

CASE REPORT

ISOLATED MALIGNANT PERIPHERAL NERVE SHEATH TUMOR OF KIDNEY CAPSULE

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Abstract: The occurrence of an isolated malignant peripheral nerve sheath tumor (MPNST) of the kidney capsule is extremely rare and its presence may only be expressed by an insidious onset of non-specific and misleading symptoms with a predominance of lower back pain. A computer tomography (CT) scan (as the imaging procedure of choice) will demonstrate the tumor location and its relation to the surrounding structures. Tumor excision *in toto* is considered the treatment of choice, but it can be hazardous, especially if the tumor is adhering to the surrounding structures. Severe bleeding complications due to the damage of venous structures have to be considered, and establishing hemostasis may pose considerable difficulties. Therefore surgery should be attempted with full precautions and the patient must receive preoperative counseling. If malignancy can safely be excluded, a laparoscopic excision should be considered as an alternative treatment as recurrence is unlikely. Definition of the originating nerve might not always be possible, and a minor degree of neurological impairment has therefore to be anticipated.

A case of an isolated MPNST of the kidney capsule without neurofibromatosis is presented. The tumor was located in the fatty and fibrous capsule. It was surgically removed. The patient was further managed with radiotherapy and chemotherapy. An MPNST in such a location is very unusual.

Key words: Malignant peripheral nerve sheath tumor (MPNST), Computer tomography (CT) scan, Retroperitoneum, Surgical complications.

Introduction

Malignant peripheral nerve sheath tumor (MPNST) is a rare tumor derived from Schwann cells or pluripotent cells of the neural crest [1]. The World

Health Organization (WHO) coined the term MPNST replacing previous heterogeneous and often confusing terms, such as malignant schwannoma, malignant neurilemmoma, and neurofibrosarcoma for tumors of neurogenic origin and similar biological behavior [2]. It arises frequently in patients with neurofibromatosis, type I (NF1) [3]. It most frequently affects patients who are between 20 and 50 years old and represents 5-10% of soft-tissue sarcomas. Its location in the retroperitoneum in a patient without neurofibromatosis is an exceedingly rare occurrence. Resection and combination chemotherapy are the accepted modes of treatment [4]. There are many histological variants, namely melanotic, epithelioid and a divergent variant with epithelial, glandular and mesenchymal differentiation.

Case report

A 65-year-old female patient presented with dull, constant pain in her abdomen for the last 2 months. It was moderate in intensity and occurred towards the left side of abdomen. It was associated with backache and nausea. There was also history of weight loss. On palpation, an abdominal mass of soft consistency was found left of the midline. Bowels were normal and there was nothing to suggest any episode of bowel or urinary tract obstruction. Her appetite was good and she did not report any chronic systemic illness. On examination, the patient was elderly, weighing 60 kg, and of average build and nourishment. She suffered insulin dependent diabetes mellitus (IDDM). Vital parameters were normal and there was no icterus or pallor. The blood analysis showed a hemoglobin (Hb) level of 97 g/L, ESR 40 mm after the end of 1 hour, total leukocyte count $8.2 \times 10^9/L$ and glycemia was 10.14 mmol/L. Total bilirubin, alkaline phosphatase, serum albumin, renal profile, serum urea and electrolytes were normal. Antibodies to hepatitis C virus (anti HCV) were negative. An ultrasound of the abdomen showed a mixed hiper and hypoechoic mass deep in the abdomen, located to the left of the spine, anterior to the psoas muscle in close contact with the left kidney. It was respiratory mobile. Areas of necrosis were seen. The mass was displacing the bowel loops without infiltrating them. However, the mass was infiltrating the retroperitoneal fat and renal capsule. A CT scan of the abdomen (Figure 1) revealed a large well-defined 20×20 cm multinodular and heterogeneous mass arising from the retroperitoneum and infiltrating the left psoas as well as the left kidney capsule. It was not possible to distinguish whether its primary origin was from the kidney. It was not infiltrating the surrounding gut. An intravenous pyelography was performed which showed a lateral dislocation of the left kidney with no invasion in renal calices, pelvis or ureter. Complete resection of the tumor was performed. The operation

