EDORIUM Journals

Int J Case Rep Images 2023;14(1):123–128.
www.ijcasereportsandimages.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Duodenal gangliocytic paraganglioma: A case report

Houssam Eddine Bitar, Mohamad Rakka,
Maureen Chbat, Rim Ibrahim, Bassam Matar, Aziz Karaa

ABSTRACT

Introduction: Duodenal gangliocytic paraganglioma (DGP) is a benign tumor that only warrants a surgical resection in the majority of cases.

Case Report: We are reporting a case of 42-year-old man who consulted for significant weight loss, fatigue, anemia, and an on–off obstructive jaundice. Imageries showed a 5 cm peri-ampullary duodenal mass. We couldn't have a diagnosis by endoscopic ultrasound (EUS), so we did a laparoscopic transduodenal tumorectomy and removed the entire mass, which came back as gangliocytic paraganglioma on pathology and immunohistochemistry. Gangliocytic paraganglioma is a benign tumor that most commonly occurs in men and is localized in duodenum. Neuroendocrine tumor, ganglioneuroma, paraganglioma, and schwannoma are the differential diagnoses. Histologic diagnosis is difficult to make. Immunohistochemistry is indispensable for the diagnosis. Often than not, we cannot make diagnosis by a simple biopsy, because the tumor has a submucosal location and three types of cells need to be present for diagnosis: spindle cells, ganglion cells, and epithelial cells. That's why en-bloc resection of the tumor is frequently needed for accurate diagnosis. Endoscopic resection or laparoscopy is used depending on the characteristics of the tumor. In our case, the 5 cm peri-ampullary tumor warranted a laparoscopic resection. Generally, there is no role for adjuvant therapy in duodenal gangliocytic paraganglioma (DGP). But sometimes, radiotherapy, surgery, somatostatin analog are used for treatment depending on the features of DGP. There is no consensus on follow-up management, but experts agree on the necessity of frequent follow-ups.

Conclusion: We believe it is important to include gangliocytic paraganglioma (GP) as a differential diagnosis in patients who present with duodenal masses, weight loss, and general state alteration.

Keywords: Benign tumor, Gangliocytic paraganglioma, Neuroendocrine tumor

How to cite this article

Bitar HE, Rakka M, Chbat M, Ibrahim R, Matar B, Karaa A. Duodenal gangliocytic paraganglioma: A case report. Int J Case Rep Images 2023;14(1):123–128.

Article ID: 101396Z01HB2023

****

doi: 10.5348/101396Z01HB2023CR

Houssam Eddine Bitar[1], Mohamad Rakka[2], Maureen Chbat[3], Rim Ibrahim[4], Bassam Matar[5], Aziz Karaa[6]

Affiliations: [1]MD, Surgery Department, Al-Zahraa Hospital UMC, Beirut, Lebanon; [2]MD, Surgery Department, Lebanese University, Beirut, Lebanon; [3]MD, Hematology Oncology, Lebanese University, Beirut, Lebanon; [4]MD, Senior Director Project Management at Biorasi, LLC, Miami, Florida, USA; [5]MD, Head of Hematology Oncology Department at the Lebanese University, Beirut, Lebanon; [6]MD, Surgery Department, Longjumeau Hospital, Longjumeau, France.

Corresponding Author: Mohamad Rakka, MD, Surgery Department, Lebanese University, Beirut, Lebanon, Email: mohamad-raka@hotmail.com

Received: 10 March 2023
Accepted: 08 April 2023
Published: 29 June 2023

INTRODUCTION

Gangliocytic paraganglioma (GP) is a benign tumor, occurring in the majority of cases in the duodenum. It is considered a rare tumor, very often misdiagnosed as neuroendocrine tumor [1].

Many controversies surround this tumor, from its origin to its prevalence, diagnosis, treatment, and follow-up. A surgical resection is enough in the vast majority of cases. But awareness of this disease is important to avoid misdiagnosis and facilitate treatment.

We are reporting a case of duodenal gangliocytic paraganglioma (DGP) with clinical follow-up after a period of seven years.

