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An Unusual Mesenteric Tumor 'Paraganglioma': A Case Report

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Introduction: Paragangliomas are mostly localized in the adrenal medulla and they are usually pheochromocytomas, derived from the neural crest, but otherwise mesenteric paragangliomas are extremely rare tumors.
Case Presentation: In this article we represent a 59-year-old female with an abdominal mass and pain due to mesenteric paraganglioma.
Conclusions: Paragangliomas can occur as mesenteric tumors; usually, preoperative accurate diagnosis is not possible with imaging methods and precise diagnosis is possible after histological evaluation.

Keywords: Abdominal Neoplasms; Paraganglioma; Tumor

1. Introduction

Paraganglia are a group of non-neural cells derived from the embryonic neural crest. Two types of paraganglia are well known; chromaffin or sympathetic paraganglia, which are made of chromaffin cells and their primary function is endocrinal; nonchromaffin or parasympathetic ganglia, which are made of glomus cells and are primarily responsible for chemoreceptor functions (1). Paraganglia contain tissues such as the adrenal medulla, carotid and aortic bodies, paraganglia in sympathetic and parasympathetic nerves and organs of Zuckerkandl. Paragangliomas are tumors originating from the neuroendocrine elements of the paraganglia. The most common localization of paragangliomas is the adrenal medulla and they are usually pheochromocytomas. Extra-adrenal localization is observed in 5-10% of all paragangliomas (2). The most common extra-adrenal paragangliomas occur as carotid bodies. Paragangliomas have been seen in the gastrointestinal tract of the duodenum. Mesenteric paragangliomas are extremely rare (3).

2. Case Presentation

A 59-year-old woman was admitted to Elazig Training and Research Hospital General Surgery Clinic (Turkey) with abdominal pain in May 2013. Onset of pain was two-three months ago but had gradually increased. Recently, the patient had noticed a mass in her abdomen. The patient's previous medical history included hypertension for 10 years, and she had laparoscopic cholecystectomy eight years ago and right tuba uterine excision due to ectopic tubal pregnancy 30 years ago. Her systolic and diastolic blood pressure was 160/100 mmHg and her pulse was 80/min on systemic examination. She had taken an antihypertensive daily. Her abdominal examination revealed a tender mass in the infraumbilical abdominal region. Other systemic examinations were normal. Laboratory findings included: white blood cell count of 11.58×10^3/L, hemoglobin of 13.6 mg/L and normal liver function. Abdominal ultrasonographic imaging (PVT-375AT, Toshiba Xario ®, Toshiba Medical Systems, Tokyo, Japan) had not resulted any pathological findings two and four months ago, yet the latest abdominal ultrasonography revealed a 42 × 28 mm hypoechoic smoothly marginated mass (Figure 1). Color Doppler ultrasonography showed a markedly hypervascular mass in the infraumbilical intra abdominal region, and the left inguinal region had a 21 × 13 mm necrotic lymphadenopathy. Abdominal computed tomography (CT) (Somatom Definition AS Plus 128, Siemens) revealed a solid mass, which was lobulary 42 × 36 mm in size. Contrast enhanced CT heterogeneously enhanced a hypervascular tumor at the pelvic mesenteric region (Figure 2). The patient underwent surgery, and a 6 cm diameter mass lying on the serosal wall of the terminal ileum originating from the small bowel mesentery was seen at the laparotomy. A section of the ileum and the mass were resected. These

features are described in Table 1. The pathological examination revealed a cellular neoplasm and included a characteristic Zellballen pattern (Figure 3). Immunohistochemically tumor cells were positive for CD138, S-100 (in sustentacular cells), CD34 (in vessels), Chromogranin and Synaptophysin. PanCK, CD68, Vimentin, Actin CD117, DOG1 were negative. Ki 67 proliferating index was found to be 1% (Figure 4).

3. Discussion

To the best of our knowledge a very small number of cases of mesenteric paraganglioma have been described in the English literature. Fujita et al reported that in total only 12 cases have been documented in the English literature. Generally these tumors are seen in females and the elderly. The patient's median age is 57.5 years; our patient was female and aged 59 years old. On the other hand retroperitoneal paragangliomas are predominantly seen in males and the median age is 39-43 years. The average size of mesenteric paragangliomas is 9.3 cm but this case had a 6 cm tumor. This is probably because our patient was slim and thus the mass was noticed earlier (2).

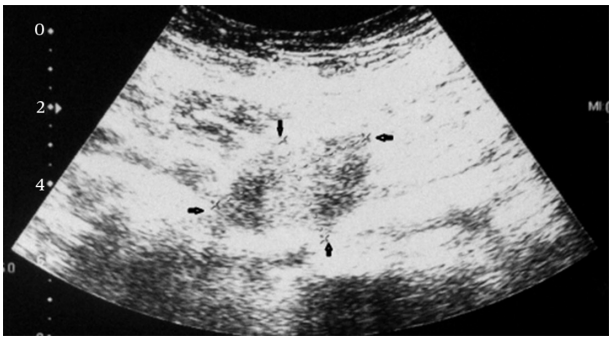


Figure 1. Black Arrows Point to the Margins of the 42 × 28 mm Hypoechoic, Smoothly Marginated Mass on the Abdominal Ultrasonography

