International Journal of Surgery Case Reports 86 (2021) 106336

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Granular cell tumor presenting with perforation of fourth part of the duodenum: A case report

Erin Templeton [a,*], Christina Eliachevsky [a], Atul K. Nanda [b]

[a] St. George's University School of Medicine, Humboldt Park Health, 1044 N Francisco Avenue, Chicago, IL 60622, USA
[b] Humboldt Park Health, 1044 N Francisco Avenue, Chicago, IL 60622, USA

ARTICLE INFO

Keywords:
Granular cell tumor
Duodenal perforation
GCT duodenum
GCT jejunum

ABSTRACT

Introduction: Granular cell tumors (GCT) are relatively rare neoplasms most commonly occurring in skin or soft tissues. GCT are thought to be of Schwann cell origin and strongly positive for s100 protein. GCT of the intestinal tract are usually asymptomatic and found incidentally in the esophagus on endoscopy.

Case presentation: Here, we present a case of GCT jejunum and the fourth part of the duodenum. The patient is a 41-year-old female who presented with abdominal pain and was subsequently found to have pneumoperitoneum with a perforation of the fourth part of the duodenum. Intraoperatively, there were multiple enlarged and hard mesenteric lymph nodes, which were found to be due to GCT involving the fourth duodenum and proximal jejunum.

Clinical discussion: The occurrence of GCT in the gastrointestinal (GI) tract are even less common accounting for 5–9% of all GCT with very few cases reported in the duodenum. GCT of the GI tract are often asymptomatic, consequently leading to misdiagnosed delays in treatment.

Conclusion: In the setting of GCT in the fourth part of the duodenum with evidence of locally advanced disease, local resection is the preferred treatment.

1. Introduction

Granular cell tumors are typically positive for s100 protein, supporting the theory that GCT are of Schwann cell origin [1]. In 1931 Abrikossof reported the first GCT of the GI tract, occurring in the esophagus [2]. GCT can occur within any organ, although most frequently they are located in the oral cavity (40%), skin (30%) and, breast (15%) [3,4]. Approximately 5–9% of GCT develop in the GI tract, of which 65% occur in the esophagus [5,6]. Most GCT are solitary submucosal tumors which are typically asymptomatic and present as incidental findings on endoscopy [6]. GCT occurring in the duodenum is extremely rare with only three cases listed in review. Johnston and Helwig reviewed 75 cases of GCT in 1981 reporting only one case in the duodenum [7]. In 1998 Odona et al. reported a benign GCT in the duodenum presenting with significant hemorrhage warranting surgical resection [8]. Most recently in 2006, Woosley and Grimm reported a duodenal GCT discovered on endoscopy requiring surgical resection for symptomatic improvement [9]. We present a unique case of GCT of the fourth part of the duodenum and jejunum managed in a community hospital. All the following work is reported in line with SCARE criteria [10].

2. Case report

A 41-year-old female with no pertinent past medical or surgical history presented to the emergency department with sudden onset abdominal pain associated with several episodes of emesis. Abdominal examination revealed diffuse tenderness and peritonitis. CT abdomen/pelvis revealed pneumoperitoneum with the most likely cause being duodenal perforation warranting emergent exploratory laparotomy [Image 1]. Intraoperative examination revealed a perforation of the anterior wall of the fourth part of the duodenum.

The mucosa proximal to site of the duodenal perforation was noted to be hypertrophic and congested. Further investigation revealed contusion and thickening of the duodenal and proximal jejunal mesentery with surrounding fibrosis and thickening of the jejunum [Image 2]. Fibrosis and multiple enlarged, hard lymph nodes in the jejunal mesentery, retroperitoneum, and lesser sac were identified [Image 3]. The perforation was repaired with an omental patch by an experienced general surgeon. Subsequently, gastrojejunostomy was performed

* Corresponding author.
E-mail addresses: etemplet@sgu.edu (E. Templeton), celiache@sgu.edu (C. Eliachevsky), ananda@nahospital.org (A.K. Nanda).

https://doi.org/10.1016/j.ijscr.2021.106336
Received 16 March 2021; Received in revised form 13 August 2021; Accepted 21 August 2021
Available online 25 August 2021