CASE REPORT

This is a case of a 42-year-old man, with no significant past medical history, no food or drug allergy, who presented to our clinic for a history of general state alteration, anemia, weight loss of 10 kg over two years, on–off jaundice over two years. He was refusing all kinds of investigations until he became very ill. He had no nausea, no vomiting, no diarrhea, or constipation. On physical exam he was jaundiced, pale, afebrile, had a pulse of 60, blood pressure 125/70, respiratory rate 20. He had good bilateral air entry, and had right upper quadrant pain. Blood tests were immediately done: bilirubin slightly elevated, Hb 9 g/dL.

Then a computed tomography scan of the abdomen was done showing an intraluminal defect in D2–D3 without obstruction (Figure 1).

An endoscopic retrograde cholangiopancreatography (ERCP) was done thereafter showing a mass and we couldn't localize the papilla.

Then an endoscopic ultrasound (EUS) was done and showed a submucosal mass of 5 cm, well circumscribed abutting the ampulla, suspended by a stalk, with iso-echogenic borders, in favor of a benign tumor (Figure 2) no lymph nodes were seen.

Biopsy by EUS was taken and came back as normal epithelium, so it was inconclusive.

A laparoscopic transduodenal tumorectomy or laparoscopic duedonotomy was then scheduled. We put an endo GIA stapler on the stalk of the mass and resected the mass and its stalk entirely. We did a cholecystectomy for the gallstones due to the history of repetitive obstructive jaundice.

The post-operatory course was uneventful.

Blood tests at d1 postoperatively were as follow: Bilirubin T 1 (*N* range 0.2–1 mg/dL), Direct 0.33 (*N* range 0–0.2 mg/dL), amylase 32 (*N* range 22–80 U/L), lipase 30 (*N* range 13/60 U/L).

An upper gastrografin imaging was performed at d5 post-operatory to see if there is any leak or stenosis after the laparoscopy. It was normal, no leakage of contrast material, no strictures. At d8 post-op, the patient was discharged home.

The final pathology came back. No malignant cells in gallbladder. There were few gallstones. The 5 cm tumor and its stalk were analyzed. Margins were negative. The 5 cm mass suspended by the stalk was consistent with a gangliocytic paraganglioma. Figure 3 shows 40* hematoxylin and eosin (HE) stain: well differentiated triphasic neoplasm consists mainly of low grade neuroendocrine cells with regular nuclei (ascending arrow), schwan cells (descending arrow), and several ganglion cells (oblique arrow). No anaplasia. No mitosis seen (Ki67 less than 1%).

20* immunohistochemical (IHC) stain showing the same tumor with diffuse intense positivity for synaptophysin (descending arrow) in submucosa of unremarkable duodenal mucosa (ascending arrow) (Figure 4).

40* IHC stain showing obvious diffusely positivity for synaptophysin in all neoplastic cells (oblique arrow) (Figure 5).

The patient was followed regularly and he remains in good health for the past seven years since he had his surgery. Physical exam remains within normal limits.

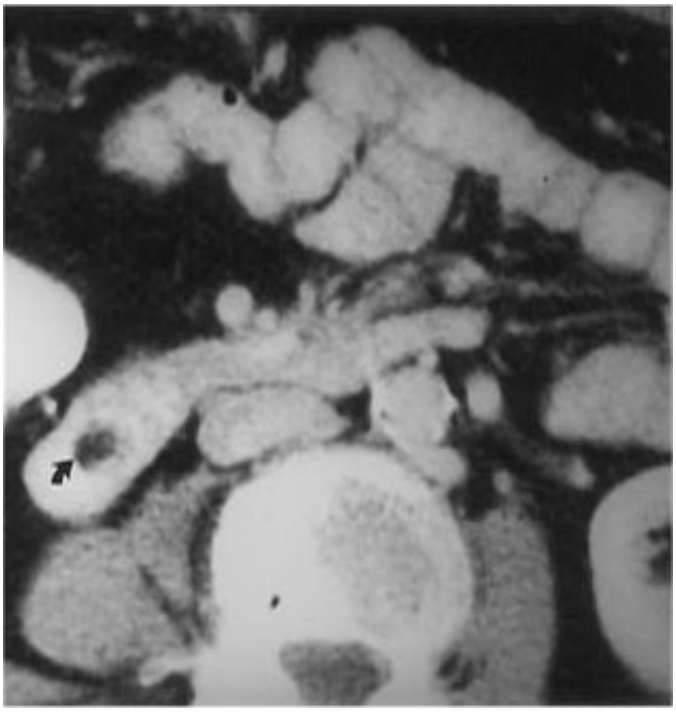

Figure 1: Computed tomography scan of the abdomen showing an intraluminal defect in D2–D3 with no obstruction.

