INFLAMMATORY MYOFIBROBLASTIC TUMOR MIMICKING GASTRIC DUPLICATION CYST: A DILEMMA IN CLINICAL AND RADIOLOGICAL DIAGNOSIS

Abstract:
Gastric duplication cyst is a rare congenital malformation of the foregut but inflammatory myofibroblastictumor (IMT) a more rare diagnosis.These are mostly asymptomatic diagnosed incidentally on imaging done for some other cause. It is very difficult to differentiate between these two conditions based on clinical and radiological studies done preoperatively. Endoscopic Ultrasound with Fine Needle Aspiration (EUS-FNA) gives us opportunity to correctly diagnose these conditons. However EUS-FNA carries a high risk of iatrogenic rupture of these lesions with significant infective potential. We present a case of IMT presenting as Gastric duplication cyst with brief review of literature.

1.Introduction:

Inflammatory myofibroblastictumors (IMT) are a very rare type of tumors comprising of myofibroblastic spindle cells and inflammaory cells. It was first observed in lungs by Bunn in 1939 . Pathogenesis of this IMT has been postulated to range from infective, reactive to a more recent belief of neoplastic. Recent change in the same is evidenced by recurrence, metastasis and cytogenetic studies doen in this field[1][2]. Definitive management still remains a mystery but surgical excision remains the treatment of choice.Gastrointestinal duplication cyst are rare congenital malformations which can occur anywhere in the gut. Foregut duplication cyst can be further divided into types based on the embryonic origin[3]. Treatment often involves surgical removal to prevent complications. Gastric duplication cyst makes up for 4 - 9% of all intestinal duplication cyst[4] Gastric Duplication cyst can be asymptomatic or can present as epigastric pain, vomiting, gastric outlet obstruction, weight loss or mass per abdomen[5] .Both of the conditions are diagnosed based on radiological diagnosis with Endoscopic Ultrasound and Computed Tomography (CT) scan. Definitive diagnosis is obtained with tissue diagnostic tests like fine needle aspiration biopsy and histopathological examination (HPE) of surgical specimen. In present case HPE was in favour of tumour consisting spindle cells like GIST and myofibroblastic tumourThis created a dilemma for definitive diagnosis in the preoperative period with contrasting findings in postoperative diagnosis. This report signifies the need for considering alternative diagnosis mentioned above in case a tissue diagnosis is not available for preoperative diagnosis and its management.

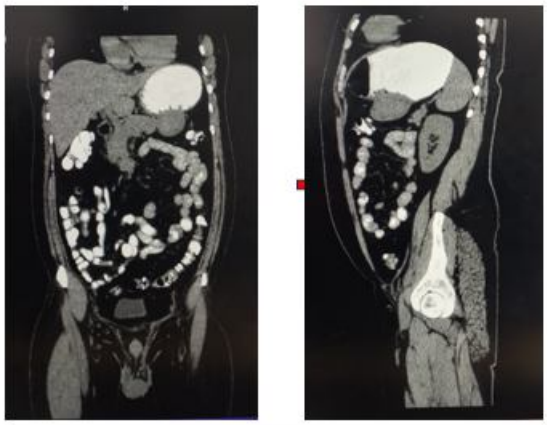
2.Case report

A 37 year old gentlemen came presented with complaints of epigastric and supraumbilical pain since 3 months insidious onset,dull aching , mild to moderate in intensity,

nonprogressive with no specific aggravating or relieving factors. The pain was non radiating and non referring in nature. He also complained of nausea for 2-3 months. There was no history of fever, retrosternal pain, vomiting.The patient had no associated systemic symptoms. Past history & family history was unremarkable. The patient denied a history of any addictions. His bowel and bladder habits were unaltered.

Clinical examination showed hemodynamically stable with no systemic signs seen. Abdominal examination was unremarkable with no tenderness, guarding or palpable abdominal lump. Contrast enhanced Computed Tomography(CECT) scan revealeda well-definedisodense lesion in the perigastric region closely abutting the posterior gastric wall and body of the pancreas (Fig. 1). The lesion was 6.1x3.9x3.6cms and showed thin peripheral postcontrast enhancement. There was no solid component noted. Possibility of malignant potential was low in the scan. His upper Gastrointestinal endoscopy showed no mucosal abnormality. Endoscopic Ultrasonography (EUS) was suggestive of small cystic lesions 3.5cm x 4.5cm arising from layer 4 of stomach at the level of cardiac end from posterior wall (Fig. 2). It showed heterogeneous contents and septations. Biopsy was attempted but yield was negative.Impression most likely of a gastric duplication cyst was given.

