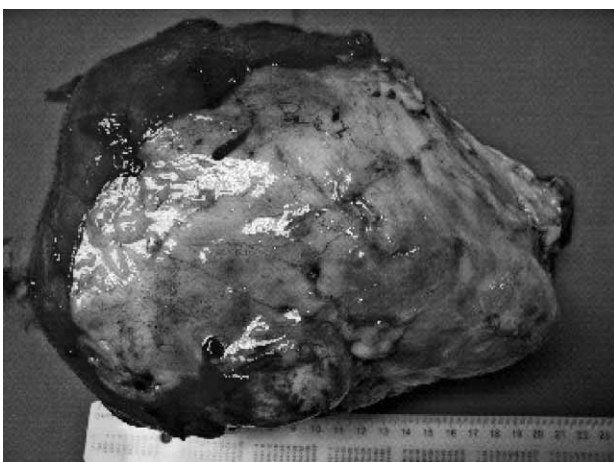


Fig. 3. Partially encapsulated tumor.

hooks are placed in their position and secured simply with a bandages and Kocher instruments (Figure 2). The system was developed for the procedures such as resection of the liver or liver transplantation and now used whenever large exposure of the abdomen is necessary.

After large laparotomy the tumor was harvested and freed from the surrounding bowel, spleen and pancreas. It was moved away from the aorta, renal artery and vein were isolated, clamped and resected. The tumor was partially encapsulated with remains of the fatty tissue. It measured 23x17x10 cm (Figure 3). On a gross section of the tumor mass the suprarenal gland was without macroscopic or histological changes. Embedded kidney measured 14x6x3 cm and was also without changes. The tumor did not infiltrate kidney or suprarenal gland. It was yellow in color, solid, centrally pseudocystic.

The material obtained from surgery was fixed in formalin and embedded in paraffin for routine histopathologic processing and immunohistochemical study. The cells were xanthomatous, foamy and rarely giant. There were sporadic storiform formations and in between abundant infiltration of neutrophils (Figure 4). The tumor cells were immunohistochemically vimentin and CD 68

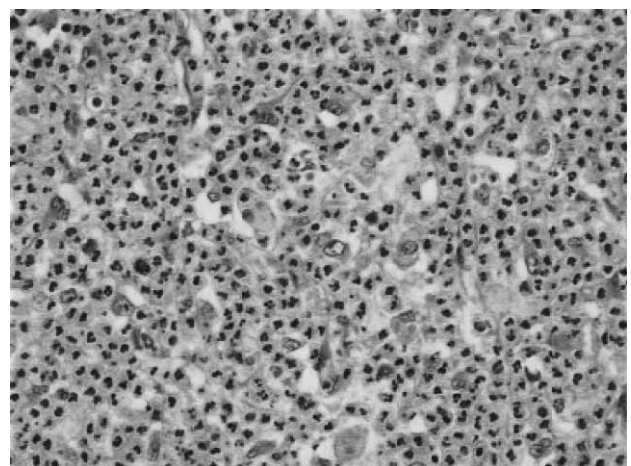


Fig. 4. The tumor cells with neutrophills.

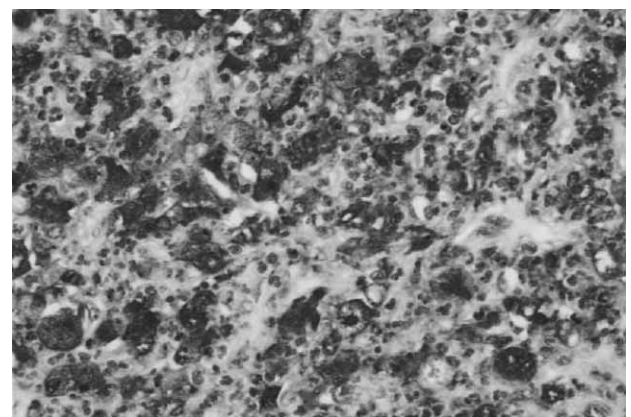


Fig. 5. CD 68 positive cells.

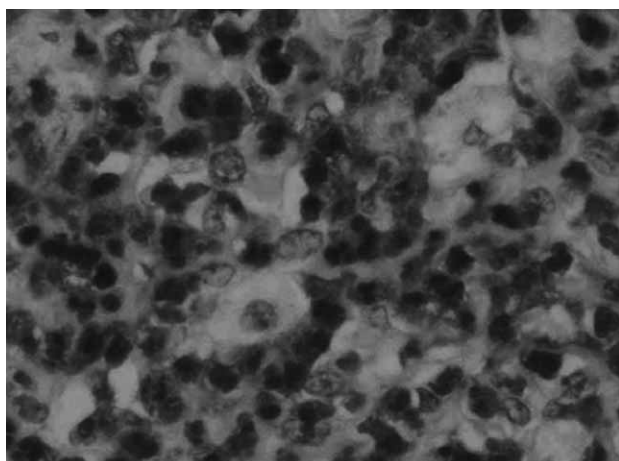


Fig. 6. CD 15 positive neutrophils.

