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# Incidental Ovarian Paraganglioma in a 62-Year-Old Woman: A Case Report of Diagnostic Challenge and Surgical Management

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**ABSTRACT:** Paragangliomas are rare neuroendocrine tumors originating from chromaffin cells of the neural crest. While they typically occur in the adrenal medulla as pheochromocytomas or along the sympathetic and parasympathetic chains, paragangliomas in the female genital tract are exceptionally uncommon. This report presents a case of a large right ovarian paraganglioma in a 62-year-old woman with a medical history of hypertension and diabetes. She presented with abdominal pain and fever, which led to a diagnosis of appendicitis, though an unexpected adnexal mass was discovered during surgery. Histological analysis confirmed the tumor as a paraganglioma, with no signs of malignancy. The patient had an uncomplicated recovery and was discharged within four days. Ovarian paragangliomas pose diagnostic challenges due to their rarity and the potential overlap of histological features with other clear-cell ovarian tumors. Despite the generally benign nature of ovarian paragangliomas, their potential for malignancy warrants complete surgical resection and long-term follow-up. This case emphasizes the importance of recognizing paragangliomas in atypical locations, aiding in timely diagnosis and appropriate management.

**KEYWORDS:** Ovarian paraganglioma, neuroendocrine tumor, abdominal mass, ovarian clear cell tumor, extra-adrenal paraganglioma,

## 1. INTRODUCTION

Paragangliomas are rare neuroendocrine tumors, with an incidence of 2–8 cases per million. These chromaffin cell tumors develop from neural crest cells and can be classified based on their origin in parasympathetic or sympathetic ganglia. They represent 10–18% of all chromaffin tumors. [1,2]

Parasympathetic paragangliomas are predominantly found in the neck and skull base, commonly arising in the carotid body or jugulotympanic region. In contrast, sympathetic paragangliomas primarily occur in the adrenal medulla as pheochromocytomas. Extra-adrenal sympathetic paragangliomas can arise anywhere along the sympathetic chain, from the skull base and neck (5% of cases) to the bladder and prostate (10%). Of those occurring along the aorta, 10% are thoracic, while 75% are abdominal, frequently originating in the organ of Zuckerkandl near the root of the inferior mesenteric artery. [3,4]

Paragangliomas in the female genital tract are exceedingly rare, with isolated cases reported in the uterus, vagina, and vulva. Ovarian paragangliomas are particularly uncommon, with only five cases documented in the literature. These tumors are often associated with hypertension and typically exhibit a benign course. Immunohistochemical analysis reveals a consistent phenotype, aiding in differential diagnosis.[5]

This report describes a case of a large right ovarian paraganglioma in a 62-year-old woman. The case is analyzed in detail, including histological and immunophenotypic features, and compared with other clear cell ovarian tumors. Pathologists should recognize the potential for paragangliomas to arise in unusual locations. [6,7]

## 2. AIM OF THE ARTICLE

This case report aims to raise awareness among pathologists and clinicians about the potential for paragangliomas to occur in atypical locations, emphasizing the need for accurate diagnosis and appropriate management.

## 3. PRESENTATION OF CASE

We report the case of a 62-year-old woman with a history of diabetes (controlled with oral medications) and hypertension (under treatment), who presented to the emergency department with right iliac fossa pain persisting for three days, accompanied by vomiting and fever. There were no reports of diarrhea, constipation, or recurrent vomiting during this time. Despite her symptoms, her overall condition remained stable.

On physical examination, the patient appeared to be in good general condition and was afebrile. Tenderness and guarding were noted in the right iliac fossa and hypogastrium, while pelvic examination revealed no abnormalities. Laboratory investigations showed elevated white blood cells (12,000/mm³), a reactive C-reactive protein level of 48 mg/L, and hemoglobin at 10 g/dL.

Abdomino-pelvic ultrasonography revealed signs of appendicitis, with an enlarged appendix measuring 9 mm in diameter, peritoneal effusion, and a left ovarian cyst measuring 5 cm in its longest axis.

The patient underwent emergency laparoscopic surgery. During the procedure, a large left adnexal mass (10 cm in its longest axis) was identified, adherent to the left colon. Due to difficult dissection, laparoscopic conversion to open surgery was required. Intraoperatively, a precise dissection revealed the large adnexal mass at the left colon root, prompting a left adnexectomy (figure 1) along with an appendectomy.

