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Case report

Duodenal teratoma: Rare case of extragonadal germ cell tumors and review of literature

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ABSTRACT

Introduction: Duodenal teratoma is a rare condition with only four cases reported in the English literature. Radiological imaging and tissue sample are necessary for diagnosis in addition to tumor markers. The most effective treatment is still complete excision with safety margins.

Case presentation: We report a case of 26 years-old-man, in whom epigastric pain, decreased appetite, and postprandial bilious vomiting had been prevalent for 5–6 months and had exacerbated prior to the emergency room.

Enhanced abdominal computed tomography revealed a 10 × 15cm heterogeneous solid mass with cystic component in the third duodenum segment. The inferior veina cava and aorta were both compressed, although there was no sign of lymphadenopathy or ascites. An ulcerating non-bleeding lesion at the D2-D3 junction of the duodenum was discovered during a gastroduodenoscopy. Biopsies and immunohistochemical investigations revealed findings that were consistent with a mixed non-seminomatous germ cell tumor. A PET-CT scan was performed, which revealed FDG uptake by the duodenal lesion but no evidence of metastatic lesions.

A distal duodenal segmentectomy is performed, and then a duodeno-jejunal anastomosis is used to restore continuity. The final diagnosis was teratomatous tumor of the duodenum without malignant changes.

Conclusion: This is the second adult case of main duodenal teratoma that has been reported. We publish it to encourage surgeons to think about this differential diagnosis and carefully plan surgery using a multidisciplinary approach.

1. Introduction

Germ cell tumors (GCTs) are neoplasms that often originate from germ cells of the gonads; but in 5% of cases they have extragonadic origins, and are then named Extragonadal Germ cell tumors (EGGCTs). They are found in kids and young adults and they originate in different anatomical locations, some of which are rare sites [1] as is the case that we are going to present.

Teratoma is one of the morphological types of EGGCTs; [1] it is made up of tissues originating from multiple germ layers, and it is divided into categories according to its degree of maturity [2]. Its embryonic origin is debatable and several concepts have been developed from 1908 to nowadays, but they can be assumed to be congenital pathologies [3].

Extragonadic teratomas are rare, and those which are rising from duodenum are among the rarest [2]. Herein, we present the fifth case of primary duodenal teratoma and the second case occurring in an adult, this is a case of 26-years-old male, who has been diagnosed with mature duodenal teratoma as a cause of abdominal pain and anemia, and treated by an oncologic resection.

This case is reported in line with the SCARE criteria [4].

2. Case presentation

A 26-years-old male with no medical or surgical history, presented to the emergency department for epigastric pain, decreased appetite and postprandial bilious vomiting that had lasted for 5–6 months and worsened. He also stated that his stools turned black, and denied the presence of fever or other symptoms.

Upon presentation, he was hemodynamically stable with moderate tachycardia of 104 beats/min, and he was afebrile. On physical

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examination, he looks exhausted and pale, but without jaundice. His abdomen was not distended, soft and tender in the epigastric and left hypochondrium. A mass is palpated at the epigastric level. The patient's laboratory studies showed anemia (Hemoglobin 7.5 g/dL), associated with elevated creatinine level (Creatinine 1.55 mg/dL); the remaining test results were all within the normal range. Enhanced abdominal computed tomography was consistent with a 10 × 15cm heterogeneous solid mass with cystic component in the third duodenal portion. Radiology also noted compression of inferior veina cava and aorta, but without evidence of lymphadenopathy or ascites (Fig. 1).

The patient was admitted, well resuscitated, and a gastroduodenoscopy has been performed which has shown an ulcerating non bleeding lesion at the D2-D3 junction of the duodenum. The biopsies and immunohistochemistry studies showed the presence of findings which are consistent with mixed non-seminomatous germ cell tumor. A PET-CT is made, and showed a FDG uptake by the duodenal lesion without demonstration of metastatic lesions (Fig. 2A–B). Testicular ultrasound and testicular cancer tumor markers (Alfa-feto protein, Beta-HCG and LDH) were insignificant and within normal range. He was prepared for surgery after multidisciplinary team planning.

A midline laparotomy was done. Abdominal exploration showed the depicted duodenal mass (Fig. 3), and did not show the presence of peritoneal carcinomatosis or liver lesions. Distal duodenal segmentectomy is performed, taking care not to breach the nearby compressed vascular structure, followed by restoration of continuity with a duodeno-jejunal anastomosis (Figs. 4–5).