(Figure 5) positive and CD 20, CD3, EMA, S-100, HMB 45, CD 34 and CD 1a negative. Neutrophils were CD 15 positive (Figure 6).

## Discussion

Sarcomas are malignant tumors arising in mesenchymal tissue. Sarcoma comes from a Greek word (sar = fleshy) because they have little connective tissue stroma and so are fleshy.

Many authors investigate prognostic variables in patients with soft tissue sarcoma. Understanding prognostic variables is important for counseling patients, selecting patients for adjuvant therapy and setting goals for patient treatment. Incomplete resection, unresectability, and high grade are the factors most consistently shown to be prognostic for poor survival in primary retroperitoneal soft tissue sarcoma<sup>5-9</sup>.

Lewis et al. analyzed correlation of tumor biology and surgical treatment with subsequent local recurrence, metastasis, and disease specific survival<sup>5</sup>. The focus was to determine the role of surgical resection in the management of primary and locally recurrent disease. The authors treated 500 patients with retroperitoneal soft tissue sarcoma by means of surgery, radiation and

chemotherapy. Resection of the tumor was done for 83% of the patients with primary disease. Among them only 80% underwent complete resection with an adequate margin of normal tissue. In conclusion out of 278 patients with primary retroperitoneal sarcoma, 185 patients or 67% underwent complete resection with an adequate margin of normal tissue. As expected, patients with unresectable disease or with incomplete resection had shorter life expectancy compared to patients with complete tumor resection. The median survival of patients undergoing incomplete resection was no different from survival of the patients with no resection at all. Some authors have reported similar rates of resection<sup>4,10,11</sup>, and some even lower<sup>12,13</sup>.

Clear operative field with enough room for harvesting is important to approach large tumor in a limited space. The Kocman's system of the retractors is a simple one that allows wide exposure of the abdominal cavity. It also makes the second assistant unnecessary or allows him to participate in the operative procedure more actively.

Histologically, these tumors are composed of benign and malignant-appearing xanthoma cells, the later often assuming a gigantic size with bizarre nuclei. Typically, these neoplastic cells display phagocytosis of neutrophils, a feature that helps distinguish them from anaplastic lymphoma. The neoplastic cells in malignant fibrous histiocytoma express vimentin (antibody that labels a wide variety of cells of mesenchymal origin) but not various leukocyte lineage markers (CD 15, CD 20, CD 45) although they may contain CD 68 (labels human monocytes and macrophages) as a reflection of their phagocytic activity<sup>14</sup>.

As the cytogenetic and molecular analysis progress, numerous chromosomal changes have been demonstrated in numerous malignant and even in some benign tumors. Chromosomal study, cytogenetics and molecular analysis may become increasingly useful in the diagnosis of tumor, especially soft tissue tumors and may help in our understanding of differentiation pathways of tumor<sup>15</sup>.

In conclusion surgical excision is the first choice of treatment for malignant fibrous histiocytoma. Even for the large tumor, ablation is possible without resection of the spleen or any portion of the pancreas. Adequate retracting system makes the procedure easier to perform.

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## **UPALNI MALIGNI FIBROZNI HISTIOCITOM RETROPERITONEUMA**

### **S A Ž E T A K**

Želimo prikazati slučaj malignog fibroznog histiocitoma koji je bio smješten u lijevom retroperitoneumu. Tumor je bio izvađen u bloku sa bubregom i nadbubrežnom žlijezdom. Tijekom resekcije tumora, bio je korišten sistem retraktora kojeg smo nazvali »Kocmanove kuke« a omogućavao je širok pristup u abdominalnu šupljinu. Tumor je mjerio 23:17:10. skupa sa lijevim bubregom i nadbubrežnom žlijezdom. Tumor je makroskopski bio žut, čvrst a u središnjem dijelu pseudocističan. Histološki je građen od ksantomskih, pjenušavih i rijetkih gigantocelularnih stanica uz stvaranje storiformnih formacija. Između se vidi obilan infiltrat neutrofilnih granulocita. Imunohistokemijski, tumorske stanice su pozitivne na vimentin, CD 68, a negativne na CD 20, CD 3, EMA, S-100, HMB 45, CD 34 i CD 1a negativne. Neutrofilni su CD 15 pozitivni.