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A Rare Case of Gangliocytic Paraganglioma presented as GIST- A Case Report

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ABSTRACT

INTRODUCTION: Paragangliomas are neuroendocrine tumors that most commonly originate in the adrenal gland, a type that is called pheochromocytoma; however, 5-10% of paragangliomas are extra-adrenal and may arise in any area between the neck and pelvic region along the sympathetic nervous system. At presentation, it may be confused with a gastrointestinal stromal tumor (GIST), but distinguishing between these tumors is critical because the natural history and treatment of these two tumors differs markedly.

CASE REPORT: A 70 year old male presented with altered bowel habits and distension of abdomen with CT scan showing GIST in the second part of duodenum.

CONCLUSION: Paragangliomas are considered benign tumors, but they occasionally display a malignant character. The most important finding in this case was the need for total resection of the tumor to avoid recurrence.

KEYWORDS Paraganglioma, Neuroendocrine Tumors, Duodenum.

INTRODUCTION

Gangliocytic paraganglioma is a rare tumor that is located in the duodenum in 90% of cases with a particular predilection to the region of the ampulla of Vater. [1] At presentation, it may be confused with a gastrointestinal stromal tumor (GIST), but distinguishing these tumors is critical because the natural history, and therefore, treatment of these two tumors differs markedly.[2] The most common clinical presentation includes melena and abdominal pain [1]. This tumor typically exhibits benign behavior with regional lymph node metastasis occurring in only 5–7% of cases with no tumor associated deaths reported –[1,38]. Recurrence after resection is rare, with only a single reported recurrence many years after initial resection [9]. Herein, we report a case of surgically resected duodenal gangliocytic paraganglioma.

CASE REPORT

A 70-year-old man was admitted to a tertiary care hospital with complaints of dyspepsia , weakness and abdominal distension. Laboratory findings showed deranged LFT with increased Total Bilirubin levels. Computed tomography (CT) suggested diagnosis of GIST in second part of duodenum. The patient underwent pancreaticoduodenectomy, regional lymphadenectomy and the mass was sent for histopathological examination.

HISTOPATHOLOGY

Grossly, a round to oval mass measuring 7 x 5 x 3 cm was received . Specimen was fixed in neutral buffered formalin and embedded in paraffin.

Tissue sections were stained with routine hematoxylin and eosin (HE). Histological examination revealed a tumor extending into the submucosa and muscularis propria. The tumor was composed of epitheloid cells, arranged in trabecular or pseudoglandular pattern, spindle cells, and ganglion cells with abundant cytoplasm and vesicular nuclei. Also seen is clusters of epithelioid cells (Zellballen configuration) and branching vascular sinusoids. There was mild pleomorphism with no significant mitotic activity or necrosis. No metastasis was found in the regional lymph nodes.

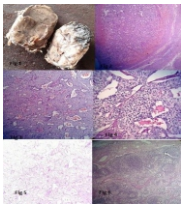

Figure 1 shows gross appearance of tumor mass

Figure 2-5 (H& E stain) shows spindle cells, ganglion cells ,epithelioid cells in Zellballen Configuration

Figure 6 (H & E stain) shows lymph node free from tumor involvement

Immunohistochemically, the epitheloid cells were positive for cytokeratin , chromogranin A, and synaptophysin . S-100 protein labeled the sustentacular cells and the spindle cell component . Ganglion cells were positive for synaptophysin .

Due to its spindle-cell predominance, our main differential diagnosis included nerve sheath tumors (schwannoma) and GIST. The lack of diffuse positivity for S-100 and the presence of scattered ganglion cells excluded the possibility of a schwannoma. Negative immunohistochemical stains for CD117 and CD34 as well as the sustentacular pattern of S-100 protein argued against a GIST.

DISCUSSION

Gangliocytic paraganglioma is an extremely rare benign neuroendocrine tumor of the gastrointestinal tract. Most GPs are characteristically located in the second portion of the duodenum with a predilection for the ampulla vateri. Duodenal GP has been reported in patients 15 to 84 years of age with a male predominance as in our case. The patients with GP clinically present with abdominal pain, gastrointestinal bleeding, or they can be asymptomatic. Duodenal gangliocytic paraganglioma consists of three distinct cellular elements: spindle cells, epithelial cells and ganglion cells.[3,4] In spindle-cell predominant tumors, the differential diagnosis includes schwannomas and GIST. Tumor behavior is usually benign with regional lymph node metastasis occurring in about 5–7% of cases [1,3–8] as in our case. Endoscopic resection of duodenal gangliocytic paraganglioma appears to be safe and effective in cases in which the tumor may be removed in its entirety by endoscopic methods [1,4,5].

It may be difficult to make a differential diagnosis, if endoscopic biopsy specimens do not contain all three histologic components. The spindle cell tumors, epithelial tumors or ganglioneuroma may be recognized according to the presence of three related different elements, as in the current case. The histological differential diagnosis of duodenal GP includes well-differentiated neuroendocrine carcinoma, ganglioneuroma, paraganglioma,

and spindle cell malignancies (nerve sheath, smooth muscle, and gastrointestinal stromal tumors)[10].

CONCLUSION

Localized duodenal gangliocytic paragangliomas may be confused with GIST and are best managed by resection with negative margins. Endoscopic removal is safe and effective in cases in which the tumor is suspended by a stalk and local and regional disease is absent. Although GPs are accepted as a benign tumors, careful assessment is necessary for recurrences or metastases.

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