

## Retroperitoneal Malignant Fibrous Histiocytoma

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## 1. Introduction

Malignant Fibrous Histiocytoma (MFH), which is also termed undifferentiated pleomorphic sarcoma or pleomorphic Spindle Cell Sarcoma (PSCS), is a type of malignant sarcoma that occurs most frequently in patients aged between 50 and 70 years. MFH occurs most commonly in the extremities and the trunk, and it is extremely rare in the retroperitoneum. The tumor is of rare, connective tissue origin with the possibility of occurrence in the retroperitoneal space tumor reached an uncommonly significant size, partly because diagnosis is difficult [1].

## 2. Case report

48 years old man was admitted to Pauls Stradins Clinical University Hospital in 2018 with complaints of severe thirst, fatigue, and episodes of subfebrile temperature. In last 6 months patient had lost weight about 10 kg. After tests and examination patient was diagnosed with decompensated primary diabetes based on following laboratory results – blood glucose 17.7 mmol/l, HbA1c 13.1% and CRP 286 mg/l. Patient started treatment with small units of intermediate-acting insulin analog, which lead to episodes of hypoglycemia, so insulin treatment was canceled. Blood glucose at the time was 4.3 – 11.1 mmol/l, C-peptide < 0.28 ng/ml.

Patient was heavy smoker for last 40 years and drank alcohol (beer) daily. His grandmother's sister had an oncological disease. There was no diabetes in family's history. Patient worked as a carpenter.

Patient underwent abdominal US that showed a large mass of the right adrenal gland. CT scan found a large mass of the retroperi-

toneal space arising from the right adrenal gland. The mass was suspected to be a liposarcoma (10 cm), so patient was sent to a surgeon (Figure 1).

In September 2018 patient had undergone lumbotomy with partial tumor excision. The resected tumor was 12 x 4 x 2 cm in size. Pathological report showed extensive necrosis, pleomorphic regions, multiple anaplastic histiocytic cells, giant cells un various atypical polynuclear giant cells with high mitotic activity (> 10 mitotic figures). Immunohistochemistry laboratory results: CD68 +, S-100 +, Vimentin +, Ki-67 65-70%. Final diagnosis was malignant fibrous histiocytoma, Grade 3.

After surgery patient received first line chemotherapy with Doxorubicin and Ifosfamide. (Table 1) After 3 chemotherapy cycles CT scan showed progression of tumor - 10 cm large tumor of the right adrenal gland with lipoid tissue infiltration incorporating right kidney.

Patient received second line of chemotherapy with Dacarbazine. After 3 cycles CT scan was performed. CT showed tumor progression 10 → 13 cm. Patient still was in good shape, without complains. After second progression he continued third line chemotherapy with Gemcitabine and Docetaxel. Third line chemotherapy was without effect. Tumor enlarged from 13 → 15 cm. (Table 1) At the end of third line chemotherapy patient started complain about pain and discomfort in the abdomen, nausea, vomiting and loss of appetite.

