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Retroperitoneal Ganglioneuroma Presenting as a Right Renal Mass in an Adult Patient: A Case Report and Literature Review

Sataa Sallami,[1] Mohamed Hmidi,[1] Sami Ben Rhouma,[1] Mohamed Chelif,[1] Adel Dahmani,[1] Ines Chelly,[2] Nidhameddine Kchir,[2] Ali Horchani,[1] Yassine Nouira[1]

[1]Department of Urology and [2]Department of Pathology, La Rabta Hospital-University, Tunis, Tunisia

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ABSTRACT

Ganglioneuromas are benign tumors arising from the neural crest. The reported case is a 47-year-old female who presented with right lumbar pain that increased progressively in intensity over the last 3 years. Computed tomography revealed a homogeneous mass located just above and behind the right kidney. The patient underwent a tumorectomy through a lumbar approach. Histology revealed a primary extra-adrenal retroperitoneal ganglioneuroma. Related literature is reviewed.

INTRODUCTION

Ganglioneuroma (GN) is a rare, primitive, benign neurogenic tumor [1]. It arises from the primordial neural crest cells that form the sympathetic nervous system [2]. The most common sites of involvement are the posterior mediastinum and retroperitoneum [3].

Most GNs are asymptomatic and found incidentally [1]. They affect newborns and infants more often than adolescents and adults [4]. They are relatively difficult to distinguish from other tumors because of the lack of specific imaging findings [1]. The patient in the present report has a primary extra-adrenal retroperitoneal GN.

CASE REPORT

A 47-year-old woman presented with the chief complaint of right upper quadrant abdominal pain that gradually increased in intensity over the last 3 years. Physical examination was unremarkable.

Ultrasonography revealed a 7 cm heterogeneous, hypoechoic mass with well-defined borders. It was located in the right lumbar region behind the upper pole and independent of the right kidney (Figure 1). Hormonal studies (24-hour urinary catecholamine) were insignificant. Computed tomography (CT) revealed a 8.6 cm x 6.6 cm x 7.9 cm solid tumor above the upper pole of the right kidney, with evidence of neovascularity within the tumor (Figure 2). No involvement of the renal vein or inferior vena cava was present. The adrenal gland was normal and no lymph nodes were enlarged.

Because the CT scan results were ambiguous, MRI was performed. The results revealed high signal intensity on T2 (Figure 3). Many diagnoses were suspected, including lymphoma, hematoma, or possibly a hydatid cyst.

Surgical excision was performed through a lumbar incision. Intraoperatively, no abnormality was observed in the right kidney. The excised tumor was well-demarcated behind the upper pole of the right kidney (Figure 4). The adrenal gland was macroscopically normal. The tumor specimen was noncystic, weighing 24 g and measuring 8 cm x 4 cm x 2 cm. It was a large, encapsulated mass of firm consistency with a

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CORRESPONDENCE: Dr. Satâa Sallami, Department of Urology, La Rabta Hospital-University, Tunis 1007, Tunisia (sataa_sallami@yahoo.fr).

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Abbreviations and Acronyms

CT = computed tomography
GN = ganglioneuroma
MRI = magnetic resonance imaging

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case report

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Figure 1. Ultrasonography Showing a Heterogeneous, Hypoechoic Mass With Well-Defined Borders.

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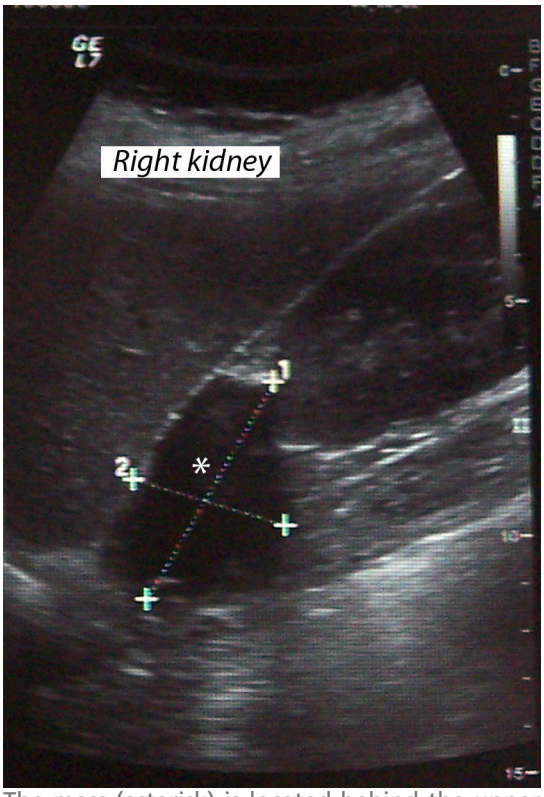

The mass (asterisk) is located behind the upper pole and independent of the right kidney.

