Original Article

Retroperitoneal tumors: a retrospective review of 14 patients

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ABSTRACT

Objective: In this study, we evaluated the pathological results and clinical presentation of patients with retroperitoneal tumors
Design: Retrospective study
Setting: Department of Urology at Hitit University, Çorum Erol Olçok Training and Research Hospital, and Department of Urology at Haydarpaşa Numune Training and Research Hospital
Subjects: Fourteen patients with retroperitoneal tumors
Intervention: Retroperitoneal mass and clinical suspicion of tumor
Main outcome measures: The symptoms at presentation and pathologic reports of the tumors were recorded in these patients.
Results: Of the patients evaluated for retroperitoneal tumor, eight patients were male. The mean age of the study group was 55.7 ± 11.4 years with a range of 41 to 75 years. Liposarcoma was the most common pathological diagnosis of the retroperitoneal tumors in these patients (35.7%). Schwannoma and gastrointestinal stromal tumor were reported in two patients. The other retroperitoneal masses were diagnosed as atypical lipomatous tumor, paraganglioma, myelolipoma, lipomatous hemanjiopericytoma and Castleman disease.
Conclusion: Retroperitoneal tumors are rare neoplasms in daily urological practice. With the development of radiological imaging techniques in recent years, most of the patients have been diagnosed incidentally at abdominal imagings for other purposes. The management of these patients is important because of the aggressive behavior of sarcomas.

KEY WORDS: liposarcoma, myelolipoma, neoplasm, paraganglioma

INTRODUCTION

The retroperitoneum is a complex potential area bounded anteriorly by the peritoneum, ipsilateral colon and mesocolon, liver, pancreas and stomach, and the posterior side is composed of the psoas, quadratus lumborum, transverse abdominal and iliacus muscles, diaphragm, ipsilateral kidney, ureter and gonadal vessels[1,2]. The retroperitoneum offers an enviroment for a wide spectrum of pathologies, including a variety of rare benign lesions and malignant neoplasms that can be either primary or metastatic. The malignant neoplasms occur four times more frequently than benign lesions[3]. Soft tissue sarcomas, 1% of all newly diagnosed malignancies, are mesenchymal tumors[4]. The retroperitoneum is the second most common site of origin of malignant mesenchymal tumours, after the lower extremites[5]. Sarcomas were reported to account for one-third of all retroperitoneal tumors. Other retroperitoneal tumors include primary lymphoproliferative tumors (Hodgkin's and non-Hodgkin's), epithelial tumors and metastatic disease from known or unknown primary sites[6]. Benign tumors of the retroperitoneum are schwannomas, neurofibromas, paragangliomas, fibromatosis, renal angiomyolipomas and lipomas[3]. We evaluated the patients who underwent retroperitoneal exploration secondary to the retroperitonel tumor retrospectively.

SUBJECTS AND METHODS

A total of twenty-two patients managed with retroperitoneal surgery between January 2011 and March 2015 were evaluated retrospectively. The patients were diagnosed by ultrasonography; computerized tomography (CT) and magnetic resonance imaging (MRI) were used for differential diagnosis. Figures 1 and 2 show the retroperitoneal tumoral mass preoperatively. The patients whose pathologic report of the surgical excision showed a

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non-tumoral mass were excluded from the study. The clinical symptoms, the age and gender of the patients, radiological and pathological findings of the 14 patients with retroperitoneal tumor were recorded. All surgical procedures were performed with open surgical techniques using subcostal Chevron incision. Total tumor excision was performed in all patients.

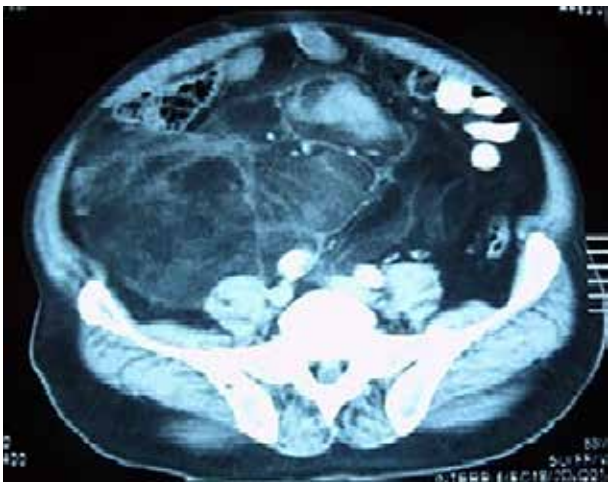

Fig 1: Computerized tomography image of the retroperitoneal mass (liposarcoma)