went well with no major blood loss (Figure 2). Post-operative recovery was uneventful and the patient is doing well, pending further follow-up. She was put on palliative chemotherapy with alternating cycles of cyclophosphamide/doxorubicin and cisplatin/etoposide. Six months after the operation the patient is still doing well; ultrasonography and a contrast-enhanced CT scan did not reveal any recurrence of the tumor (Figure 3). Histopathology of the resected mass revealed a left nephrectomy specimen with fibrous and fat kidney capsule weighing 1820 g and measuring $20 \times 15.5 \times 11$ cm (Figure 4). After removing the fat and fibrous capsule, a process easily performed, the kidney weighed 120 g, measuring $13 \times 4 \times 6$ cm with normal macroscopic morphology. However, the capsule contained a large tumor mass that encased the whole kidney but did not infiltrate the kidney. On cut surface the tumor was gray-white with a sarcomatous (fish-flesh) (Figure 5) appearance with some necrotic areas. On routine hematoxylin-eosin (HE) examination, the tumor was composed of monotonous tumor cells arranged in a storiform to whorled pattern focally with frequent mitotic figures. Areas of hyalinization were noted. Lymphocyte infiltration was present in multiple foci but the highest concentration of these cells was noted in the front of the tumor growth. Differential histochemical stainings (Reticulin stain; Van Gieson stain) revealed focal presence of collagen and abundant reticulin network. Immunohistochemistry was performed using antibodies for S-100 protein (Figure 6), Vimentin (Figure 7), Actin (Figure 8), Desmin, CD31, CD34 and Synaptophysin. Tumor cells were positive for S-100 protein, and Vimentin, there was focal positivity for Actin and there were no positive tumor cells stained for Desmin, Synaptophysin, CD31 and CD34. We reached a diagnosis of MPNST that originated from the tumor capsule.

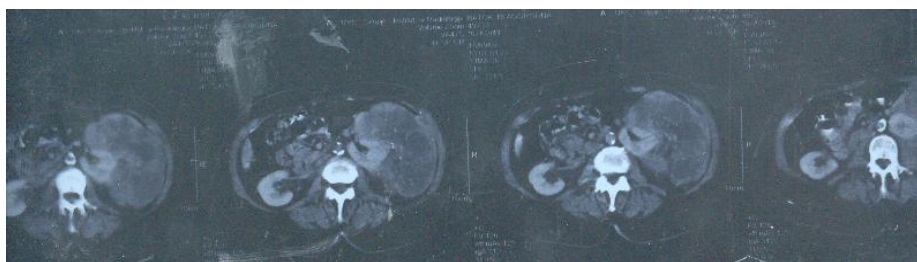


Figure 1 – CT scan of the abdomen revealed a large well-defined, lobulated and heterogeneous mass

Слика 1 – Компјутеризирана томографија на абдомен со приказ на голема, добро ограничена, лобулирана маса

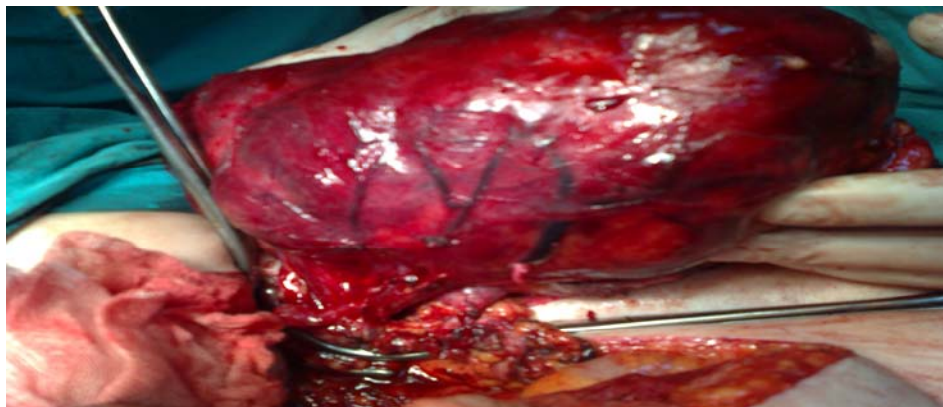


Figure 2 – Intraoperative view
Слика 2 – Интраоперативен изглед

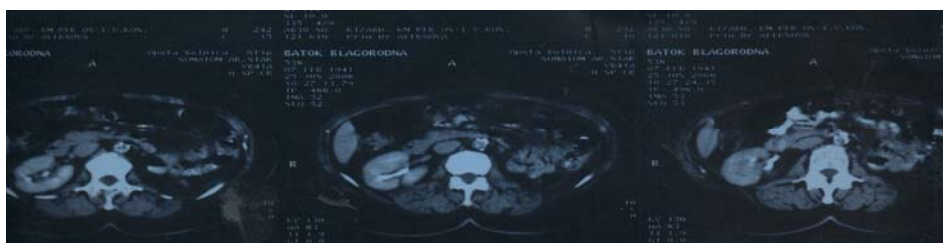


Figure 3 – CT scan control after 6 months did not reveal recurrence of the tumor
Слика 3 – Контролната компјутеризирана томографија по 6 месеци не покажува рецидив на туморот