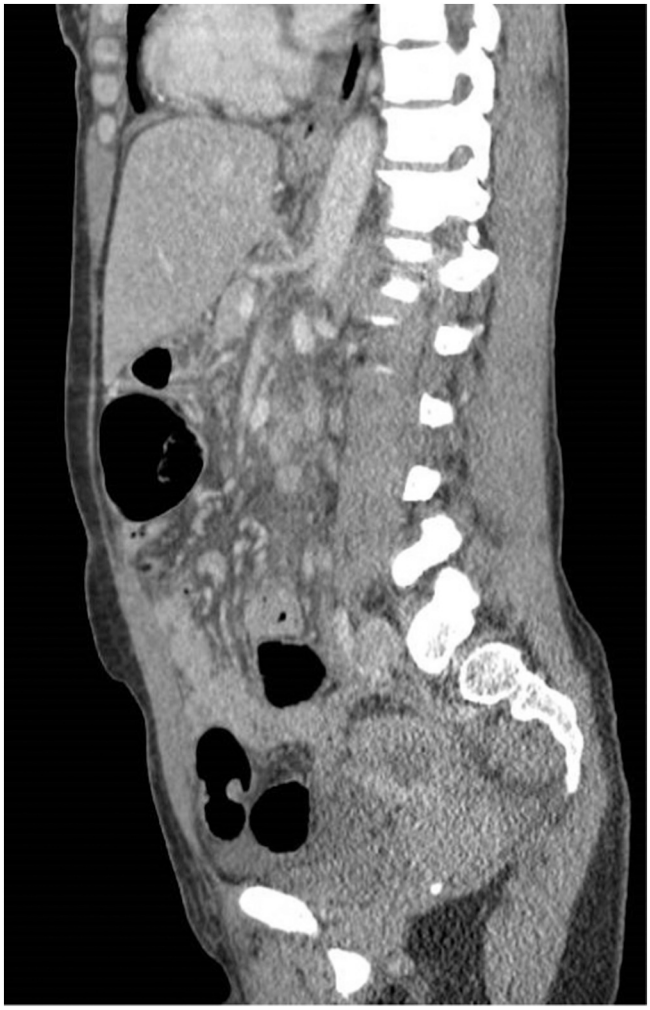

Image 1. Transverse plane CT of abdomen demonstrating small foci of pneumoperitoneum.

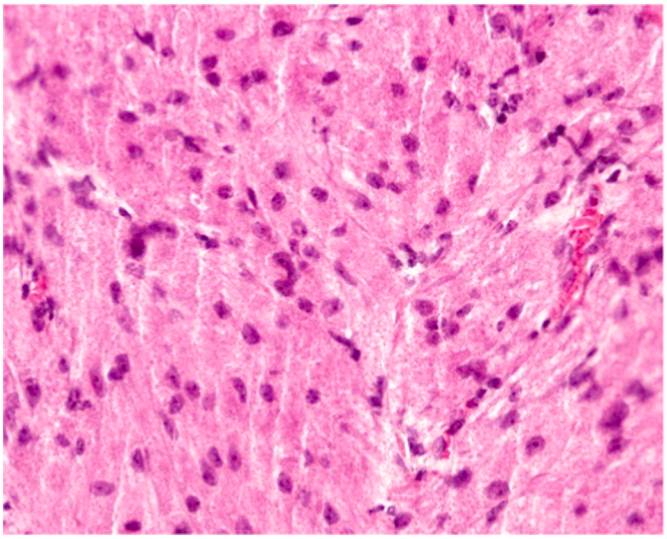

Image 2. Coronal plane CT of abdomen/pelvis demonstrating nonspecific, mild to moderate edema of the mesentery and air around the duodenal perforation.

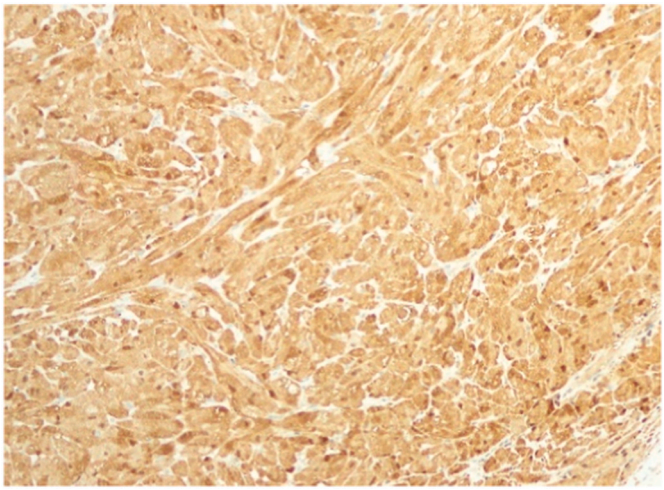

Image 3. Sagital plane CT of abdomen/pelvis demonstrating edema of jejunal mesentery and enlarged mesenteric and retroperitoneal lymph nodes.

bypassing the perforation and surrounding area of involvement.