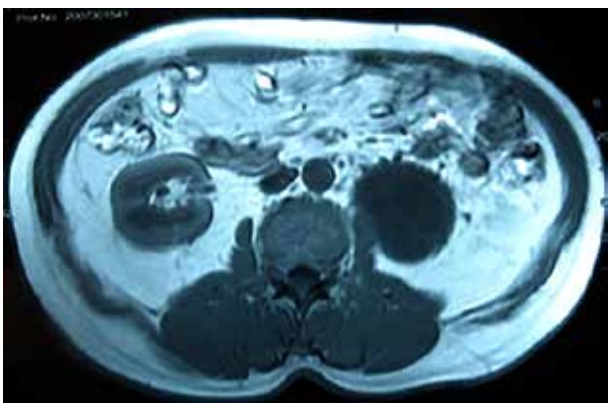

Fig 2: Magnetic resonance imaging showing the lesion (Schwannoma)

The data were expressed as mean ± standard deviation using statistical software program.

RESULTS

The mean age of the patients was 55.7 ± 11.4 years (range: 41 – 75 years). Of the patients, eight were male and the mean tumor size from the pathological gross specimen was 14.9 ± 9.9 cm with a range of 4 to 25 cm. Nine patients were diagnosed by ultrasonography. As the clinical presentation of the patients were evaluated, the history and physical examination of the patients revealed flank pain in five, abdominal mass in two, abdominal pain in three, hematuria and hot flushes in two patients, respectively. Two of the patients had non-specific symptoms.

Of the study group, liposarcoma was reported in five patients with four well differentiated and one undifferentiated histology. Lipomatous hemangiopericytoma, atypical lipomatous tumor, paraganglioma, myelolipoma and Castleman diseases (CD) were detected in one patient histologically. Schwannoma and gastrointestinal stromal tumor were diagnosed in two patients. Pathological results and clinical presentations are shown in Table 1. Immunohistochemical study was performed in seven patients. Smooth muscle actin (SMA), CD34,99,117, desmin, S-100, Ki67, Bcl-2, melanoma-specific melanoma marker (MART), inhibin, synaptophysin and chromogranin were used for immunohistochemical examination.

Table 1: Presentations and pathological results of the patients

Gender	Age	Symptom	Diagnosis
Male	75	Flank pain	Lipoma
Male	48	Nonspecific	Leiomyosarcoma
Male	58	Abdominal swelling	Hemangiopericytoma
Female	65	Hot flushes	Paraganglioma
Male	41	Abdominal pain	Liposarcoma
Female	44	Flank pain	GIST
Male	63	Abdominal swelling	Liposarcoma
Female	65	Nonspecific	Myelolipoma
Male	45	Flank pain	Schwannoma
Male	55	Abdominal pain	Schwannoma
Female	48	Flank pain	GIST
Female	55	Abdominal pain	Liposarcoma
Female	44	Hematuria	Castleman
Male	75	Flank pain	Liposarcoma

GIST: gastrointestinal stromal tumor

The mean follow up was 61.14 ± 6.93 months. During the follow up period, there was only one recurrence in a patient who was diagnosed as liposarcoma. A second surgical operation was done. One patient died three years after the operation because of myocardial infarction.

DISCUSSION

The retroperitoneum offers an environment to the tumors to originate and grow to a large size before the patients become symptomatic. Retroperitoneal tumors are usually diagnosed or identified with cross sectional imaging for other problems[4]. Most of the patients present with abdominal mass, increase in girth, palpable mass, early satiety and abdominal discomfort[7]. Patients may present with signs of bowel and/or ureteral obstruction, because of either compression or invasion of nearby structures[4]. Although the most common complaint is abdominal or back pain, weight loss and anemia are the other symptoms. Flank pain and abdominal complaints (71.42%) are the main symptoms of the patients in this study.

