Case Report

Gastric Adenomyoma: A Rare Cause of Pyloric Mass in Children

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INTRODUCTION

Gastric adenomyoma is a rare tumor with a benign course. Children may present with nonspecific symptoms but can also exhibit urgency such as melena and gastric outlet obstruction (GOO). Surgical resection is the cornerstone in its treatment. Histologically, it is composed of epithelial elements surrounded by a smooth stroma. Herein, we present a rare case of gastric adenomyoma in a 12-year-old child to highlight the diagnostic dilemma and the importance of surgical resection and histopathology in management.

CASE REPORT

A 12-year-old boy presented with complaints of episodic, dull-aching pain in the left upper abdomen for the past 1 month. There was associated history of episodes of nonbilious vomiting with the pain. The child underwent upper gastrointestinal endoscopy elsewhere, where a submucosal bulge in the pyloric region was documented, and an endoscopic biopsy from the site of the bulge was suggestive of a spindle cell neoplasm. He was referred to our center for further management.

On presentation, his general condition was well preserved. On examining the abdomen, no lump or organomegaly was found. Ultrasonography (USG) of the abdomen revealed circumferential thickening in the distal body of the stomach. Subsequently, a contrast-enhanced computed tomography (CECT) scan of abdomen confirmed the presence of a heterogeneously enhancing mass in the anteroinferior wall of the gastric antrum causing luminal obstruction [Figure 1a]. This mass also showed focal fluorodeoxyglucose (FDG) uptake [Figure 1b] on positron emission tomography (PET scan). Investigations were done to rule out tuberculosis and lymphoma as a possible cause of distal gastric and pyloric thickening.

With a working diagnosis of gastrointestinal stromal tumor (GIST), an exploratory laparotomy was planned. A circumferentially thickened and bulky pylorus was found with normal duodenum and proximal stomach. The abnormal pylorus was completely excised [Figure 2a], and a gastroduodenostomy was performed with a transanastomotic tube. The histopathology revealed the expansion of the submucosa and muscularis by smooth muscle cells [Figure 2b]. The presence of Brunner’s glands [Figure 2c], positive staining for periodic acid-Schiff [Figure 2d] and CK7 [Figure 2e] and negative staining for synaptophysin [Figure 2f] were consistent with the diagnosis of gastric adenomyosis. The margins of resection were free.

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The patient was asymptomatic after a year of follow-up and a PET scan done (3 months after resection) did not reveal any FDG uptake. Serial long-term follow-up with USG has been planned to rule out local recurrences if any.

**DISCUSSION**

Gastric adenomyoma, also known as adenomyosis and myoepithelial hamartoma is a rare benign entity. Only 52 cases were reported in the literature till 2017. Although the age at presentation may vary, most cases present between fourth to sixth decades of life. The youngest case described in literature is a 1-week-old neonate. No gender preponderance has been seen for this tumor. These tumors most commonly involve the antrum (85%) and pyloric (15%) regions of the stomach.

The etiopathogenesis of gastric adenomyoma is a gray zone, and there are two schools of thought. While the majority consider its occurrence as a developmental aberration, there is considerable evidence of it being an abortive variant of heterotopic pancreas. The presence of epithelial and smooth muscle components supports the former ideology and coexistence with heterotopic pancreas and histology justifies the latter. It can also present with other conditions such as gastric duplication, annular pancreas, and duodenal adenomas.

Children with gastric adenomyoma usually present with nonspecific clinical features such as episodic pain, nausea, vomiting, and melena. There are some reports of intermittent pyloric obstruction in patients with gastric adenomyoma as well. Imaging techniques such as USG and CECT of the abdomen are usually noncontributory, and the diagnosis of gastric adenomyoma is made only by histopathology in majority of the cases. Similarly, investigations such as UGI contrast series and endoscopy cannot differentiate between conditions such as gastric adenomyoma, GIST, leiomyoma, lymphoma, and lipoma and carcinoid tumor. Thus, it is not uncommon to misdiagnose it as some other pathology. In our case also, these investigations did not lead to a diagnosis. One peculiar finding was the focal FDG uptake by the thickened pylorus which has not been described previously in gastric adenomyoma. The mechanism behind this uptake is a matter of speculation since a benign process like adenomyomas is unlikely to be FDG avid. Despite this biological behavior, a repeat
PET scan was performed, and it did not show any FDG uptake.

The management consists of surgical resection of the tumor, and postoperative histopathology is recommended for making a correct diagnosis.[5] While resection of the tumor with an adequate margin is indicated in most of the cases, limited resection should be preferred in tumors with involvement of the periampullary region. In these cases, a frozen section can aid in making a preemptive diagnosis and can avoid the morbidity of a more radical surgery.[3] Endoscopic submucosal resection is also a feasible treatment modality in adults as reported by Wang et al. in their series of 15 cases of gastric adenomyoma.[1] The risk of malignant transformation is rare (<2%) and is similar to that of hamartomatous polyps. Histopathology plays a vital role in making a diagnosis of gastric adenomyoma. On gross examination, the mass arises in the submucosa and protrudes into the lumen of the stomach. Microscopically, the tumor shows the presence of epithelial component (tubules, Brunner’s glands, and Pancreatic acini) surrounded by smooth muscle stroma.[6]

**CONCLUSION**

A gastric adenoma is a rare cause of pediatric GOO. Endoscopic biopsy may miss the diagnosis, and therefore, complete surgical resection should be attempted after ruling out other common causes of GOO. The presence of characteristic histopathological findings should confirm the diagnosis.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Wang S, Cao H, Zhang Y, Xu M, Chen X, Piao M, et al. Endoscopic submucosal dissection for gastric adenomyoma: A rare entity of 15 cases among 571 patients with gastric submucosal eminence lesions. Medicine (Baltimore) 2017;96:e6233.
2. Duran Álvarez MA, Gómez López JR, Guerra Garijo T. Gastric adenomyoma: The unexpected mimicker. GE Port J Gastroenterol 2017;24:198‑202.
3. Nabi J, Authoy FN, Akhter SM. Atypical presentation of myoepithelial hamartoma in the antrum of the stomach, mimicking a gastrointestinal stromal tumor: A case report. J Med Case Rep 2012;6:382.
4. Lasser A, Koufman WB. Adenomyoma of the stomach. Am J Dig Dis 1977;22:965‑9.
5. Hedenbro JL, Ekelund M, Wetterberg P. Endoscopic diagnosis of submucosal gastric lesions. The results after routine endoscopy. Surg Endosc 1991;5:20‑3.
6. Cimmino CV. Gastric adenomyosis vs. aberrant pancreas. Radiology 1955;65:73‑7.