Figure 4 – Gross specimen
Слика 4 – Макроскопски изглед

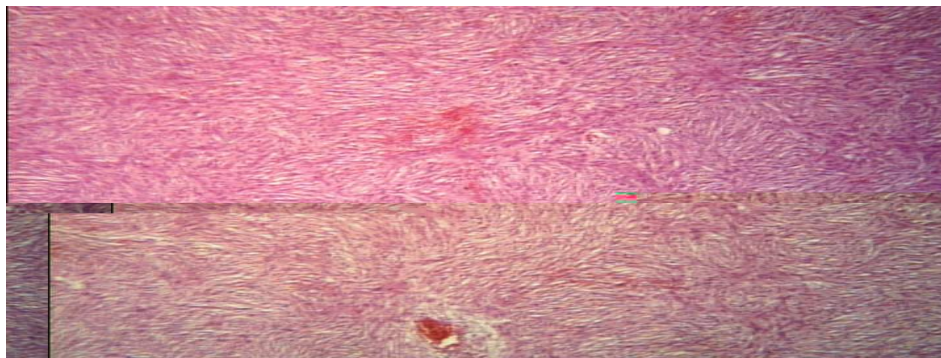


Figure 5 – Tumour was with sarcomatous (fish-flesh) appearance with some necrotic areas
Слика 5 – Туморот беше со саркома тоиден (како рибино месо) изглед со некои
некротични зони

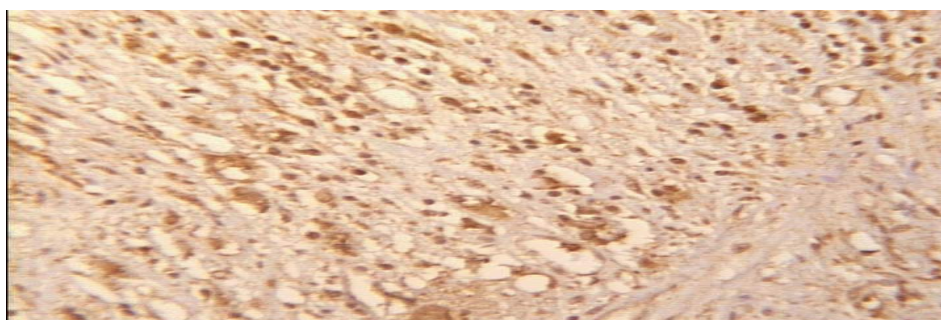


Figure 6 – S-100 protein $\times 200$, positive staining of tumour cells
Слика 6 – С-100 протеин $\times 200$, позитивно пребојување на туморски клетки

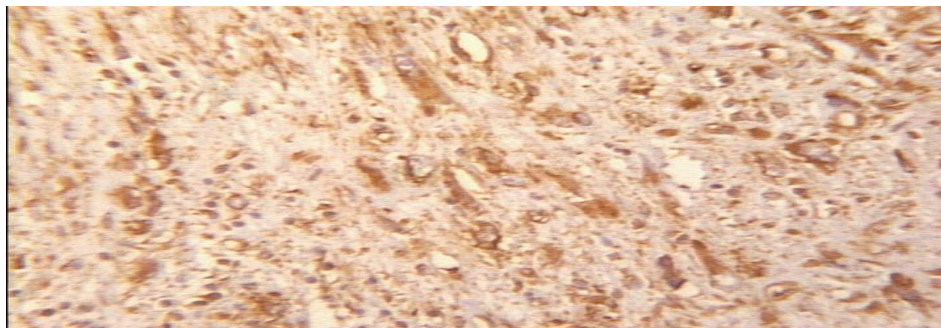


Figure 7 – Vimentin $\times 200$, positive staining of tumour cells
Слика 7 – Виментин $\times 200$, позитивно пребојување на туморски клетки