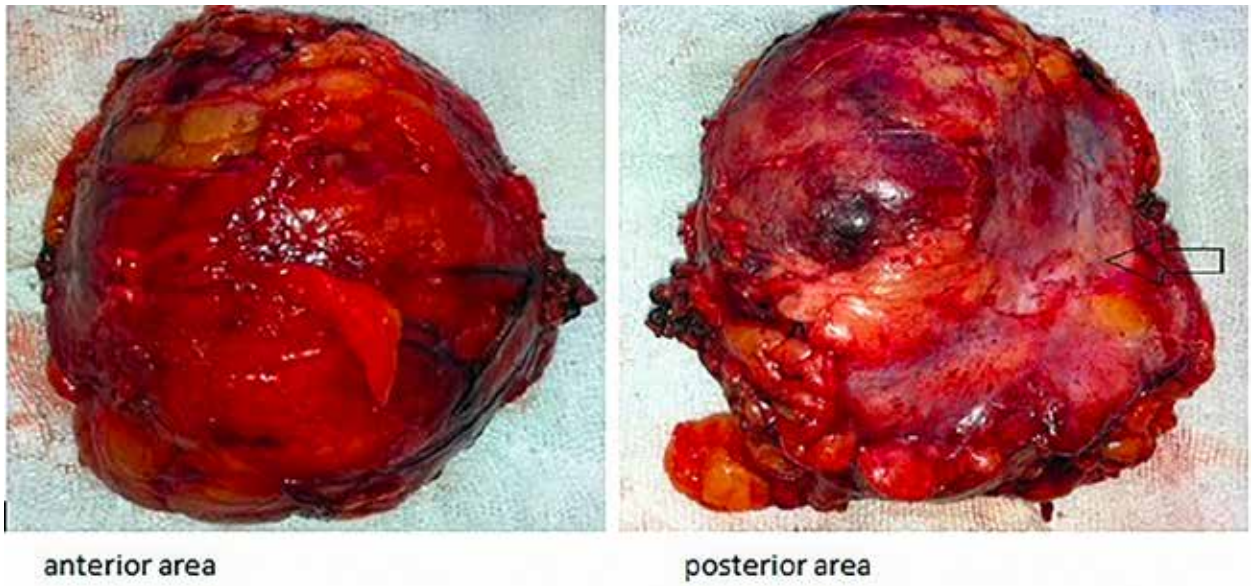

Fig 3: Complete resection of the retroperitoneal liposarcoma (Arrow shows the renal vascular area: posterior surface)

Sarcomas account for 33% of all retroperitoneal tumors, with two histological subtypes predominating, namely liposarcoma (70%) and leiomyosarcoma (15%)[6]. There is no predominance in terms of gender and race. The patients are usually diagnosed between 54 and 65 years of age[8]. These tumors can be found anywhere in the body, with 50% in the extremites, 10-15% in the trunk, less than 10% in the head and 15% in the neck retroperitoneum. Sarcoma was diagnosed in five patients, comprising 35.71% of the patients in the present study. Most of the patients were male (80%) and the mean age was 56.4 years. All patients had liposarcoma and the ratio of well differentiated to poorly differentiated was 4:1. Nephrectomy was performed in two patients because of local invasion of the sarcoma, the other patients were treated with tumor removal without nephrectomy (Fig 3).

Schwannoma is usually a benign tumor that originates from Schwann cells of the peripheral nerve sheath[9]. These tumors can occur in any neural tissue where Schwann cells are present[10]. Schwannomas are commonly found in cranial and peripheral nerves; the retroperitoneal schwannomas comprise 3% of all schwannomas[11]. Retroperitoneal schwannomas are approximately 1 - 5% of all retroperitoneal masses (Fig 2). The patients are usually asymptomatic. Although retroperitoneal schwannomas are almost benign, they may very rarely undergo malign transformation[12]. Schwannomas are composed of Schwann cells with regions of high and low cellularity termed Antoni A and Antoni B areas, with positive staining of S-100 microscopically[11]. Two patients had schwannoma in this study, with an incidence of 1.4%. The patients presented with flank pain and abdominal pain.