Perforation site biopsy revealed acute erosive duodenitis with full thickness ulceration. The mesenteric lymph node biopsy revealed GCT which demonstrated nests of round polygonal cells with abundant granular eosinophilic cytoplasm on histology [Fig. 1]. Immunohistochemical stains performed indicated s100 positive (4+), negative CD34, Ki67, DOS-1, c-KIT and PAS [Fig. 2]. According to the Fanburg-Smith criteria, if three of more of the following histologic criteria are met, a GCT is considered malignant: necrosis, spindling, vesicular nuclei with large nucleoli, increased mitotic activity, high nuclear: cytoplasmic ratio, and pleomorphism [11]. According to this classification system, this patient's GCT is benign as no criteria was met. Post-operatively the patient is scheduled for CT abdomen with contrast and esophagogastroduodenoscopy for further investigation of possible additional tumor burden. Follow-up has been limited due to patient compliance and COVID-19 pandemic.

3. Discussion

GCT of the GI tract are most commonly benign, slow growing, and submucosal in location [12]. GCT possess malignant potential, typically in larger lesions. In a review of 183 cases 4% were malignant, all of

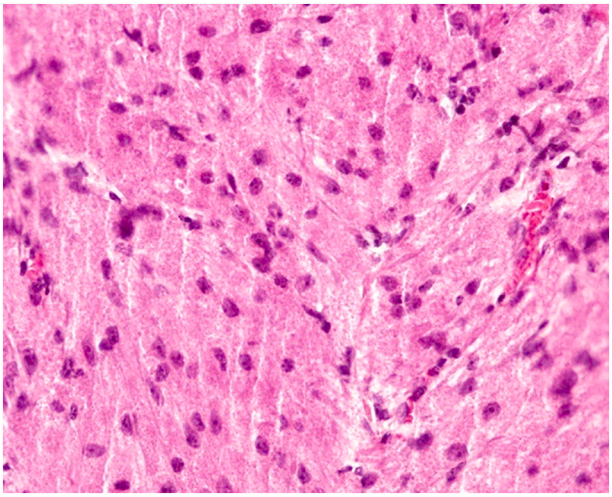

Fig. 1. Light-microscopy view of mesenteric lymph node demonstrating nests of polygonal cells with abundant granular eosinophilic cytoplasm.

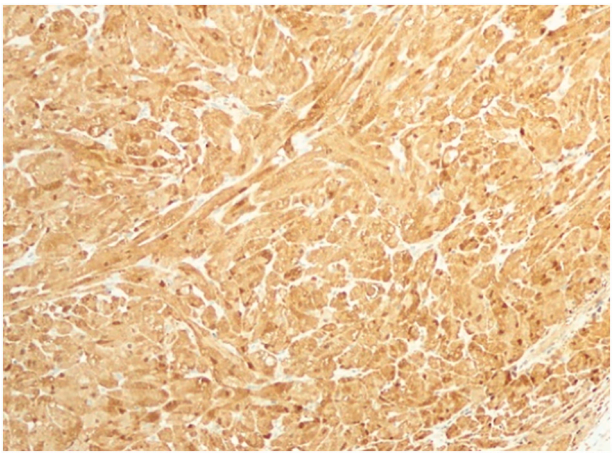

Fig. 2. Light-microscopy view of mesenteric lymph node demonstrating strong (4+) s100 immunohistochemical positivity.

which were greater than 4 cm in size [13]. Malignant GCT tend to be aggressive with high rates of metastasis and recurrence, thus having a poor prognosis [14]. Conversely, benign intestinal GCT have excellent prognosis with low recurrence rate after surgical resection, with a curative rate of 92% and a recurrence rate of 2–8% [3]. Due to their rarity, treatment of intestinal GCT treatment is controversial. Whether tumors should be monitored or surgically resected is debated [15]. Historically, local excision has been the preferred treatment for intestinal GCT [15]. Complete local surgical excision is curative for benign granular cell tumor, while excision with wide margins is recommended for malignant lesions [14].