Figure 2. CT Scan Showing a Homogeneous Mass Arising in the Retroperitoneum (circle).

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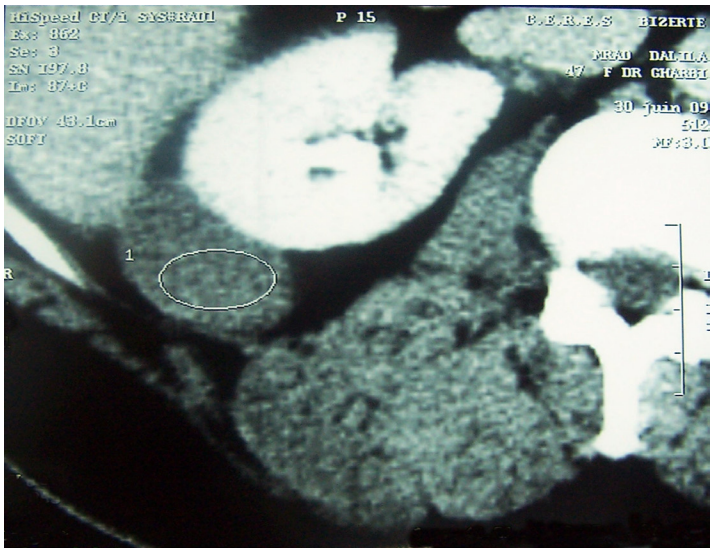

Figure 3. T2-Weighted Magnetic Resonance Image of the Mass.

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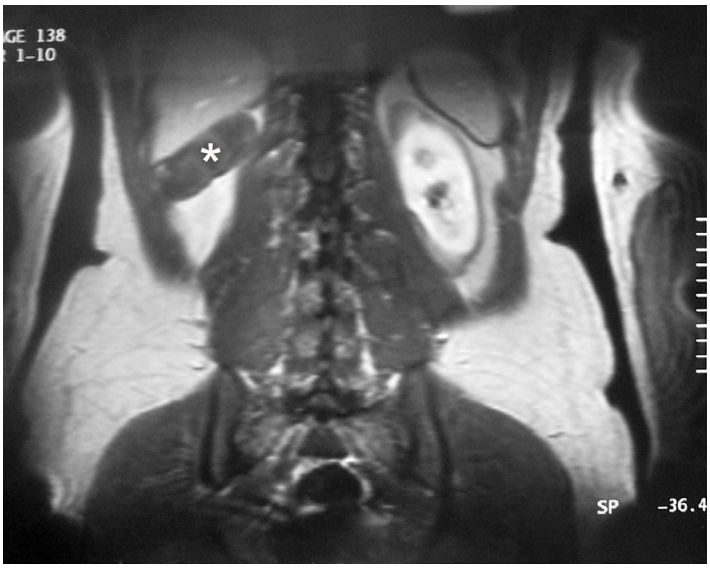

The mass (asterisk) is heterogeneous, with high signal intensity (ie, greater than that of the liver).

Figure 4. Intraoperative View of the Tumor (asterisk)..

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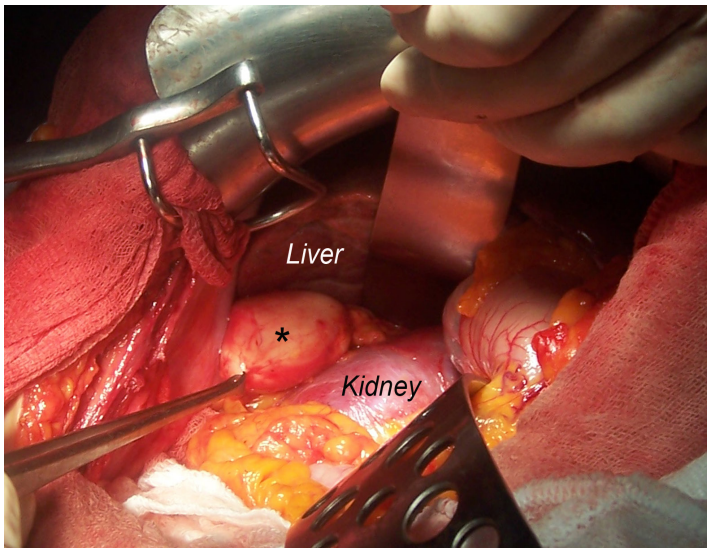