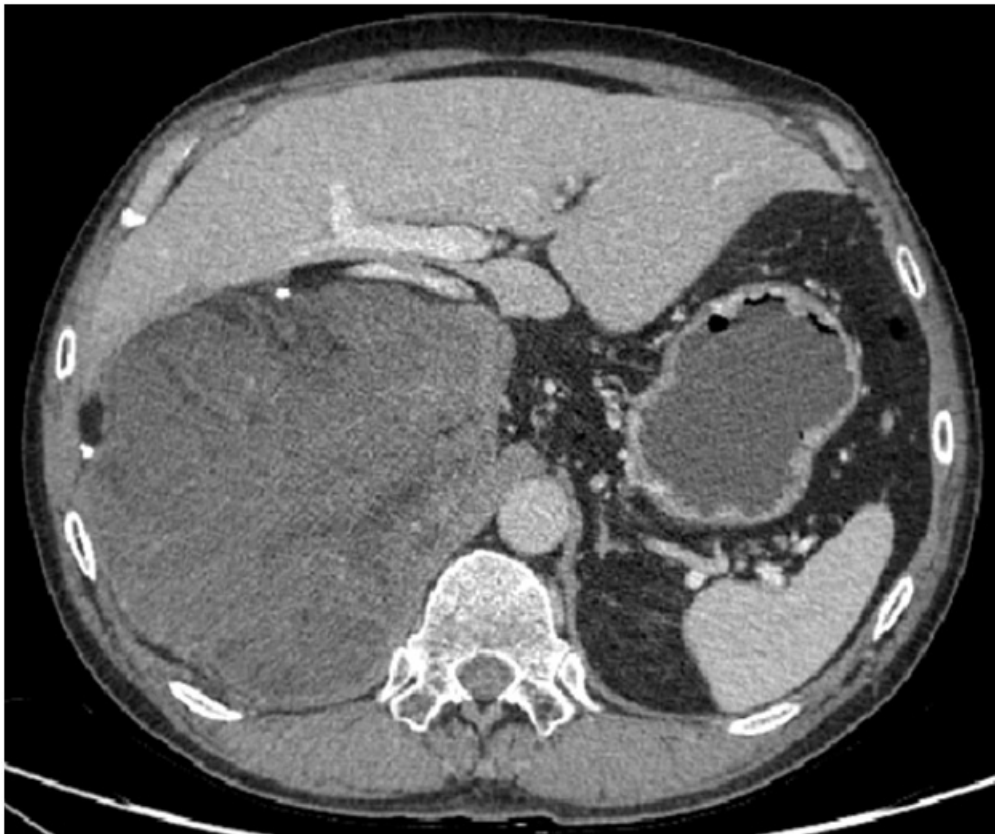
Due to symptoms and ineffective third line treatment in Novem-

ber 2019 patient repeatedly underwent lumbotomy and excision of the retroperitoneal tumor. During operation tumor was found to be closely adherent to the diaphragm, pleura and back muscles. (Figure 2) Tumor was removed partially along its metastasis from the mesentery. No additional treatment was given.

After half a year on CT scan was found multiple new pathological nodes in the both sides of the retroperitoneal space, more extensively on the left side along stomach large curvature and in the right renal region, infiltrating stomach, right kidney and basal lobes on the right side of liver (Figure 3).



**Figure 1:** CT scan before first surgery. A large mass of the retroperitoneal space arising from the right adrenal gland



**Figure 2:** CT scan before second surgery. Tumor closely adherent to the diaphragm, pleura and back muscles



**Figure 3:** CT scan before third surgery. Multiple new pathological nodes in the both sides of the retroperitoneal space, more extensively on the left side along stomach large curvature and in the right renal region, infiltrating stomach, right kidney and basal lobes on the right side of liver

**Table 1:** Chemotherapy sequence

Chemotherapy line	Chemotherapy drugs	Tumor size (at the beginning of treatment)
1 <sup>st</sup> line chemotherapy – 3 cycles	S. Ifosfamid 1500 mg/m <sup>2</sup> for 4 days S. Doxorubicini 20 mg/m <sup>2</sup> for 3 days	10 cm
2 <sup>nd</sup> line chemotherapy – 3 cycles	S. Dacarbazine 1000 mg/m <sup>2</sup> for 1 day	13 cm
3 <sup>rd</sup> line chemotherapy – 3 cycles	S. Gemcitabine 1000 mg/m <sup>2</sup> for 1 day S. Docetaxel 80 mg/m <sup>2</sup> for 1 day	15 cm

On October 2020 patient underwent third tumor resection operation with right side nephrectomy and splenectomy.

After one month he was diagnosed with progressive tumor growth with extensive metastatic process and dissemination in the retroperitoneal and intraperitoneal spaces along with multiple nodes in various sizes.

Since all treatment possibilities were exhausted patient tumor specimen was tested using OncoDNA test. OncoDNA test showed sensitivity for CDK4 inhibitors. Patient started treatment with CDK4 inhibitors–Ribociclib. Unfortunately patient at that moment was already weak and died shortly due to tumor progression.

### 3. Conclusion

Histiocytoma is a rare and aggressive tumor that is insensitive to traditional chemotherapy regimens that are recommended by

guidelines. Patients with this diagnose should be considered for alternative solutions and modern treatment possibilities. Gene panel sequencing, immunohistochemistry assays and other unique and specific biomarker tests can be very helpful in choosing the right treatment for each patient individually.

### References:

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