The patient had an unremarkable recovery. Liquids were tolerated on postoperative day two, and she was discharged on postoperative day four.

Histological analysis identified a large ovarian paraganglioma and chronic appendicitis. No additional immunohistochemical markers were positive.

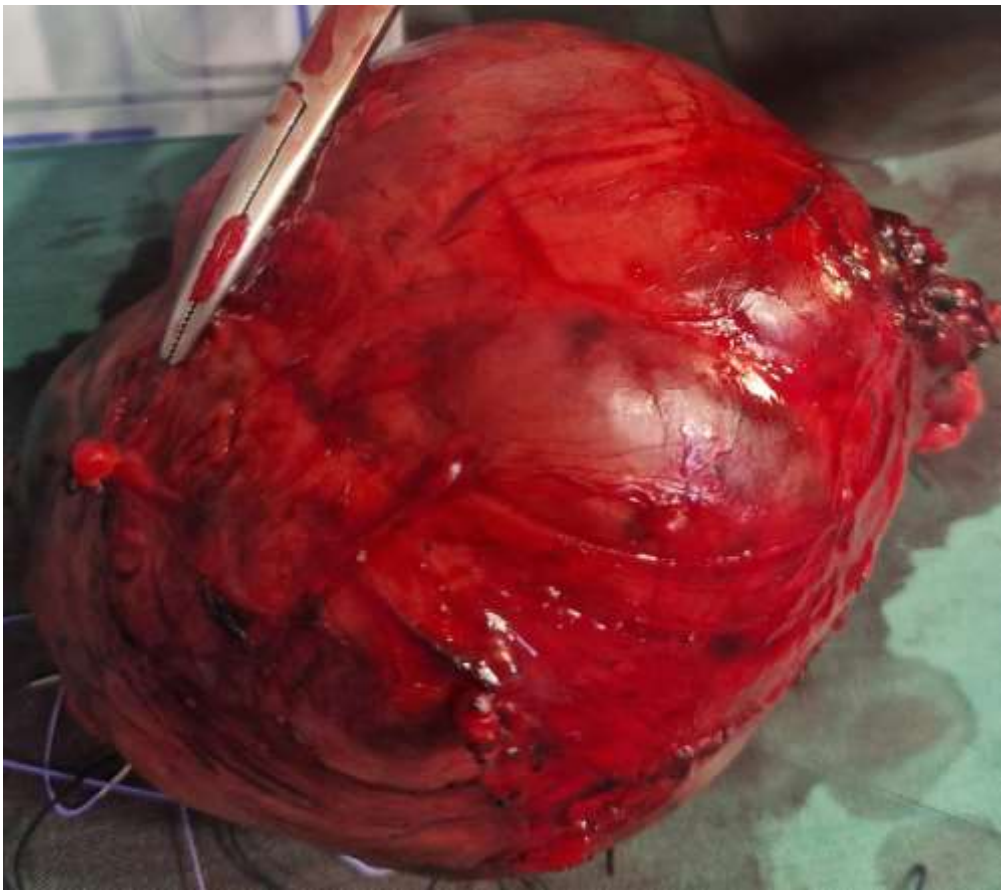

**Figure 1: surgical specimen of the left adnexectomy with the large left ovarian mass.**

## 4. DISCUSSION

Paragangliomas are rare neuroendocrine tumors that originate from chromaffin cells of the neural crest. While most are located in the adrenal medulla (referred to as pheochromocytomas), extra-adrenal paragangliomas may develop along sympathetic or parasympathetic chains. Sympathetic paragangliomas typically produce catecholamines, causing symptoms like hypertension, while parasympathetic tumors are generally non-functional. Despite their relatively well-documented behavior in classic anatomical sites, paragangliomas in unusual locations, such as the ovary, remain poorly understood.[8]

The occurrence of paragangliomas in the female genital tract, including the ovary, is exceedingly rare. Cases reported in the literature have primarily been sporadic, with a range of presentations. In the five documented ovarian cases, the tumor was unilateral in four cases and bilateral in one, with sizes varying from incidental microscopic findings to large masses presenting as palpable abdominal tumors. Some cases were associated with hypertension, while others were asymptomatic. [8,9]