Figure 5. Postoperative Photographs of the Specimen.
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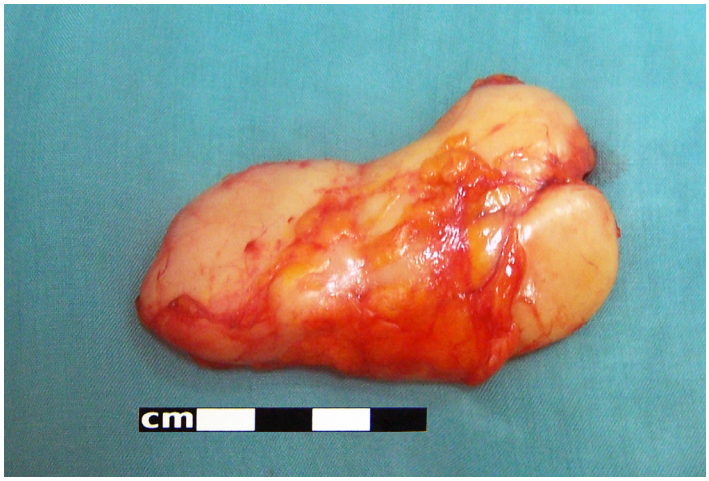
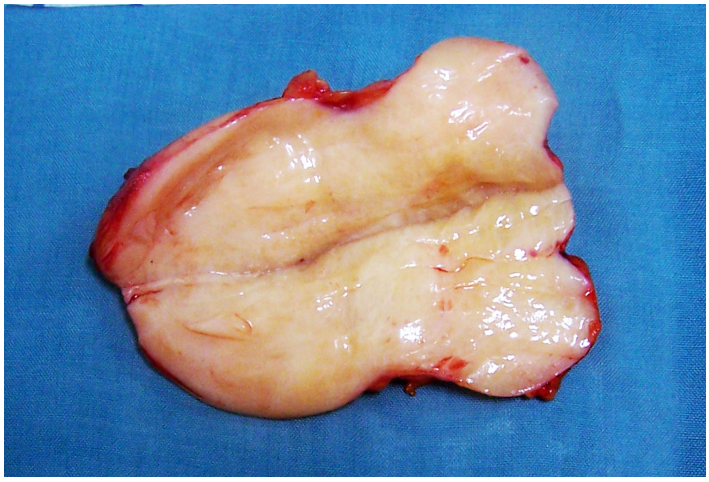
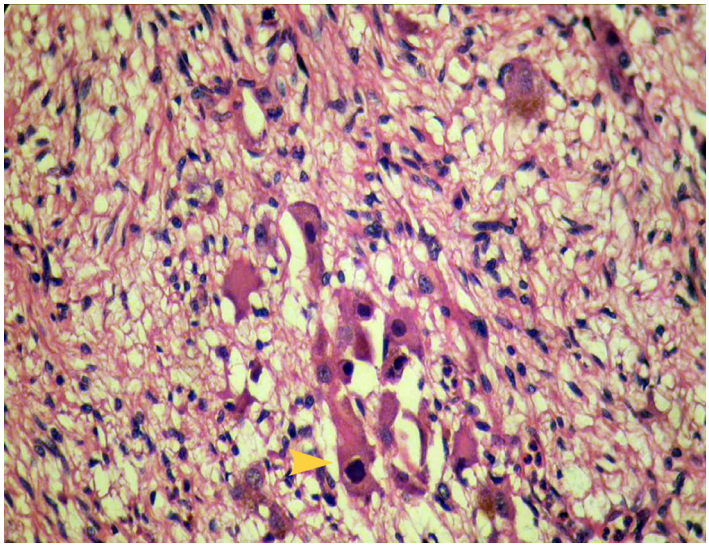




homogeneous, solid, grayish-white cut surface and a focally edematous appearance (Figure 5).

The postoperative histologic examination revealed a mixture of immature and mature ganglion fibers and neuronal cells that were suggestive of retroperitoneal GN, without any signs of malignancy. Pathological findings showed spindle cells with abundant cytoplasm, big nuclei, and prominent nucleoli without atypia (Figure 6).

Figure 6. Pathological Evaluation (Hematoxylin and Eosin, 100x). doi: 10.3834/uij.1944-5784.2010.10.03f6



Mixture of immature and mature ganglion fibers and neuronal cells, suggestive of retroperitoneal ganglioneuroma. Spindle cells with abundant cytoplasm, big nuclei and prominent nucleoli (arrow) without atypia.

The postoperative period was uneventful, and the patient was discharged on the 3rd postoperative day. The patient was relieved of her complaints after surgery, and no evidence of recurrence was found at the 9-month follow-up evaluation.

DISCUSSION

GN is a rare, benign tumor that is usually asymptomatic [1,5]. Although GNs usually appear in children, *retroperitoneal* GNs appear more often in adults [2]. The older mean age of appearance for retroperitoneal GNs is probably explained by the localization of the tumor, its tendency to produce few symptoms, and its slow growth [2]. It has no sex predilection, having a male:female ratio of 1:1 [1].

