Case Report

Extra adrenal retroperitoneal paraganglioma

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ABSTRACT

We herein report a case of a 45-year-old Saudi lady not diabetic nor hypertensive who presented to the emergency room with a one day history of severe central and lower abdominal pain. On examination, she was hemodynamically stable and abdominal examination showed tenderness in the lower abdomen. Her hematological and biochemical investigations were normal. Computed tomography of the abdomen showed an 8 x 7 cm retroperitoneal mass located at the aortic bifurcation. The patient had exploratory laparotomy and complete excision of the mass. The histopathological study showed a paraganglioma. The patient had an uneventful postoperative period and follow up.

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Paragangliomas are tumors of specialized cells (chief cells of the paraganglia) derived from the neural crest. These cells are symmetrically distributed along the aorta in close association with the sympathetic chain. When this tissue aggregates in the adrenal medulla it gives rise to an intra adrenal paraganglioma, known as a pheochromocytoma. When it remains in para aortic sites it may develop into an extra adrenal retroperitoneal paraganglioma. The diagnosis of a paraganglioma is infrequently made preoperatively unless the tumor is functional. Most patients with nonfunctional tumors present with abdominal pain and a mass and occasionally have distal metastasis.[1] We present here a case of a Saudi lady who presented with severe acute abdominal pain and was found at exploratory laparotomy to have a retroperitoneal mass at the aortic bifurcation. Complete excision was performed, and the histopathological study showed a paraganglioma.

Case Report. A 45-year-old, otherwise, healthy Saudi lady, presented to the emergency room with severe central and lower abdominal pain. There was no associated vomiting and no change of bowel habits. There were no previous similar attacks. The patient was not known to be hypertensive and not on any medication. On physical examination, the patient looked ill and in pain but was hemodynamically stable. Abdominal examination showed tenderness in the hypogastric region, there was no evidence of peritonitis, and no palpable mass. Bowel sounds were normal and rectal examination was normal. Her hematological and biochemical profile was normal. Chest and abdomen radiographs revealed no abnormalities. The patient was admitted as a case of acute abdomen. Computed tomography (CT) scan of the abdomen showed the presence of a well defined 8 x 7 cm retroperitoneal cystic mass located at the aortic bifurcation with peripheral rim enhancement and central low attenuation. No calcification or lymphadenopathies were detected. The differential diagnosis according to the CT report was: a hematoma, an abscess or a necrotic lymph node **(Figure 1).** According to the clinical presentation and, the CT findings a paraganglioma was not suspected and therefore, serum and urine catecholamine levels were not performed. Further, investigations including fine needle aspiration biopsy and aortic angiography were

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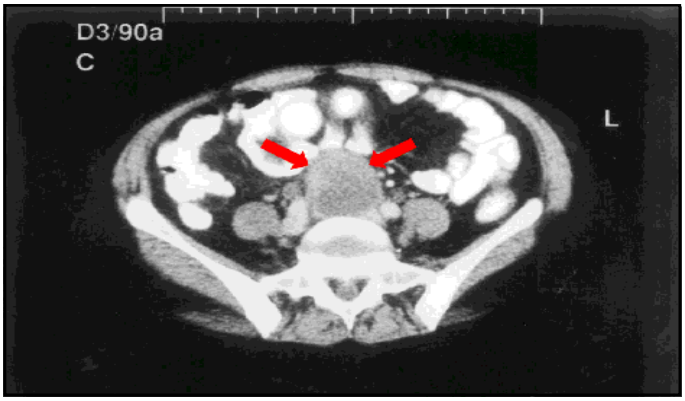

Figure 1 - Computed tomography of the abdomen showing an 8 x 7 cm retroperitoneal mass located at the aortic bifurcation.

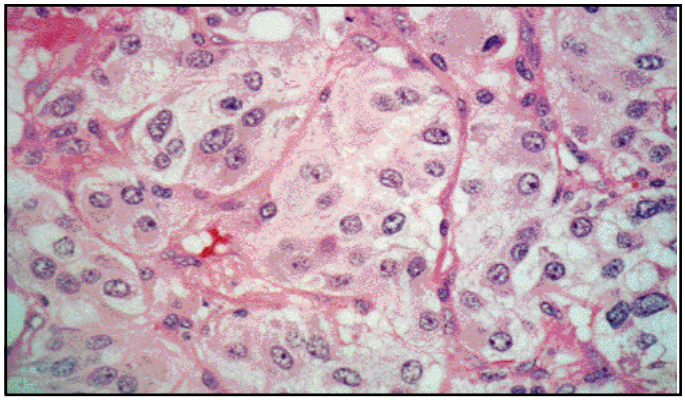

Figure 2 - Paraganglioma: photomicrograph showing packets of neoplastic cells with vesicular nuclei and abundant cytoplasm, blood vessels are also seen between the described group of cells. Hematoxylin and eosin stain x 200.

planned, however, the patient continued to have severe abdominal pain that was not relieved by high doses of analgesic (pethidine). For that reason, she was taken to the operating room the next day where an exploration was carried out through a lower midline incision. An 8 x 7 cm firm, oval retroperitoneal mass was found, it was brownish in color and encapsulated. It was located at the aortic bifurcation superior to the left iliac vein. The mass was dissected carefully; the abdominal aorta and both common Iliac arteries were controlled proximally and distally to control any massive bleeding during dissection. The mass was excised completely. The patient had a smooth postoperative recovery and was discharged in good general condition. She was followed in the clinic for 12 months and was normotensive and free of symptoms. The histopathological study showed a paraganglioma **(Figure 2)**.