The patient tolerated the surgery well and was discharged home on the tenth day after a simple stay without complications following surgery.

The pathology report revealed a 14 cm small intestine polypoid intraluminal mass, with a large pedicle; which shows surface erosion with inflammatory changes. It is composed of a mixture of mature epithelial and somatic elements; various types of ducts and cysts showing epidermoid, columnar, mucinous and endocrine type, with some atypia and rare mitosis. The stroma around is formed of fibrous, cartilaginous and neuroid elements. The proximal and distal margins are free. The final diagnosis was teratomatous tumor of the small bowel without malignant changes.

The patient presented to the outpatient clinic two weeks following the operation. He doesn't complain, eats well, and maintains a normal bowel habits. His abdomen is soft with a nice scar. A follow-up meeting is scheduled after 3 months, with a request for abdominal imaging and a blood test.

3. Discussion

GCTs are tumors that derive from neoplastic germ cells, and they have their extragonadal counterpart; they are morphologically similar [1]. GCTs usually originate in the gonads, whereas EGCTs are found in different places, the majority of which are found at the midline level of the body; as if the germ cell precursors had mistakenly stopped there as they migrated to their final position during embryogenesis [7], via dorsal mesentery of the hindgut from endoderm of yolk sac to gonads; [5] several extragonadic locations are reported from which we mention head and neck, brain, mediastinum, abdominal wall, retroperitoneum and sacrococcygeal region [2].

There are some distributions and subdivisions of these tumors, which vary between benign and malignant types. The two essential subdivisions are germinomatous and non-germinomatous; the latter is the commonest and is likewise divided into some subtypes including embryonal carcinoma, yolk sac carcinoma, choriocarcinoma, mixed and teratoma [6].

Teratomas are therefore neoplasms formed by a mixture of dermal tissue of two or three origins [11], deriving from the three germ layers: the ectoderm, the endoderm and the mesoderm, and with different level of maturity and differentiation [2]. Only 1% will undergo a malignant transformation mostly in squamous cell carcinoma [2], they risk having an aggressive and metastatic pattern [1].

They can be found at any age, with essentially two peaks of incidence, at the age of two and early adulthood [2], and they can be found

