Case study

Retroperitoneal Non-functioning Paraganglioma: A Case Report with Review of Literature

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ABSTRACT

Background : Paragangliomas (PGs) are rare neoplasms arising from cells of the primitive neural crest. These tumours are infrequently diagnosed and often difficult to diagnose and treat.

. The radiological diagnosis is based on computed tomography (CT) and magnetic resonance imaging (MRI). We report a case of a 41 years old male presenting with abdominal pain who had a retroperitoneal tumour situated at the left anterior para renal space. Complete resection of the tumour was performed. The histological examination and immunohistochemical analyses concluded the diagnosis of paraganglioma.

Case report : We aim to describe a case report of an incidental finding of left retroperitoneal paraganglioma in a young man who presented with left sided abdominal pain. We also aim to emphasize the importance of diagnosis and the malignant potential of these tumours. Conclusion : Retroperitoneal paragangliomas are most commonly benign with good prognosis, but can be locally invasive and metastasize as well. The possibility for malignant transformation of paragangliomas makes surgical excision the treatment of choice.

Keywords: Retroperitoneal tumours, paraganglioma, neuroendocrine tumor, immunohistochemistry.

1. INTRODUCTION

Paragangliomas are relatively rare endocrine tumours that arise from paraganglionic tissue, a widely dispersed collection of specialized neural crest cells [1]. Paragangliomas, also known as extra-adrenal pheochromocytomas, account for 5-10% of pheochromocytomas and can occur in any position between the neck and the base of the pelvis [2].The retroperitoneal paraganglioma accounts for over 50% of all paragangliomas [3].

within the para-aortic and perinephric spaces [4]. It can occur at any age, most commonly in young adults and tend to be sporadic. However, they can be linked to syndromes in approximately 30% [5].

Different imaging modalities play a crucial role in detecting these tumors.

The typical CT appearance of paraganglioma is that of a well limited isodense mass with an intense contrast enhancement. On MRI, the mass presents an iso signal or hyposignal in T1 and hypersignal in T2 with an early and intense enhancement..Consequently, these tumors may be mistaken for other primary epithelial or mesenchymal abdominal tumors. Since benign and malignant retroperitoneal PG have both the same histological appearance, the best predictor of malignancy is metastasis or recurrence.

The possibility of malignant transformation of paragangliomas makes surgical excision the treatment of choice.

The present case is reported to describe the clinical presentation, radiological features, and histological features of a nonfunctioning retroperitoneal paraganglioma discovered during investigation of abdominal pain.

2. Case report:

A 41 year old male was referred with vague abdominal pain of 4 month duration. On further inquiry, there was no history of hypertension, chest tightness, palpitations, anxiety or weakness. He had undergone a ventral hernia repair a few years ago. He was taking no regular medications. There was no family history of endocrine disorders. His vitals remained stable during the hospital stay.

The physical examination revealed a palpable and tender mass of the upper left abdominal quadrant, the pain was sudden in onset and progressive in nature. The rest of the physical examination was otherwise normal.

Initial investigations including biological assessments and X-ray were all normal. However, Abdominal computed tomography (CT) was performed and revealed a well-defined retroperitoneal mass lateralized on the left, round shaped, with regular contours, spontaneously hypodense and heterogeneous, intensely and heterogeneously enhanced after injection of PDC in the arterial phase with a close connection to the anterior para renal space and the psoas muscle, measuring 82*71*100 mm, without locoregional invasion, suggestive of liposarcoma or neuroendocrine tumors (Figure. 1). On MRI, the mass presents an iso signal or hyposignal in T1 and hypersignal in T2 with an early and intense enhancement. A diagnosis of GIST was made (Figure. 2). Consequently, these tumors may be mistaken for other primary epithelial or mesenchymal abdominal tumors.

Through a midline laparotomy incision, after mobilization of the left colon we found a very vascularized retroperitoneal tumour of about diameter 10 cm that was encapsulated and soft, with a close proximity to the left ureter and the psoas muscle. The lesion was completely resected after a laborious ureterolysis (Figure. 3). The lateral boundary of the dissection was at the left ureter (Figure. 4).

There were no intraoperative or postoperative complications and He was discharged home on the 6th postoperative day.

On macroscopic examination the mass was encapsulated with a lobulated and central areas of hemorrhage. Histological examination of the surgical specimen coupled with immunohistochemical study was consistent with the diagnosis of paraganglioma.

Figure 1. Abdominal CT scan with PDC injection : well-defined retroperitoneal mass (white arrow)

Figure 2. MRI image show the paraganglioma (yellow arrow), with a close connection to the anterior para renal space (K : the kidney)

Figure 3. The excised paraganglioma

Figure 4. Operative photo : note the paraganglioma (P) and close proximity of the ureter (yellow arrow) and the psoas muscle (white arrow)

3. discussion

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Those arising from the adrenal medulla are named phaeochromocytoma and comprise approximately 80% of these tumours. Extra-adrenal paragangliomas (10%–20%) are found arising from chromaffin tissue along the autonomic nervous system and can thus be found in the head, neck, thorax, abdomen and pelvis.

the abdomen, usually located in the organ of Zuckerkandl at the aortic bifurcation, consistent with the distribution of paraganglia first characterized in the human fetus by Zuckerkandl in 1901 [6].