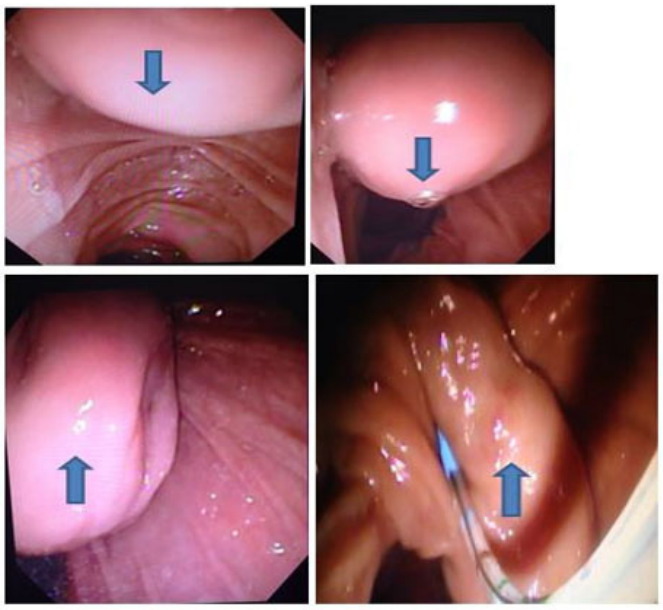

Figure 2: Endoscopic ultrasound showing a submucosal mass of 5 cm, well circumscribed abutting the ampulla, suspended by a stalk, with iso-echogenic borders.

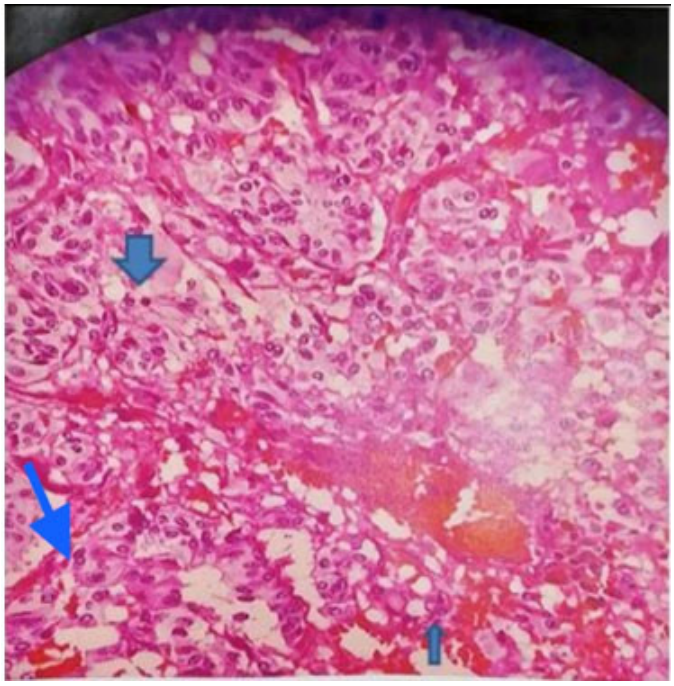

Figure 3: At 40* hematoxylin and eosin (HE) stain: well differentiated triphasic neoplasm consists mainly of low grade neuroendocrine cells with regular nuclei (ascending arrow), schwan cells (descending arrow), and several ganglion cells (oblique arrow). No anaplasia. No mitosis seen (Ki67 less than 1%).