GCT have traditionally been considered radioresistant [16]. The use of radiotherapy and chemotherapy for benign lesions is not recommended. The role of adjuvant radiotherapy and chemotherapy for the treatment of malignant GCT is a matter of dispute. Adjuvant radiotherapy should be considered in cases of malignant GCT characterized as large in size, with positive postresection margins, and those with high mitotic rates [17]. In 1990 Rosenthal reported a case where adjuvant radiotherapy was used in the treatment of a large locally recurrent cutaneous GCT with clinical features suggestive of malignant potential, with good response and no recurrence [16]. However, it should be noted there is little evidence to support the use of neoadjuvant radiotherapy in the treatment of intestinal GCT prior to complete resection.

GCT are usually solitary neoplasms. In review, 32 cases have been reported of multiple GCT of the esophagus [18]. Only a few cases report multiple solitary GCT occurring throughout the colonic tract. In 1993, Melo et al. reported a case of 52 GCT spanning from the cecum to the sigmoid [19]. A conservative approach was employed, opting for observation with regular colonoscopies, reserving surgical management once the patient became symptomatic. In 2009, Saleh reported multiple synchronous GCT involving the colon, appendix and mesentery [20]. In review, Saleh reported the first case of multiple colonic GCT extending to the mesentery. On thorough review, we are reporting the first case of a GCT of jejunum and the fourth part of the duodenum with involvement of mesentery, and lesser sac.

In the setting of this patient's GCT in the fourth part of the duodenum, local resection would have been the preferred treatment. However, due to the location of the tumor and given its local spread evidenced by enlarged and hard lymph nodes of the in the jejunal mesentery and lesser sac, complete surgical resection is technically challenging. Radiotherapy was considered but due to the proximity of the lesion to the small bowel, the risk of perforation was too high. There are insufficient data to predict the prognosis of benign small intestine GCT. However, it might be expected to have a favorable prognosis, similar to GCT at other locations [15].

4. Conclusion

We reported a case of a GCT of the fourth part of the duodenum presenting with perforation. Acute management focused around repair of the duodenal perforation with biopsy. This case reports exemplifies the role of intraoperative biopsy as the patient was incidentally found to have locally advanced GCT. GCT are rare soft-tissue tumors with few cases reported occurring in the duodenum. After stabilization, further tumor work up is required to determine the appropriate surgical management. The recommended treatment of GCT of the intestinal tract is local resection. However, due to location and evidence of advanced local pathological process, surgical resection and radiotherapy held too great of a risk for the patient presented in this case report.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Ethical approval

The study is exempt from ethical approval in our institution.

Registration of research studies

Not applicable.

Guarantor

Erin Templeton
Christina Eliachevsky
Nanda K Atul

CRediT authorship contribution statement

Erin Templeton: Writing – Original Draft, Writing – Review & Editing.

Christina Eliachevsky: Writing – Review & Editing.

Atul K. Nanda: Methodology, Writing – Review & Editing, Supervision

Declaration of competing interest

The authors have no competing interests.

Acknowledgement

Thank you to pathologist, Aisha M Jaleel, for providing the histology slides.

References

[1] K. Stefansson, R. Wollmann, M. Jerkovic, S-100 protein in soft-tissue tumors derived from Schwann cells and melanocytes, Am. J. Pathol. 106 (2) (1982) 261–268.

[2] A. Abrikossoff, On fibroids, Virchows Arch. Path Anat. 260 (1926) 215–233, https://doi.org/10.1007/BF02078314.

[3] E.E. Lack, G.F. Worsham, M.D. Callihan, et al., Granular cell tumor: a clinicopathologic study of 110 patients, J. Surg. Oncol. 13 (4) (1980) 301–316, https://doi.org/10.1002/jso.2930130405.