GNs are mostly sporadic, but there are a few reports of GNs associated with neurofibromatosis type II and multiple endocrinologic neoplasia type II [6]. The most common site of occurrence is the posterior mediastinum and the retroperitoneum [2,3]. Other sites of occurrence include the gastrointestinal tract [7], parapharyngeal area [8], bone [9], supraclavicular area [10], and pelvis [11]. Retroperitoneal GNs are usually nonfunctioning and asymptomatic until they reach large sizes, when they cause symptoms due to local expansion and compression of adjacent structures [12].

Diagnosis of Ganglioneuroma

GN is difficult to distinguish from other tumors due to lack of specific image findings [13]. Ultrasonography reveals a homogeneous, hypoechoic mass with well-defined borders

[5]. CT is the most commonly used imaging modality for assessment because it reveals the extent of the tumor, organ of origin, regional invasion, vascular encasement, adenopathy, and calcification [11].

MRI typically shows that GNs are homogeneous with relatively low signal intensity on T1-weighted images [14]. Several reports indicate that relatively high, heterogeneous signal intensity on T2-weighted images correlates with GN; the appearance is presumed to be caused by a combination of myxoid material and relatively few ganglion cells. The MRI may also show curvilinear bands of low signal intensity on T2-weighted images, which give the tumor a whorled appearance [14].

MRI enhancement varies from mild to marked. Typically, GNs demonstrate gradually increasing enhancement rather than early enhancement during contrast-enhanced dynamic MRI, which is similar to the results from contrast-enhanced CT [15]. With either imaging method, retroperitoneal GNs appear as well-circumscribed oval, crescentic, or lobulated masses [14]. They usually reveal some degree of hemorrhage, necrosis, and calcification [1]. A well-circumscribed mass with a tendency to partially or completely surround blood vessels without compressing the lumen was the mainstay of these findings [16].

Increasing numbers of GNs are being found incidentally by ultrasonography or CT [5]. The literature also reports some functional GNs that were found to release peptides such as vasoactive intestinal peptides (VIP), somatostatins, and neuropeptide-Y (NPY) [17,18]. These tumors may cause some symptoms like diarrhea, sweating, and hypertension related to the peptides [2].

Although catecholamine synthesis is an almost constant feature of all neurogenic tumors, GNs rarely lead to symptoms [19]. Consequently, surgeons should be aware of the possibility of hypertensive crisis during the surgery [17,18].

Interestingly, GNs tend to partially or completely surround major blood vessels, with little or no compromise of the lumen [14]. Ichikawa et al [20] reported an interesting contrast enhancement pattern in GN that consisted of delayed heterogeneous uptake. These enhancement features are explained by the presence of an abundance of myxoid matrices in the tumors, resulting in delayed progressive accumulation of contrast material in the extracellular space.

The definitive diagnosis is usually based on histopathological findings after surgical excision of the tumor. Grossly, GNs are large, encapsulated masses of firm consistency with a homogenous, solid, grayish-white cut surface [2].

GNs are fully differentiated and invariably benign. They can be multiple and/or associated with other independent types of neural or neuroendocrine neoplasms [21]. Pathologically, GNs should be differentiated from ganglioneuroblastomas, which have a ganglioneuromatous component of more than 5%; the ganglioneuromatous component of GN is minimal [1].

Management of Ganglioneuroma

Treatment of GN consists of complete surgical resection of the tumor, when possible [5]. Preoperative or postoperative chemotherapy or radiotherapy has no value, unless the GN was associated with ganglioneuroblastoma changes. In this case, there might be some role for chemotherapy [12]. Some investigators have indicated that conservative management in cases that are not amenable to complete resection still results in a good prognosis [3,12]. However, surgery should be performed for the following conditions: symptoms resulting from the tumor, encroachment on vertebral foramina, marked growth in size, and increased secretory activity of catecholamine [12]. It should be kept in mind that GNs have a metastatic potential [10]. For example, one study reported that removal of an abdominal GN with the surrounding lymph nodes revealed another GN in one of the lymph nodes [12].

The possibility of slow progression and late recurrence of GNs has been reported; therefore, long-term periodic radiologic surveillance postoperatively is necessary to assess the malignant potential of these tumors [5,22]. GNs are compatible with long-term, disease-free survival even with unsatisfactory surgical treatment [12]. The prognosis is good [2]. In the case of GN recurrence, surgery should be considered [22].

CONCLUSION

GNs are slow-growing, benign tumors that have a tendency to remain clinically silent for a considerable period of time. The radiological aspect is not specific. Treatment is surgical. Most patients have prolonged survival without any evidence of progression after surgery, but long-term follow-up is recommended.

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