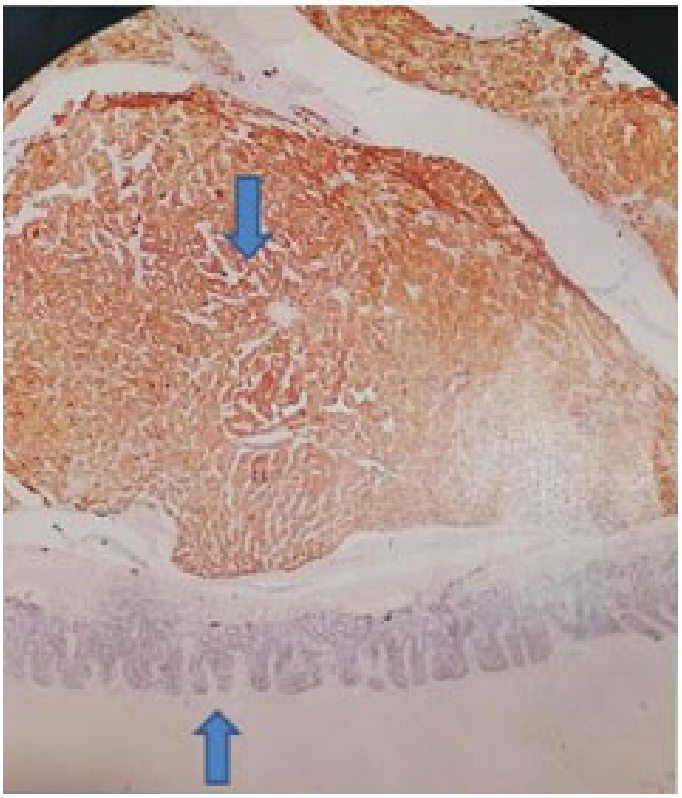

Figure 4: At 20* immunohistochemical (IHC) stain showing the same tumor with diffuse intense positivity for synaptophysin (descending arrow) in submucosa of unremarkable duodenal mucosa (ascending arrow).

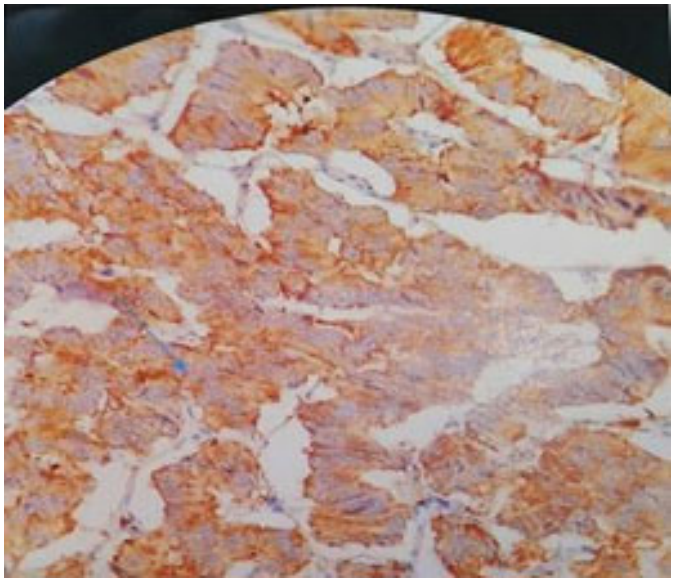

Figure 5: At 40* IHC stain showing obvious diffusely positivity for synaptophysin in all neoplastic cells (oblique arrow).

DISCUSSION

Gangliocytic paraganglioma is a benign tumor, occurring in the majority of cases in the duodenum (89% of cases). It can also occur in the respiratory tract, spinal cord, pancreas, jejunum, esophagus, and appendix [1, 2].

The first description of this tumor was in 1957 by Dahl et al. and the term gangliocytic paraganglioma was adopted in 1971 by Kepas Zacharias based on common features with paraganglioma and ganglioneuroma. Buchler et al. reported the first case of GP with positive lymph nodes [2].

The mean age at diagnosis is 54 years and males are more affected than females with a ratio of 1.5:1. The mean tumor size is 2.5 cm [3]. Although GP is a benign tumor as we saw in our case, it has a capacity to spread to lymph nodes and distant organs [2]. Some experts even name this disease “tumor with uncertain malignant potential” [4] as it led to death in 3 reported cases [1]. We note that GP from other sites of duodenum have a greater potential for metastasis [5].

Even though considered a rare tumor, we believe that DGP is an under-diagnosed tumor [6], and the incidence of positive lymph nodes is under evaluated [2, 7]. It is often misdiagnosed as neuro-endocrine tumor [8].

Its occurrence is almost always sporadic as in our case but there are some reported cases of gangliocytic paraganglioma in the context of a Von Recklinghausen disease [9].