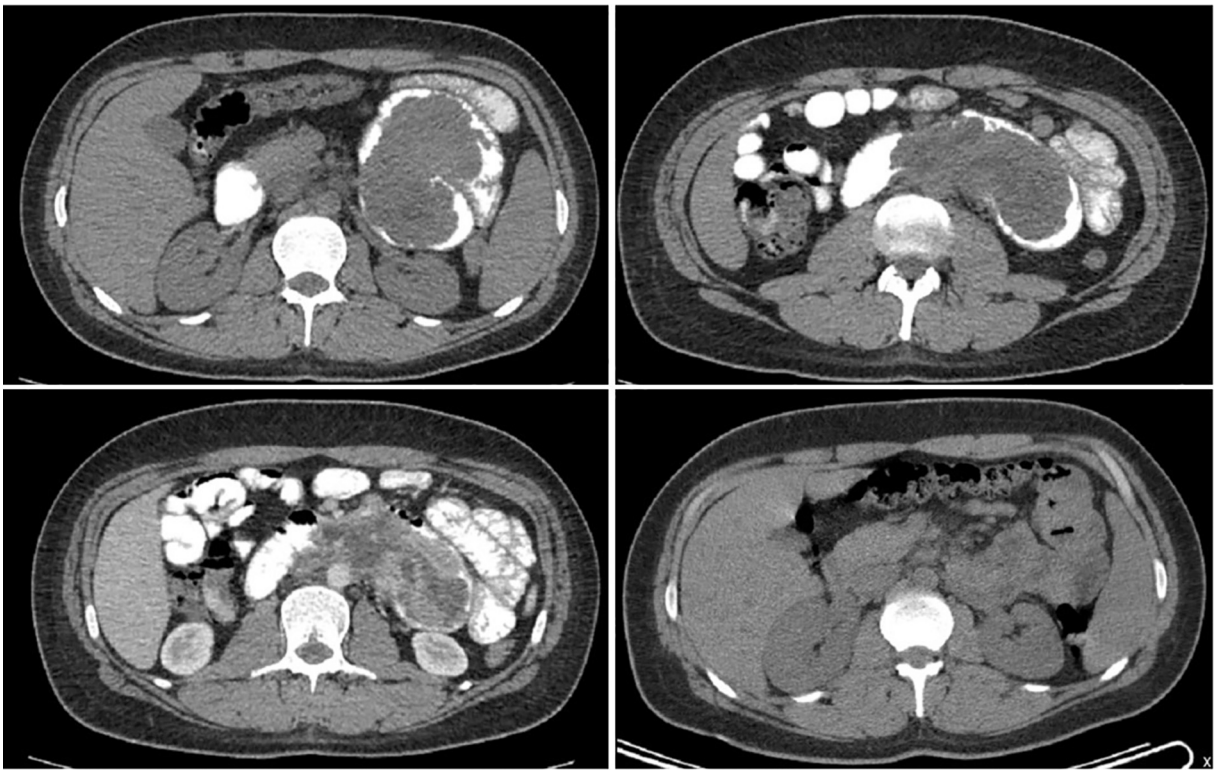

Fig. 1. Enhanced abdominal computed tomography.