Occasionally, these tumors are multiple, or they may be associated with paragangliomas of other sites or with other tumors such as gastrointestinal stromal tumors of the stomach and pulmonary chondromas as a component of Carney's triad. Patient age at diagnosis is usually about 40 years, and the incidence is similar between men and women. A large proportion of paragangliomas are related to disease-causing mutations or hereditary syndromes such as Von Hippel–Lindau (VHL) gene mutations [7].

Patients with functional paragangliomas that produce catecholamines may experience hypertension, flushing, tachycardia, palpitations, and/or anxiety [8] and they are easy to diagnose because of elevated urinary catecholamine. Those with non-functioning retroperitoneal paragangliomas are characterized by their asymptomatic profile and normal levels of catecholamines in the urine and blood. And may be diagnosed incidentally or present with compressive symptoms such as abdominal pain as was the case in our patient, or may be associated with nausea, vomiting, abdominal distension and weight loss [9].

Diagnosis of retroperitoneal paraganglioma is infrequently made unless the tumour is functional. Owing to their location, non secreting character and slow growth, they are clinically latent tumors.

Ultrasound may be used as a first-line investigation however CT and MRI have higher sensitivity. Computed tomography scans typically show a solid round or oval mass that is heterogeneous but that may show central necrosis or hemorrhage, calcification and enhancement. Ultimately although imaging techniques are helpful, the diagnosis of paragangliomas can only be confirmed with careful histological and immunohistochemical evaluation [10] as is the scenario in the present case. Histologically paragangliomas are diagnosed by their highly vascular appearance, with chief cells and sustentacular cells arranged in clusters called zell-ballen. It is this rich vascular component of the tumor that explains the intense contrast enhancement at CT or MRI imaging. Specific antibodies for neuroendocrine markers such as synaptophysin and chromogranin, as well as S-100 protein, may also be used to confirm the diagnosis [11].

Due to the malignant potential of paragangliomas, Surgical resection offers the only chance of cure. Resection is often challenging as these highly vascular tumours are frequently located near multiple vital blood vessels. If a tumour is felt to be unresectable, attempts to reduce its size using chemotherapy, radiation therapy or embolisation may be indicated because resection offers the only chance of cure [12]. These complementary therapies can ensure a positive response in 50% of the cases but without any significant influence on prognosis [13]. Surgery is the only means that allows a significant improvement in prognosis with a five-year and ten year survival rate without relapse of 75% and 45% respectively.

It is reported that extra-adrenal tumours are more likely to be malignant than adrenal tumours [14]. Distant sites for metastasis include bone, liver, peritoneum, pelvis, cervical lymph nodes, ovaries and lungs. As paragangliomas are rare tumours, there is lack of large prospective trials and all available series have limited power to detect significant differences regarding survival in patients with or without metastasis at initial presentation. In a series of 22 cases of extra-adrenal retroperitoneal paraganglioma from Memorial Sloan-Kettering Cancer Center, the 5-year survival for tumours not resected was 19% compared with 75% after complete resection [15]. This is consistent with the 5 years survival rate of 73%, reported by Cunningham et al. [16]. Metastasis could be seen up to 7 years after resection. Once metastasis occurs, survival is usually less than 3 years [15]. No difference in the survival period was reported between functional and nonfunctional paragangliomas.

Postoperative monitoring with CT/MRI and PET scans is recommended in order to detect malignant potential or progression of the disease. This should begin three months after surgery and continue biannually for the first three years. The total duration of follow- up recommended is at least ten years for PGs and if there is a high risk of malignant PG, the followup must be extended [17].

In conclusion, our case emphasizes the necessity to include extra-adrenal paraganglioma in the differential diagnosis and management of retroperitoneal tumours, despite its rarity. Imaging studies have greatly contributed to a more accurate diagnosis of paraganglioma. Yet the gold standard of diagnosis remains a pathological one. Lifelong follow-up of

patients with retroperitoneal paragangliomas is essential as metastasis and recurrence may occur.

4. Conclusion

Recurrence and metastasis define the malignant forms. Patients should be initially evaluated with catecholamine levels, followed by CT or MRI to locate the primary lesion. The final diagnosis is based on immunohistochemical findings. The surgical resection remains the treatment of choice for the localized tumors. Advances in genetic testing and discovery of new molecular markers are contributing to increased understanding of paragangliomas, however at present there is no way to definitively predict metastatic risk.

Regular follow-up with ultrasound and CT scan are necessary to detect early tumor recurrence.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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