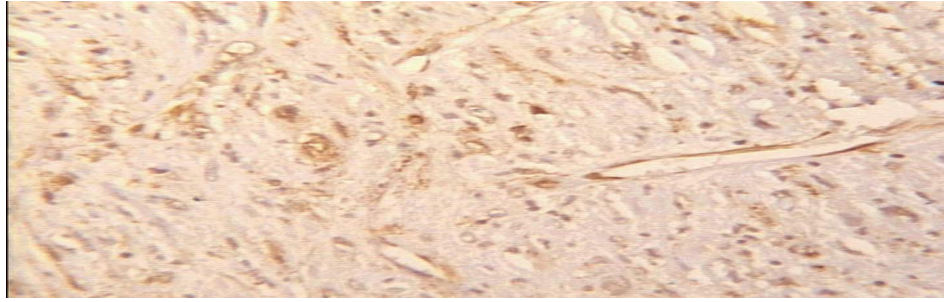


Figure 8 – Actin $\times 200$, positive staining of tumour cells

Слика 8 – Актин $\times 200$, позитивно пребојување на туморски клетки

Discussion

Malignant peripheral nerve sheath tumor is biologically an aggressive tumor for which the treatment of choice is surgery. A combination of gross, histopathological, immunohistochemical were all needed for diagnosis and grade of the MPNST [5]. Isolated neurogenic tumors, without any manifestation of neurofibromatosis, are reported rarely. Shahab *et al.* [6] described a case of neurofibroma of ileum without any manifestation of neurofibromatosis. Contrast enhanced CT and magnetic resonance imaging (MRI) are the most helpful imaging techniques to clearly identify tumor extent and to suggest its neurogenic origin. The presence of heterogeneity with evidence of necrosis and hemorrhage on MRI, and increased uptake on positron emission tomography scan, may prove helpful in detecting malignant change. Malignant peripheral nerve sheath tumors should be approached by a multi disciplinary team, assuring the complete surgical removal of the lesion. Disease-free and overall survival statistics reinforce the aggressive nature of this unique soft tissue tumor [7]. Histological diagnosis is required, but diagnosis can be compromised by the heterogeneous nature of the tumor. Surgical treatment should be undertaken whenever possible. This type of sarcoma is often very aggressive with frequent development of local recurrences and metastases [8]. Magnetic resonance imaging and CT scans are the first line procedures for localizing these tumors. Computer tomography scans can also be a guide for biopsies. A T1-weighted MR image reveals low-to-intermediate signal intensity, and T2-weighted MR images show high signal intensity in the lesion. Malignant peripheral nerve sheath tumors can arise within a previous radiation field. Malignant peripheral nerve sheath tumors tend to be larger (> 5 cm). They may exhibit ill-defined margins suggesting infiltration of adjacent tissues and associated edema. Heterogeneity with central necrosis on cross-sectional imaging is common in malignant lesions. Malignant

peripheral nerve sheath tumors generally involve the major nerve trunks and present with pain and neurologic symptoms, as well as a possible soft-tissue mass. Secondary MPNSTs can arise from prior radiation treatment, with a latency period of longer than 10 years [9]. The treatment for MPNST has been combined modality that includes aggressive surgery with a wide margin of clearance or mass. Several cycles of adjuvant chemotherapy with cyclophosphamide/doxorubicin have been advocated [4]. However, Wanebo *et al.* [2] in their study of 28 cases advocate radiotherapy as adjuvant.

Conclusions

We suggest including MPNST of the kidney capsule in the differential diagnosis of abdominal masses. We also suggest, in a case of malignancy or large tumor, that an open surgical procedure should be applied. According to the degree of malignancy, postoperative radiochemotherapy is indicated.

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Резиме

ИЗОЛИРАН MALIGNANT PERIPHERAL NERVE SHEATH TUMOR НА БУБРЕЖНАТА КАПСУЛА

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Појавата на изолиран Malignant Peripheral Nerve Sheath Tumor (MPNST) на бубрежната капсула е извонредно ретка, а неговото присуство се изразува со појава на неспецифични и неповрзани симптоми со преминација на болка во грбот. Компјутеризираната томографија, како imaging процедура на избор ја демонстрира локацијата на туморот и неговата поврзаност со околните структури. Екцизијата in toto на туморот се смета за процедура на избор која ќе ја покаже локацијата на туморот и неговиот сооднос со околните структури. Треба да се совладаат тешките компликации како што е крвавењето заради оштетување на околните структури. Воспоставувањето на хемостаза може да претставува сериозен проблем. Затоа, хируршката интервенција треба да се изведува со голема претпазливост и договор со пациентот. Ако со сигурност се исклучи малигнитетот, може да се размислува и за лапароскопска екцизија како алтернативен третман. Дефинирањето на нервот од кој потекнува туморот не секогаш се идентификува и затоа може да се очекува помал степен на невролошка дисфункција.

Прикажавме случај на Malignant Peripheral Nerve Sheath Tumor (MPNST) на бубрежната капсула без неурофиброматоза. Туморот беше лоциран во масната и фиброзната капсула. Истиот хируршки беше отстранет. Подоцна

пациентот беше подложен на хемотерапија. MPNST со оваа локализација е екстремно редок.

Клучни зборови: Malignant Peripheral Nerve Sheath Tumor (MPNST), компјутеризирана томографија, ретроперитонеум, хируршки компликации.

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