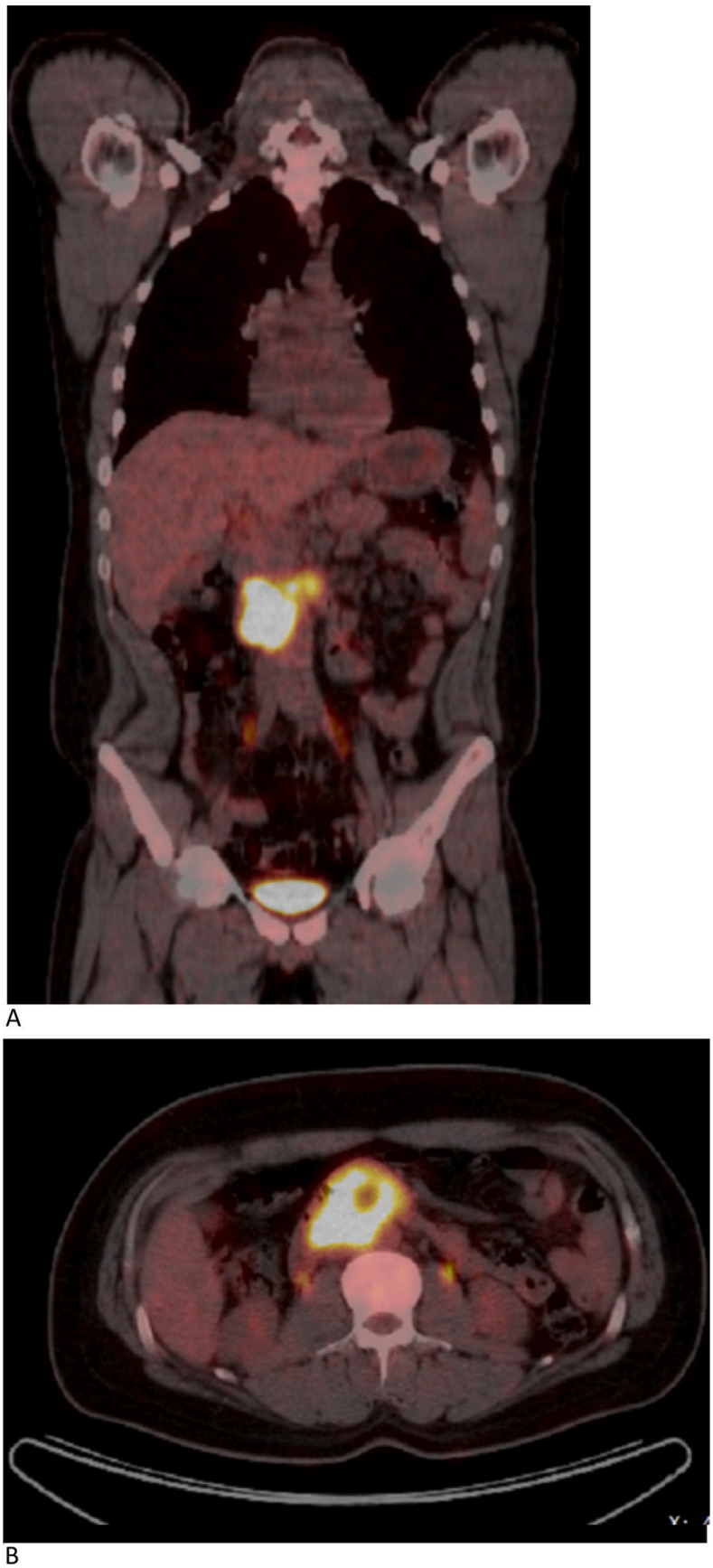

Fig. 2. A PET-CT with FDG uptake.
B PET-CT with FDG uptake.

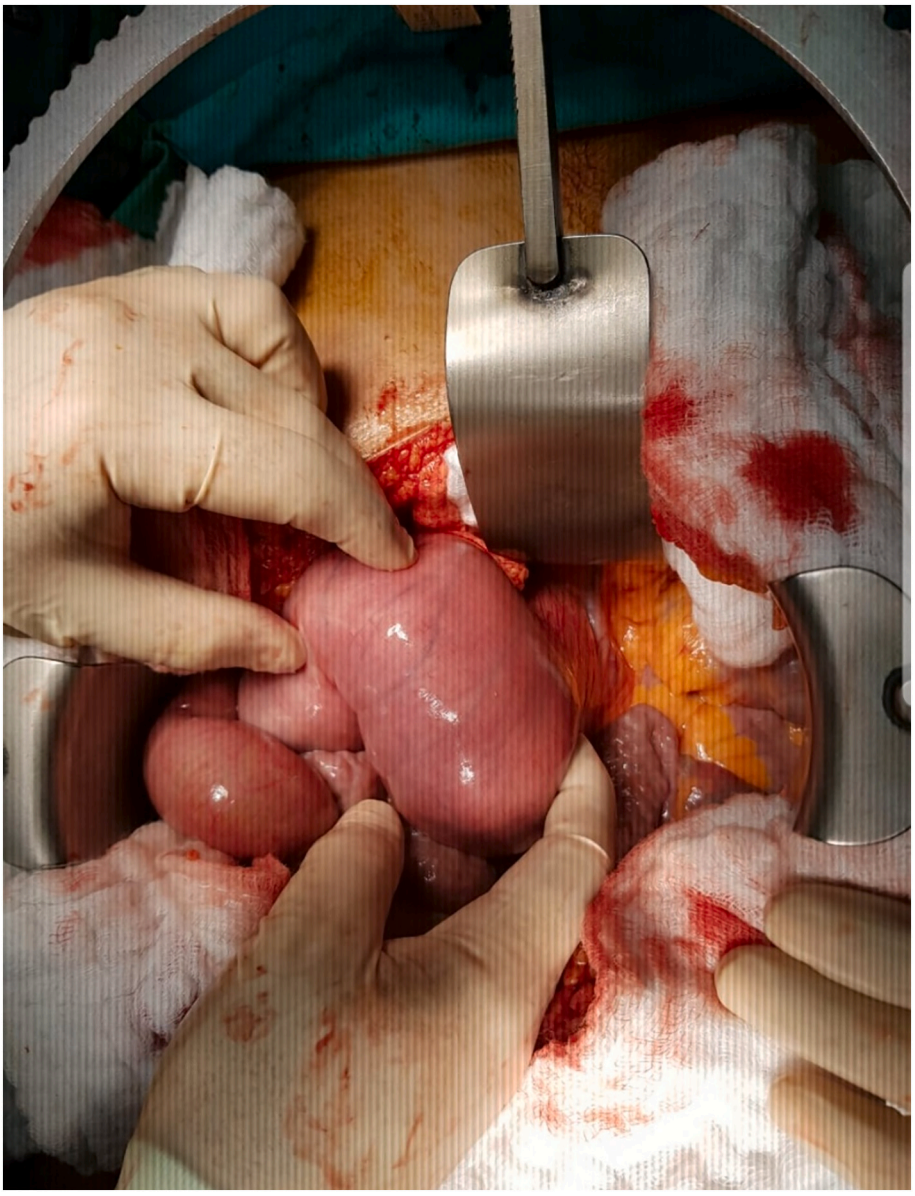

Fig. 3. Intraoperative photo of the duodenal mass.

in different places, but with varying frequencies; foregut teratoma are quite rare, of which the rarest are those found at the level of the duodenum [3].