Many theories concerning its origin exist: first some experts think that it has a hamartomatous origin [1]. Other experts say that it has and ectodermal origin. In the other hand there are experts who say that it has an endodermal origin for epithelial cells and neuroectodermal origin for ganglion and spindle cells [6, 10].

Patients present with abdominal pain or gastrointestinal (GI) bleeding and consequent anemia as in our case. The lesion can be found incidentally. It can present also with nausea, weight loss, fatigue, or jaundice [11].

Computed tomography (CT) scan of the abdomen and pelvis and endoscopic ultrasound (EUS) are done for staging and serve to R/o invasion of lymph nodes, as 47% and 64% of lymph nodes positivity are detected respectively by CT and EUS [11]. Our patient didn't have positive lymph node (LN) on scan that's why a LN dissection wasn't performed. Other types of imaging have been used to detect a DGP such as gastrografin swallow, magnetic resonance imaging (MRI), intraoperative US, positron emission tomography (PET), Octreotide scan, but they are less specific than CT and EUS for detecting LN [11].

This tumor has a submucosal location as we saw in our patient. It is a well demarcated not encapsulated solid mass [2] with an overlying mucosal ulcer. Bleeding is common even without mucosal ulceration [2]. It is most frequently polypoid pedunculated or sessile [10]. Thus, the pre-op diagnosis by EUS is difficult [3] as it can contain 1 or 2 types of cells [2] and only 19% of cases are diagnosed preoperatively [1] by fine needle aspiration (FNA) or core biopsy. We couldn't had a diagnosis preoperatively with our patient that's why we've done a laparoscopy to be able to have an en-bloc resection of the totality of the mass for accurate diagnosis.

Our patient didn't have the risk factors for lymph node positivity: young age [3], tumor invasion beyond the submucosa [10]. So we didn't do a LN dissection.

The size of the tumor and the depth of its invasion will indicate the treatment modality [10].

If the tumor is <5 cm, suspended by a stalk, and with no suspicion of positive lymph nodes, then an endoscopic resection can be conducted, although many experts argue against this technique [11]. If the tumor is peri-ampullary, >5cm, no stalk or suspicion of positive lymph nodes by imaging then a surgical resection with lymph node dissection must be conducted. In our case, the tumor was 5 cm diameter and peri-ampullary, we decided to do laparoscopic duodenotomy for a complete en bloc resection. In case of positive lymph nodes experts agree on the importance of doing a curative pancreatico-duodenectomy even though this operation carry its own risk of morbidity and mortality. Doing it otherwise will require further studies and long periods of surveillance [12].

The final diagnosis is done only after the tumor is completely resected. This tumor consists of three different cellular types: spindle cells, epithelial cells, and ganglion cells, that can exist in different proportion and we need the presence of all three types of cells to be able to make the final diagnosis of GP [3, 12].

Immunohistochemistry (IHC) is indispensable in the case spindle and ganglion cells are scarce. Immunohistochemical stain differs for each cell type. Epithelial cells are often positive for synaptophysin, cytokeratin, CD56, chromogranin, neuron-specific enolase, pancreatic polypeptide, progesterone receptor. Ganglion-like cells are positive for synaptophysin, CD56, neuron-specific enolase, somatostatin, and pancreatic polypeptide. Spindle cells are positive for S100, bcl2, CD56, neuron-specific enolase, and synaptophysin [2, 6, 8].

As for the differential diagnosis, low grade neuroendocrine tumor will not contain spindle and ganglion cells, neither progesterone receptor. But it will be positive for synaptophysin, specific-enolase protein, and S100. As for ganglioneuroma, we'll see only ganglion cells without spindle or epithelial cells. On the other hand, there is no ganglion cells in paraganglioma. Finally, in schwannoma we'll see only spindle cells [1, 3, 6].

It's imperative to get an accurate diagnosis because the clinical course, prognosis, follow-up and treatment will differ depending on the type of tumor.

There is no role for adjuvant chemotherapy (cyclophosphamide vincristine dacarbazine) [12] and radiotherapy (RT) for DGP even in the metastatic cases [4]. But RT can be used if the patient is unfit for pancreatico-duodenectomy [13]. In fact, a study done by Van et al. showed that 14 sessions of RT can control bleeding in a patient having a peri-ampullary GP with multiple comorbidities who is unfit for surgery.