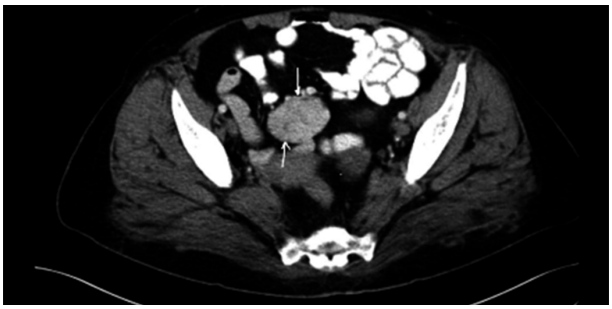
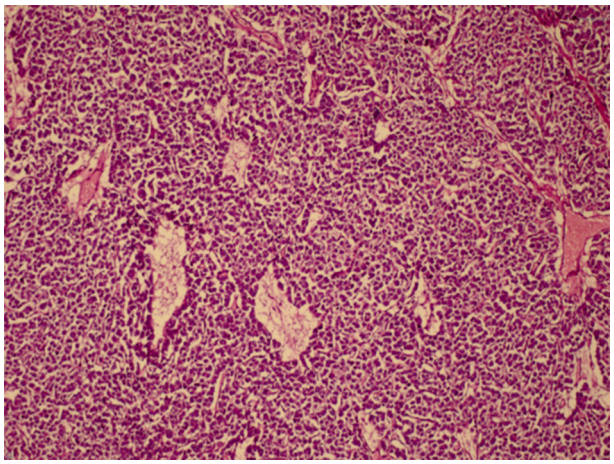


Figure 2. Contrast Enhanced Axial Abdomen CT: Arrows Represent a Heterogeneously Enhanced Hypervascular Tumor in the Pelvic Region

Figure 3. Microscopic View of the Specimen With Hematoxyin and Eosin Staining, 100 ×



Group of cells were arranged with a characteristic 'nested Zellballen' pattern and separated with fibrovascular connective tissue.

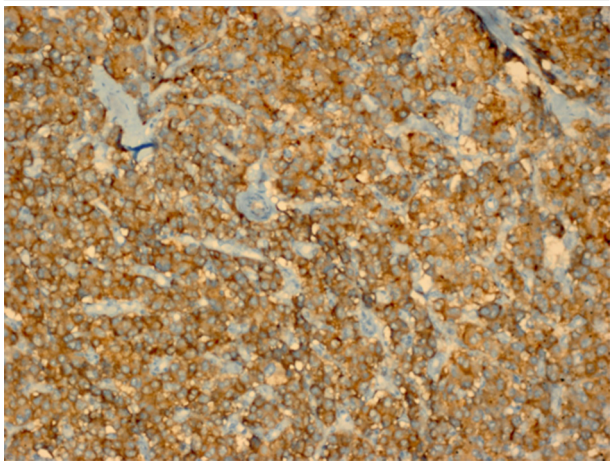


Figure 4. Confirmation of Neuroendocrine Origin Was Done by Identification Of Synaptophysin, Supporting the Diagnosis of Paraganglioma, 100 ×

Table 1. Main Features of the Subject of This Study and Previous Cases Found from the Literature

Age	Gender	Size	Location Size	Surgical Procedures	Prognosis
Other cases (n = 12)	Male/Female 3/9	9.1 cm	mesentery of small intestine/ colon 10/2	intestinal and mass resection/ mass resection 9/2	16.5 month (8 documented survivals), no recurrence
This case	Female	6 cm	mesentery of small intestine	intestinal and mass resection	14 month alive, no recurrence

Paragangliomas can be functional such as paroxysmal episodic hypertension, palpitations, headache, increased sweating due to secretion of catecholamines, yet extra-adrenal paragangliomas are generally nonfunctional as in our case (4). Only 25% of mesenteric paragangliomas have functional features (5). In our case, blood pressure was rising mildly and became stable with antihypertensive drugs. This condition continued after surgery. Diagnosis of functional paragangliomas is easier than non-functional tumors; analysis of plasma or urinary metanephrines is very sensitive when there is doubt about paraganglioma. Otherwise, imaginary techniques such as ultrasound (US), CT, magnetic resonance imaging (MRI) are almost equally effective for the detection of non-functional tumors. Scintigraphy or Positron emission tomography (PET) scan can be effective for the detection of the features of masses (4). We could not identify the tumor type preoperatively because of our technical issues. Laparoscopy and biopsy are recommended for diagnosis, and tumor resection with laparoscopy or laparotomy is essential for treatment. Tumor recurrence has not been reported in the current literature. We performed laparotomy and extended bowel and mesentery resection in this case and she had no recurrence for 14 months. We could not perform a laparoscopic operation because the tumor size was large and we had technical insufficiencies. Extra-adrenal paragangliomas are more aggressive than other paragangliomas, malignancy rates vary between 14, 7 and 50% (3). Histological evaluation is not enough for differentiation of benign or malignant tumors but presence of metastasis is essential for malignancy. The recent World Health Organization classification indicates that mitotic counts and the Ki-67 labeling index are significant for determining the presence of malignancy (2). We did not detect any metastasis in our case and Ki-67 labeling index was low and mitosis was rare. Mesenteric paragangliomas are extremely rare tumors, and usually require differential diagnosis from gastrointestinal stromal tumors, lymphoma and leiomyoma. Preoperative accurate diagnosis is important yet precise diagnosis is obtained by histological evaluation. Surgical intervention is essential for treatment.

Authors' Contributions

Study concept and design: Zeynep Ozkan. Acquisition of data: Yeliz Gull and Mustafa Koc. Analysis and interpretation of data: Onder Altas and Cengizhan Ozdemir; drafting of the manuscript: Zeynep Ozkan; critical revision of the manuscript for important intellectual content: Emre Durdag; administrative, technical and material support: Gunay Yasar; study supervision: Emre Durdag.

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