Gastrointestinal stromal tumor (GIST) is the most common mesenchymal tumor of the gastrointestinal tract and represents less than 1% of all malignancies[13]. GIST arises from the wall of gastrointestinal tract and is thought to originate from the cells called Interstitial Cells of Cajal and regulate the motility of the gastrointestinal tract. These tumors originate from the stomach, small intestine, colon and esophagus with a ratio of 40 - 70%, 20 - 40%, 5 - 15% and 5% respectively. These tumors may rarely occur outside the gastrointestinal tract and are called extra gastrointestinal stromal tumor[14]. CD 117 protein is the most specific and important marker, expressed in more than 95% of cases. GIST were reported in two patients in pathological evaluation. CD 117, Ki-67, S 100, CD 34, SMA and desmin were performed in these patients. CD 117 was positive (Fig 4a), S-100 and desmin were negative in two patients. CD 34 (Fig 4b) and SMA were positive in one patient. Ki-67 was less than 10% in the patients. Using the Fletcher classification scheme[15], one patient's tumor had high risk, and the other had intermediate risk for aggressive behaviour.

Myelolipoma is an uncommon benign tumor of the adrenal gland which consists of mature fat and mixed myeloid and erythroid cells[16]. Proportion of fat and myeloid tissue is the key factor for preoperative diagnosis by CT or MRI. Presence of calcifications, necrosis and intratumoral hemorrhage may confuse the diagnosis. Differential diagnosis include adrenal adenomas, retroperitoneal lipomatous masses such as liposarcoma, lipoma and angiomyolipoma. One patient was diagnosed with adrenal myelolipoma in the present study.

CD is an unusual benign lymphoid tumor[17]. It is classified into two groups with localized and multicentric subtypes. Three histological variants

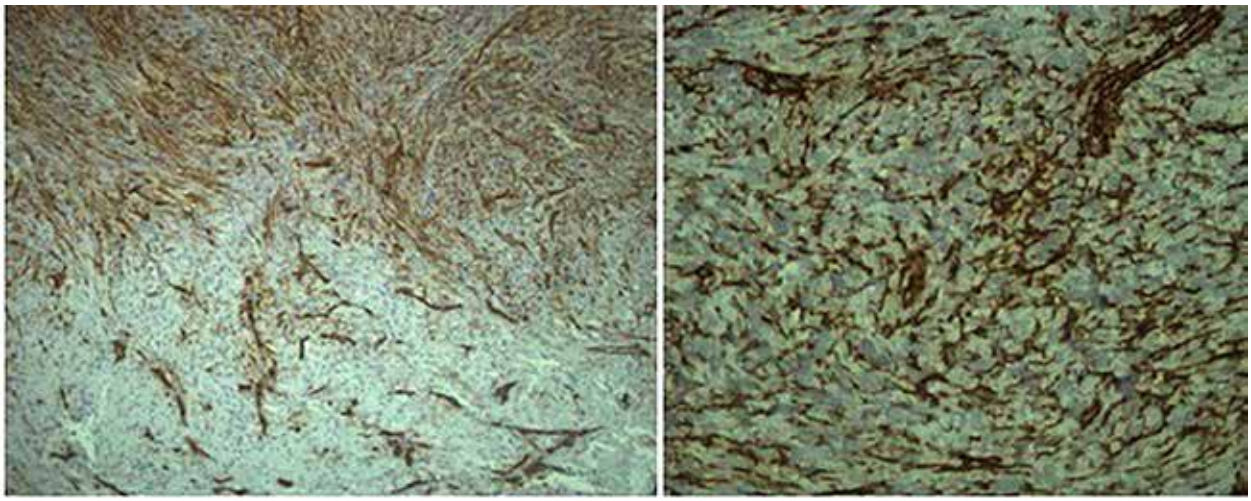

Fig 4: Immunohistochemical study of gastrointestinal stromal tumor. **a)** Positive staining of CD34 X40 magnification; **b)** Positive staining of CD 117 X 100 magnification

(hyaline-vascular, plasma cell and mixed types) have been described. Multicentric type of CD has a worse prognosis than localized type. CD may arise wherever lymphoid tissue is found. The most common site of origin is mediastinum; and the retroperitoneum is an uncommon localization which accounts for 7% of cases[18]. Complete surgical resection is an effective treatment and suggested for localized disease[17]. Chemotherapy and immunotherapy are effective for multicentric types of CD, if the diagnosis is confirmed histologically by lymphnode biopsy[19].