A paraganglioma is an unusual tumor arising from chemoreceptor cells derived from the neural crest. Although these tumors generally occur in the head and neck, where the term "carotid body tumor" applies,[2] Olson and Abell[3] reported 21 cases of retroperitoneal paraganglioma in 1969. The adrenal medulla bilaterally presents the largest collection of this paraganglionic tissue from which pheochromocytoma arises much more frequently than its extra adrenal counterpart.[4] Only 10% of retroperitoneal paraganglioma occur outside the adrenal gland,[5] most commonly adjacent to the aorta and vertebral column and in particular the area corresponding to the organs of Zuckerkandl. Zuckerkandl in 1901 was the first who described the distribution of paraganglia in the human fetus. The organ of Zuckerkandl referred to as "aortic bodies" is represented by multiple dispersed paraganglia between the origin of the inferior mesenteric artery and the aortic bifurcation. This paraganglia are histologically similar to carotid body and adrenal medullary tissue.[4] This tissue regresses in children after the age of 12-18 months. Lack et al[4] described the macroscopic and microscopic appearances of 12 retroperitoneal paragangliomas. They found the majority of tumors to be partially encapsulated and some showed areas of necrosis and hemorrhage. The functionally active tumors were small in size. The microscopic patterns ranged between zellaballen pattern (the characteristic pattern of carotid body and related head and neck paraganglioma) and the broad anastomosing or trabeculor pattern. Frozen section diagnosis is often difficult.[13] The cytologic diagnosis of paraganglioma is difficult as these tumors exhibit a plethora of features that overlap with other neoplasms.[14] Retroperitoneal paragangliomas can be detected early if clinical findings caused by excess secretion of Catecholamines are present,[6] which occurs in 60% of patients.[4] Urinary catecholamines are elevated, usually with a predominance of norepinephrine.[7,8] The diagnosis of nonfunctioning tumors is difficult and infrequently made preoperatively. Most of these patients have abdominal pain and palpable masses and occasionally symptoms of distal metastases.[1,19] Abdominal pain and discomfort was the most frequent complaint.[4] There appears to be slight predilection for males, which contrasts with the marked female predominance in cases of head and neck paragangliomas.[4] Paragangliomas are generally considered more likely to be malignant than pheochromocytomas with a malignancy rate of 24% to 50%[1,9,15] and 10%.[10] A CT scan is the first modality used to localize functional tumors.[1,16] Metaiodobenzylguanidine (MIBG) may help to delineate multiple tumors and small tumors not seen on CT scan.[11] Leslie et al[12] compared the magnetic resonance imaging (MRI) with CT and I^{-131} MIBG scintigraphy and he found that MRI was preferable than CT in the evaluation of primary pheochromocytoma particularly in patients with hypertension and borderline catecholamine levels. In the patients with recurrent or metastatic disease, the data suggests the I^{131} MIBG scintigraphy is the examination of choice. Arteriography provides the most valuable preoperative information. The highly vascular

nature of these tumors is usually apparent, and major vessel involvement may predict the need for a major vascular procedure.[13] Wendelin et al[6] studied 28 cases with retroperitoneal paraganglioma and found that benign and malignant lesions could not be distinguished on the basis of histopathological criteria only. Distant metastasis and local invasion of adjacent organs were the only reliable indicators of malignancy. The malignancy rate in their study was 14%. The average diameter of all tumors was 8.6 cm. The malignant paraganglioma are larger in size (an average diameter of 13.8 cm) and have irregular margins, and in 3 of the 4 tumors, low density areas of necrosis. Sclafani et al,[1] reviewed 22 cases between 1949 and 1990 at Memorial Sloan-Kettering Cancer Center. Complete surgical resection could be carried out in only 13 patients and those patients had better overall survival compared to patients whose tumors were not completely resected. They had a malignancy rate of 50% and patients developed metastases to bone, liver, peritoneum, pelvis, ovaries, cervical lymph nodes and lung. They considered the presence of distant metastases as a criteria for malignancy. The 5-year-old and 10 year disease free survival rates were 75% and 45%. Once diagnosed with metastatic disease 50% of patients will die of their disease within 3 years. Adjuvant chemotherapy and radiotherapy may be used for palliation of symptoms.[17,18]

In conclusion, retroperitoneal paragangliomas should be kept in mind when a Surgeon is confronted with a retroperitoneal mass. Other tumors of neurogenic origin may resemble paragangliomas in both distribution and appearance; these include neurofibromas and neurilemomas. In addition, primary retroperitoneal tumors like poorly differentiated liposarcomas and leiomyosarcomas[6] must be included in the differential diagnosis. Surgical resection offers the only chance of cure in this disease.[1,16] Lifelong follow up is essential as metastasis may occur late.[9]

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