[4] G.R. McSwain, R. Colpitts, A. Kreutner, et al., Granular cell myoblastoma, Surg. Gynecol. Obstet 150 (1980) 703–710.
[5] J.R. Goldblum, T.W. Rice, G. Zuccaro, J.E. Richter, Granular cell tumors of the esophagus: a clinical and pathologic study of 13 cases, Ann. Thorac. Surg. 62 (3) (1996) 860–865, https://doi.org/10.1016/s0003-4975(96)00443-2.
[6] O. David, S. Jakate, Multifocal granular cell tumor of the esophagus and proximal stomach with infiltrative pattern: a case report and review of the literature, Arch. Pathol. Lab. Med. 123 (10) (1999) 967–973, https://doi.org/10.1043/0003-9985(1999)123<0967:MGCTOT>2.0.CO;2.
[7] J. Johnston, E.B. Helwig, Granular cell tumors of the gastrointestinal tract and perianal region: a study of 74 cases, Dig. Dis. Sci. 26 (9) (1981) 807–816, https://doi.org/10.1007/BF01309613.
[8] N. Onoda, H. Kobayashi, K. Satake, et al., Granular cell tumor of the duodenum: a case report, Am. J. Gastroenterol. 93 (10) (1998) 1993–1994, https://doi.org/10.1111/j.1572-0241.1998.00566.x.
[9] J.T. Woosley, I. Grimm, Granular-cell tumor of the duodenum: a case report, Gastrointest. Endosc. 63 (2) (2006) 339–341, https://doi.org/10.1016/j.gie.2005.07.023.
[10] for the SCARE group, A. Agha Riaz, R. Borrelli Mimi, Farwana Reem, Koshy Kiron, J. Alexander, Dennis Fowler, P. Orgill, The SCARE 2018 statement: updating consensus surgical Case Report (SCARE) guidelines, Int. J. Surg. 60 (2018) 132–136.
[11] J.C. Fanburg-Smith, J.M. Meis-Kindblom, R. Fante, L.G. Kindblom, Malignant granular cell tumor of soft tissue: diagnostic criteria and clinicopathologic correlation [published correction appears in am J Surg Pathol 1999Jjan; 23(1): 136], Am. J. Surg. Pathol. 22 (7) (1998) 779–794, https://doi.org/10.1097/00000478-199807000-00001.
[12] F. Radaelli, G. Minoli, Granular cell tumors of the gastrointestinal tract: questions and answers, Gastroenterol. Hepatol. (N Y) 5 (11) (2009) 798–800.
[13] J. Orlowska, J. Pachlewski, A. Gugulski, E. Butruk, A conservative approach to granular cell tumors of the esophagus: four case reports and literature review, Am. J. Gastroenterol. 88 (2) (1993) 311–315.
[14] V.A. Singh, J. Gunasagaran, J. Pailoor, Granular cell tumour: malignant or benign? Singap. Med. J. 56 (9) (2015) 513–517, https://doi.org/10.11622/smedj.2015136.
[15] M. Barakat, A.A. Kar, S. Pourshahid, et al., Gastrointestinal and biliary granular cell tumor: diagnosis and management, Ann. Gastroenterol. 31 (4) (2018) 439–447, https://doi.org/10.20524/aog.2018.0275.
[16] S.A. Rosenthal, V.A. Livolsi, A.T. Turrisi 3rd., Adjuvant radiotherapy for recurrent granular cell tumor, Cancer 65 (4) (1990) 897–900, https://doi.org/10.1002/1097-0142(19900215)65:4<897::aid-cncr2820650413>3.0.co;2-1.
[17] Arvind Krishnamurthy, et al., Malignant granular cell tumor of the tongue: a clinico-pathological challenge, Indian J. Surg. Oncol. 5 (1) (2014) 71–74, https://doi.org/10.1007/s13193-013-0283-2.
[18] J. Szumilo, D. Skomra, K. Zinkiewicz, W. Zgodzinski, Multiple synchronous granular cell tumours of the esophagus: a case report, Ann. Univ. Mariae Curie Sklodowska Med. 56 (2001) 253–256.
[19] C.R. Melo, I.S. Melo, F.C. Schmitt, R. Fagundes, D. Amendola, Multicentric granular cell tumor of the colon: report of a patient with 52 tumors, Am. J. Gastroenterol. 88 (10) (1993) 1785–1787.
[20] H. Saleh, M. El-Fakharany, M. Frankle, Multiple synchronous granular cell tumors involving the colon, appendix and mesentery: a case report and review of the literature, J. Gastrointestin. Liver Dis. 18 (4) (2009) 475–478.