Paragangliomas are rare tumors which arise from the neural crest tissue that develops into sympathetic and parasympathetic paraganglia throughout the body[20]. Paraganglioma of adrenal medulla is known as pheochromocytoma, while paragangliomas located outside of the adrenal gland are called as extra adrenal paragangliomas. These tumors can be divided into functioning and non-functioning based on their ability to secrete hormones. Extra adrenal paragangliomas account for 10 - 15% of all paragangliomas and the age of the diagnosed is between 30 and 45 years[21]. The most common site for retroperitoneal paragangliomas is between the origin of inferior mesenteric artery and the aortic bifurcation that is known as organ of Zuckerkandl. Clinical presentations of retroperitoneal paragangliomas are based on location and ability to secrete hormones such as fluctuating or episodic hypertension, headache, and sweating. Nonfunctioning paragangliomas are usually asymptomatic and diagnosed incidentally as a mass. While CT scan also has a sensitivity of around 90% for identifying extra-adrenal paragangliomas, MRI is recommended for first imaging modality. Metaiodobenzylguanidine scintigraphy (MIBG scintigraphy) is usually used for the diagnosis of neuroendocrine tumors, but it lacks sensitivity for extra adrenal paragangliomas. PET scan has more sensitivity when compared to MIBG scintigraphy[22]. Our patient presented with sweating and hot flushes for three months. MIBG scintigraphy was used for diagnosis and detected positive findings for paraganglioma. Immunohistochemical study was performed with MART, inhibin, synaptophysin, chromogranin and Ki-67. Synaptophysin and chromogranin were positive (Fig 5); MART and inhibin were negative; and Ki-67 was less than 1%.

Hemangiopericytoma is a rare benign tumor derived from a type of smooth muscle cell attached to pericytoma, capillaries also known as Zimmerman pericytes and initially described by Stout and Murray in 1942[23]. The new edition of the World Health Organization Classification classified hemangiopericytoma into three groups; solitary fibrous tumor, including giant cell angiofibroma and lipomatous hemangiopericytoma[24]. Lipomatous hemangiopericytoma is a rare benign mesenchymal neoplasm that grows slowly and consists of mature adipocytes and hemangiopericytoma like areas[25]. The common sites of this tumor are the deep soft tissues of the lower extremity and the retroperitoneum. Nielsen described this unique variant of hemangiopericytoma in 1995[26]. Positive staining of CD 34 and 99 can be helpful in immunohistochemical study for the diagnosis[25]. The tumor size of the patients was 16 cm and immunohistochemical study revealed positive staining for CD 31, 34, 99, Bcl-2, SMA, desmin, S100 and negative staining for CD 117 in pathological evaluation.

The retrospective design of the study and small number of patients are the main limitations of this study. The patients were treated and diagnosed by different pathologists and surgeons respectively.

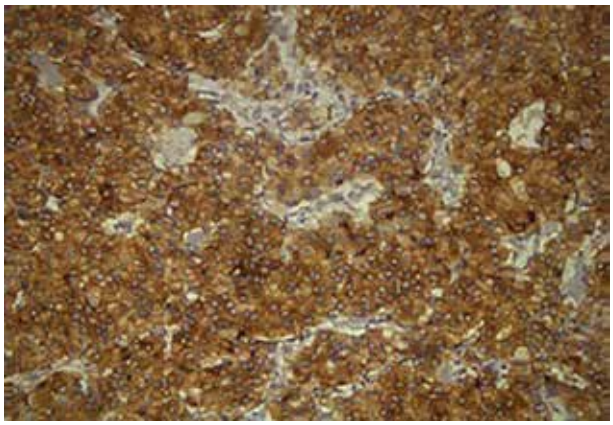

Fig 5: Histological examination of paraganglioma, synaptophysin X100 magnification

CONCLUSION

The retroperitoneal tumor is a rare condition in urology practice. With widespread use of ultrasonography and other imaging techniques, retroperitoneal tumor incidence is increasing in urology practice. Preoperative evaluation is very important because of limited diagnostic accuracy.

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