The majority of tumors are endocrinologically inactive. Some secrete somatostatin and pancreatic polypeptide [2]. These levels, if elevated, can be used to monitor recurrence or metastasis after a surgical resection [3]. Katayama et al. reported a case of DGP with catecholamine secretion during the surgery, revealing that pheochromocytoma could be in the differential diagnosis [14].

Studies showed that there could be a role for somatostatin analogs in the treatment of DGP [12] specially in patients who are unfit for surgery or those who still have positive margins after surgery.

Concerning the follow-up of this disease there are no consensus on how and when to do imaging studies, but the experts agree on the necessity of a more frequent follow-up including clinical and imaging studies, in the cases where there are lymph nodes involvement [12].

CONCLUSION

Duodenal gangliocytic paraganglioma is a rare tumor most frequently benign located in the duodenum, arise in mid-age man. Surgery is needed in the case the tumor is more than 5 cm, peri-ampullary, and also in the rare cases of lymph node positivity. A good physical exam, complete resection, IHC, and surveillance are key in this disease.

REFERENCES

1. Okubo Y, Yoshioka E, Suzuki M, et al. Diagnosis, pathological findings, and clinical management of gangliocytic paraganglioma: A systematic review. Front Oncol 2018;8:291.
2. Park HK, Han HS. Duodenal gangliocytic paraganglioma with lymph node metastasis. Arch Pathol Lab Med 2016;140(1):94–8.
3. Loftus TJ, Kresak JL, Gonzalo DH, Sarosi GA Jr, Behrns KE. Duodenal gangliocytic paraganglioma: A case report and literature review. Int J Surg Case Rep 2015;8C:5–8.
4. Li B, Li Y, Tian XY, Luo BN, Li Z. Malignant gangliocytic paraganglioma of the duodenum with distant metastases and a lethal course. World J Gastroenterol 2014;20(41):15454–61.
5. Henry C, Ghalel-Méchaoui H, Bottero N, Pradier T, Moindrot H. Gangliocytic paraganglioma of the pancreas with bone metastasis. [Article in French]. Ann Chir 2003;128(5):336–8.
6. Okubo Y, Nemoto T, Wakayama M, et al. Gangliocytic paraganglioma: A multi-institutional retrospective study in Japan. BMC Cancer 2015;15:269.
7. Witkiewicz A, Galler A, Yeo CJ, Gross SD. Gangliocytic paraganglioma: Case report and review of the literature. J Gastrointest Surg 2007;11(10):1351–4.
8. Adams L, Friedman TM, Shaver TR, Younan G. Duodenal gangliocytic paraganglioma requiring a pancreaticoduodenectomy: A case report and review of the literature. Case Rep Surg 2018;2018:6292789.
9. Relles D, Baek J, Witkiewicz A, Yeo CJ. Periampullary and duodenal neoplasms in neurofibromatosis type 1: Two cases and an updated 20-year review of the literature yielding 76 cases. J Gastrointest Surg 2010;14(6):1052–61.
10. Bakshi N, Rao S, Dhawan S, Singla V. Duodenal gangliocytic paraganglioma: A rare cause for gastrointestinal polyp. Indian J Pathol Microbiol 2018;61(4):580–2.
11. Hartford L, Sherazadishvili A, Leslie K. Duodenal gangliocytic paraganglioma with lymph node metastasis: A systematic case review. Int J Hepatobiliary Pancreat Dis 2017;7:53–61.
12. Cathcart SJ, Sasson AR, Kozel JA, Oliveto JM, Ly QP. Duodenal gangliocytic paraganglioma with lymph node metastases: A case report and comparative review of 31 cases. World J Clin Cases 2017;5(6):222–33.
13. Van J, Esteban MJ, Oikonomou I. 1979 GI bleed secondary to gangliocytic paraganglioma treated with radiation monotherapy. The American College of Gastroenterology 2019;114:S1105–6.
14. Katayama T, Iwasaki E, Minami K, Fukuhara S, Kanai T. Transient hypotension during endoscopic resection of gangliocytic paraganglioma with ampullary involvement. VideoGIE 2019;5(1):26–8.

****

Author Contributions

Houssam Eddine Bitar – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Mohamad Rakka – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Maureen Chbat – Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Rim Ibrahim – Acquisition of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Bassam Matar – Conception of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Aziz Karaa – Conception of the work, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement

Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

Copyright