The clinical presentation of teratomas is related to their size, location and mechanical compression exerted by them [8], while the physical examination is not specific, yet it may involve a palpable cystic-solid mobile mass with [5]. Note that sometimes they present as hemorrhagic mass [3], associated with iron deficiency anemia [8].

The initial step of the investigations is to identify whether it is an extragonadal primary tumor or if it is a secondary to a coexisted primary gonadal pathology [9] The diagnosis is based on the histological analysis, in addition to tumor markers (Alfa feto protein, Lactate Dehydrogenase and Beta Human Chorionic Gonadotropin), may be essential, especially when high levels of human chorionic gonadotropin are found; [1] at this point, its value will be important during follow-up for early detection of recurrence or malignant transformation [3], but none of them is a teratoma's specific tumor marker [9].

Imaging methods retain an important role in the diagnosis and characterization of teratomas, and they play an essential role in the planning and guide of the subsequent operative management. Certain pathognomic radiological features are found, but they are often nonspecific. A simple x-ray can show a mass of soft tissue surrounded by calcification of the edge. Ultrasound can show the mass and its echogenicity which can be complex. Computed tomography usually reveals a cystic mass with varying attenuations consistent with the combination of different tissues and it also allows a better illustration of anatomy and evaluation of loco-regional and remote invasion. Magnetic resonance imaging tends to be superior to Computed tomography in assessing the constituents of the mass and providing more detailed imaging of the tissue. Positron emission tomography can help identify malignant teratomas [10].

Teratoma exhibit distinct macroscopic features, including an encapsulated mass, with fleshy and cystic portions that may contain mucoid, keratin debris, serous, and/or hard component such as bone or teeth. Microscopically, they are composed of many types of somatic tissues arranged in a chaotic architecture, with varying degrees of maturation, allowing mature teratomas to be distinguished from juvenile teratoma. Neuroectodermal tissue is the most prevalent type of tissue seen in the immature form [7].