Interestingly, hypertension has been inconsistently observed in ovarian paragangliomas, likely due to variability in catecholamine secretion. In one instance, hypertension resolved postoperatively, supporting a causal relationship. However, in this case, the patient's longstanding hypertension could not definitively be linked to the tumor due to her age and pre-existing cardiovascular condition. [10,11,12]

The origin of ovarian paragangliomas remains speculative. In cases associated with teratomas, the tumor likely arises from neural components within the germ cell tumor. However, in non-teratomatous cases like ours, alternative mechanisms are proposed:

1. Ectopic Paraganglionic Tissue: Migratory failure of neural crest-derived cells during embryogenesis may lead to ectopic paraganglionic tissue within the ovary or other genital sites.
2. Stem Cell Differentiation: Stem cells in the ovary may undergo differentiation into neuroendocrine tissue under specific genetic or environmental influences, a mechanism shared with other rare neuroendocrine tumors such as extra-axial ependymomas.[12]

Ovarian paragangliomas must be differentiated from other clear-cell ovarian tumors, including:

1. Luteinized Sertoli Cell Tumors: These present with clear cuboidal cells in tubular formations and Leydig cells, absent in paragangliomas. Sertoli cell tumors can express neural markers like synaptophysin and chromogranin, making immunohistochemistry challenging.
2. Extra-Axial Ependymomas: These tumors can also display clear polygonal cells but typically show ependymal rosettes and fibrillary cytoplasm, features not present in paragangliomas.[13]

Classic histological examination often remains more reliable than immunohistochemistry, especially in rare cases where staining patterns overlap or contradict established norms. For example, paragangliomas may exhibit low molecular weight cytokeratin positivity, also seen in Sertoli cell tumors, but inhibin—a hallmark of sex-cord stromal tumors—is inconsistently expressed in paragangliomas, adding diagnostic complexity.

Abdominal paragangliomas, particularly extra-adrenal tumors, have a higher likelihood of malignant behavior compared to their adrenal counterparts. In the context of ovarian paragangliomas, malignancy was observed in two of five reported cases: one exhibited lymph node metastasis, and the other invaded adjacent uterine tissue. Both tumors were large (>8 cm), reinforcing the potential association between tumor size and malignant behavior.[14]

In contrast, our case demonstrated benign histological features without metastasis or local invasion. The presence of sustentacular cells in histological examination often indicates benignity, although exceptions exist. Given the unpredictable behavior of these tumors, long-term follow-up is essential.

Imaging is essential for the identification and characterization of paragangliomas. Contrast-enhanced CT and MRI are routinely used but lack specificity for this condition. MRI is particularly sensitive in detecting extra-adrenal tumors. Functional imaging, such as MIBG scintigraphy, is invaluable for distinguishing functional from non-functional tumors, although it was not performed in this case due to the incidental nature of the finding.[15]

Surgical resection is the mainstay of treatment for paragangliomas. Complete excision is critical, particularly for large tumors, given the risk of malignancy. Postoperative management includes regular biochemical testing (plasma metanephrines, urinary catecholamines) and imaging to monitor for recurrence or delayed metastasis.

In our patient, the tumor was discovered incidentally during surgery for appendicitis. Despite its large size (10 cm), there was no evidence of malignancy or recurrence at short-term follow-up. Nevertheless, the potential for late metastases mandates life-long surveillance.

This case emphasizes the importance of considering paragangliomas in the differential diagnosis of rare ovarian masses, particularly those with clear cell morphology and neuroendocrine differentiation. It also highlights the diagnostic challenges posed by overlapping histological and immunohistochemical features with other ovarian tumors.[16]

## 5. CONCLUSION

Ovarian paragangliomas are exceedingly rare, with diverse clinical presentations and variable outcomes. Histological and immunohistochemical analysis remains the cornerstone of diagnosis, complemented by imaging and clinical correlation. Although often benign, their potential for malignancy necessitates thorough surgical excision and life-long follow-up. This case contributes valuable insight into the clinical, pathological, and surgical management of ovarian paragangliomas, furthering understanding of this rare entity.

**PROVENANCE AND PEER REVIEW:**

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**CONSENT**

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

**ETHICAL APPROVAL**

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

**CONFLICTS INTERESTS**

Authors have declared that no competing interests exist.

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