The main therapy for a duodenal teratoma is total surgical excision

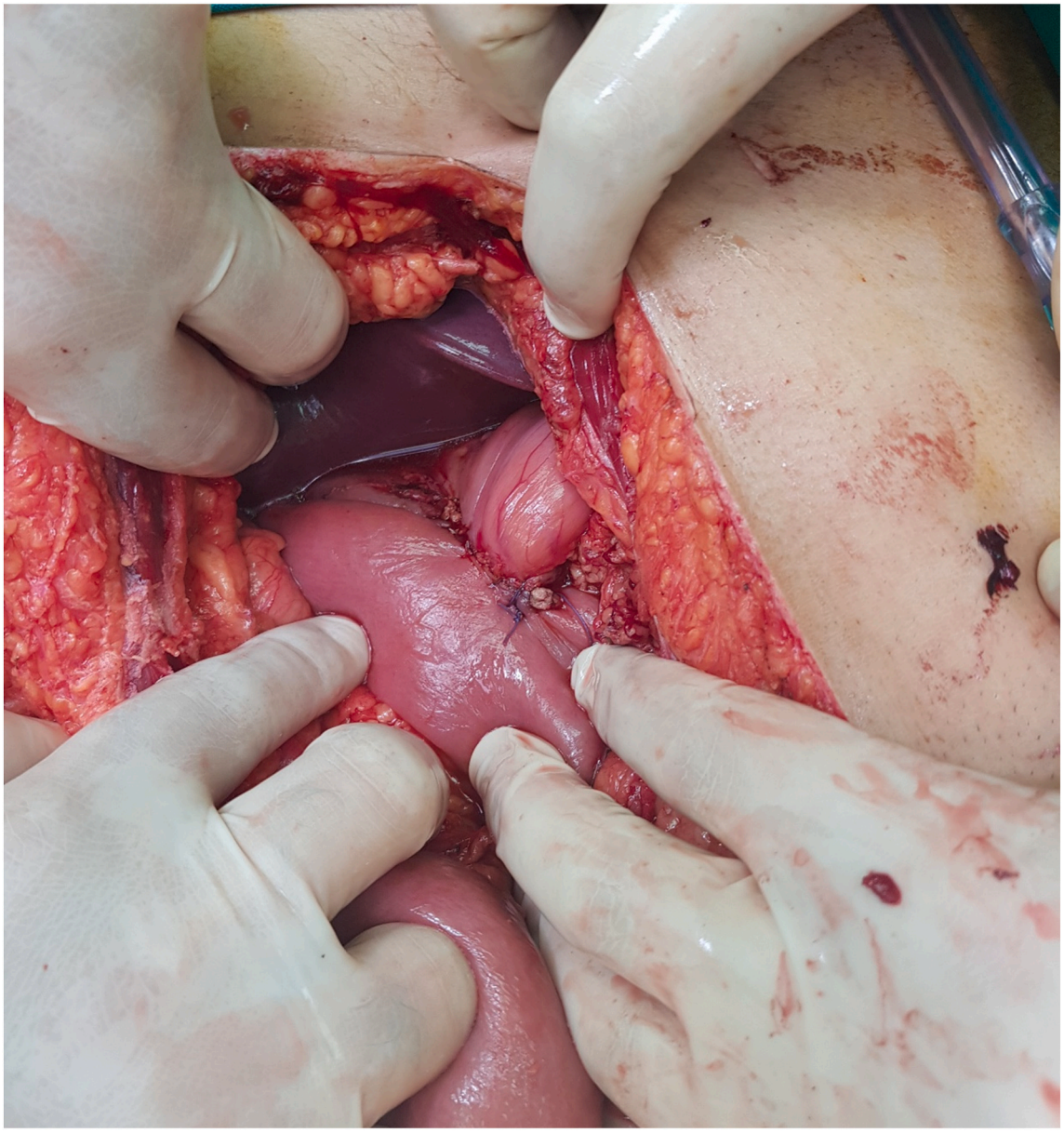

Fig. 4. Intraoperative photo of the duodeno-jejunal anastomosis.

with safe margins; however, because of its close proximity to the pancreas and if the tumor is abutting the ampulla of vater, reconstructive surgical operations such as pancreaticoduodenectomy may be required [10]. Mature teratomas have a good prognosis and can be removed surgically with clear margins. In the case of immature malignant variants, however, chemotherapy based on cisplatin, etoposide, and bleomycin must be added to the treatment plan [10].

Duodenal teratoma is an extremely rare pathology, of which only four cases have been published in the English literature, and only one case in an adult (Table 1). Then the case presented is the 5th in history and the second in an adult.

4. Conclusion

Duodenal teratoma is an uncommon disorder that has only been reported four times in the English literature. In addition to tumor markers, radiological imaging and tissue biopsy are required for diagnosis. Complete excision with safety margins is still the most effective procedure.

This is the fifth case of primary duodenal teratoma to be reported, and the second in adults. We publish it to encourage surgeons to consider this differential diagnosis and to plan surgery carefully using a multidisciplinary approach.

Ethical approval

The study type is exempt from ethical approval.

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CRediT authorship contribution statement

All authors were involved with the design, drafting, revision, and final approval of this case.

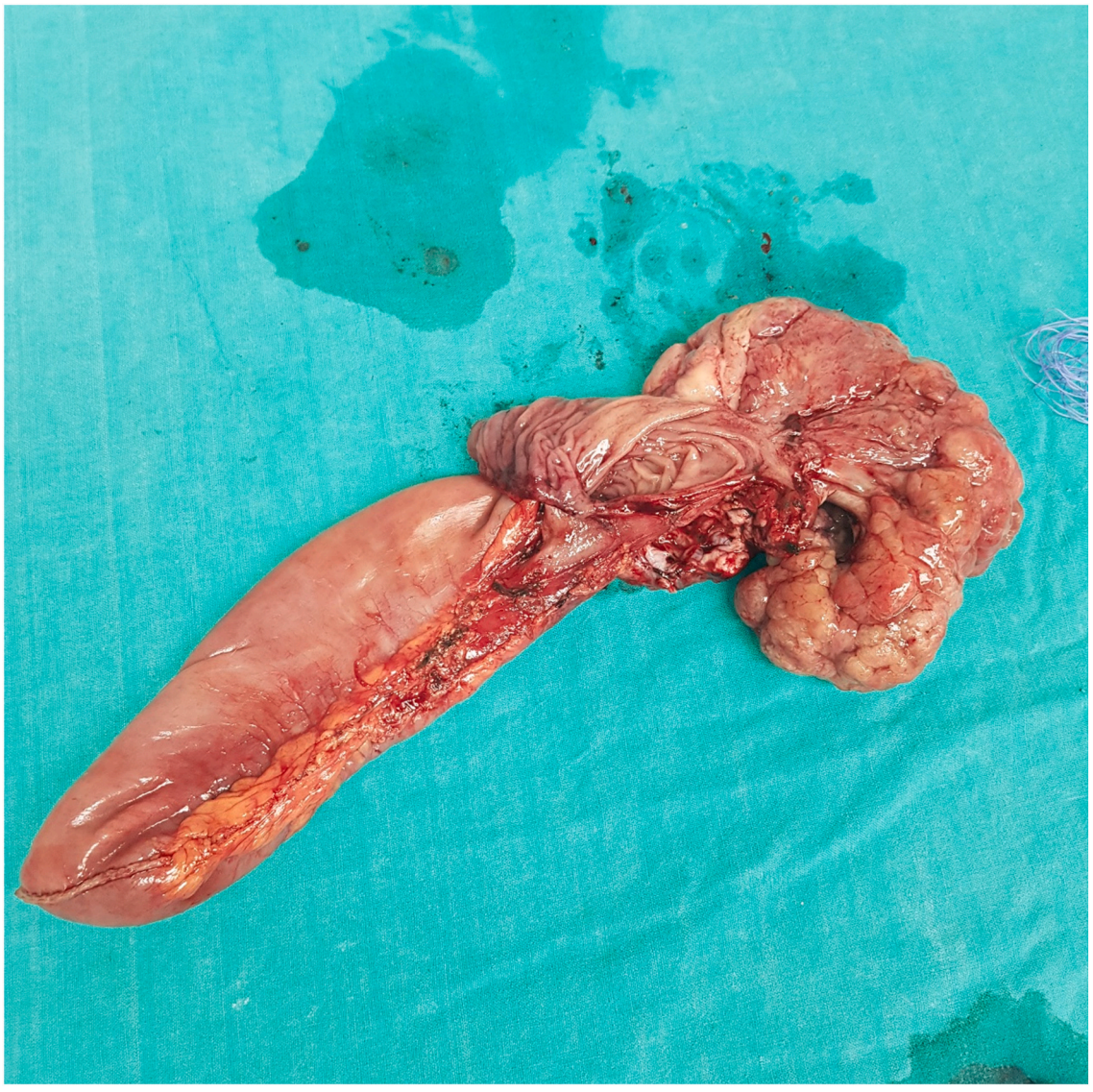

Fig. 5. Intraoperative photo of the excised segment of duodenum.

Table 1
Reported duodenal teratoma in English Literature.

Features	Case 1	Case 2	Case 3	Case 4
Authors	Joshi et al.	Chaudhari et al.	Puri et al.	Chansoon et al.
Year	2014	2015	2018	2020
Age	7 days	7 days	2 years	31 years
Ml	Female	Female	Female	Male
Presentation	Bowel obstruction and dehydration	Bowel obstruction	Severe anemia, generalized body swelling and incidentally detected mass	Partial bowl obstruction
Physical exam/ associated findings	Exomphalos	Exomphalos	Well circumscribed, firm mobile mass in right hypochondrium	Palpable ill-defined mass on the left side of the abdomen
Diagnostic imaging	No detected mass	Detected mass	Complex bilobed solid-cystic mass	Multiseptated cystic tumor at the retroperitoneum
Treatment	Urgent laparotomy – complete excision	Urgent laparotomy – complete excision	Elective laparotomy – complete excision	Two stage surgery, first gastrojejunostomy; second complete excision
Finding	Solid-cystic mass in the second part of the duodenum	Hard cystic mass attached to duodenum	Large intraluminal mass in duodenum	Solid cystic mass originating from the duodenum
Tumor markers	Normal	Normal	Not available	Not available
Reference	[3]	[11]	[5]	[2]

Guarantor

Dr Mostapha Mneimneh

Informed 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.

Declaration of competing interest

This article has no conflict of interest with any parties.

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