Fig. 1. Lesion in the perigastric region closely abutting the posterior gastric wall and body of the pancreas



a) Coronal view b) Sagittal view

Figure 2 Stomach at the level of cardiac end from posterior wall

Considering the inability to acquire tissue diagnosis the decision to do a laparoscopic excision of the cyst was taken. Nathanson retractor was inserted through the epigastric port and liver was retracted away from the operative site. The cyst was found to be ruptured and present in the lesser sac continuous with the posterior wall of the stomach (Fig.3). The cyst was excised in toto and separated from the gastric wall leaving a rent in the muscularis layer of the gastric wall.The cyst was placed in an endobag and sent for histopathology (Fig. 4). The opening in muscularis was closed with polydiaxone 3-0 simple interrupted sutures.

Intraoperatively upper Gastrointestinal endoscopy was done to rule out mucosal breach and to check the adequacy of the repair. Stomach was insufflated with air and leakage of gastric repair was tested from the laparoscope by keeping the suture line dipped in normal saline to look for any bubbling (Fig. 5). Repair was found to be adequate. An abdominal drain was placed in the lesser sac.Check dressing was done on day 2. The drain was removed on day 5. Liquid diet was started on day 3 and shifted to full diet on day 5.

Histopathology report was suggestive of spindle cell tumor most likely a GIST or myofibroblastictumor. IHC reporting gave us the definitive diagnosis of inflammatory myofibroblastictumor.Follow up of 6 months has shown him to be disease & symptom free.

Figure 3 lesser sac continuous with the

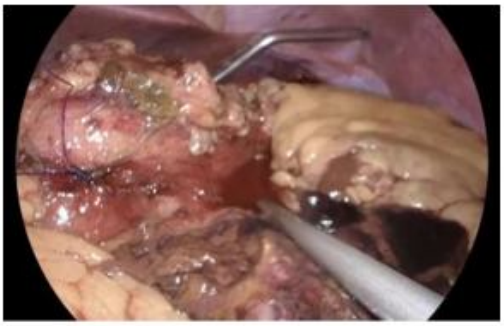


Figure 4 cyst placed in an endobag

Figure 5 Line dipped in normal saline to look for any bubbling

3.Discussion

Gastric duplication cyst, Gastrointestinal stromal tumors and Inflammatory myofibroblastictumors. Are rare tumors of the Gastrointestinal tract. Present case highlights this varied differential diagnosis based on preoperative evaluation and post operative histopathological reports.Gastric duplication cyst generally present in children and young adults and rarely in forth decade of life like the IMT[3][6]. Duplication cysts are thought to arise due to a developmental anomaly during the embryonic stage of growth.Endoscopic ultrasound is the investigation of choice. Typically seen as a anechoic to isoechoic cystic lesion arising from layer 3-5 of stomach usually arising from the muscularis propia[5][7].Lungs and gastrointestinal tract are the most common tissue of origin in either of the swellings and IMT rarely occur in the oral cavity and maxillofacial areas [8].While

some authors suggest that EUS-FNA is necessary to rule out other conditions and malignant transformation within the cyst whilst ensuring a definitive diagnosis.IMT might require multiple biopsies to make a definitive diagnosis Role of fine needle aspiration is controversial as it known risk of rupture and can result in dissemination in case of a malignancy. In present case, the cyst ruptured due to attempted EUS guided FNA.

Surgical excision remains the treatment of choice in such cases. This may result in a rent in the gastric wall which should be closed with sutures. It is imperative to rule out leak after gastric wall repair. This can be done with simultaneousintraoperative upper gastrointestinal endoscopy and laparoscopy. Air insufflated through endoscope and dipping the suture line in normal saline laparoscopically is a useful test. Leak can be detectedwith evidence of any bubbling present which is seen laparoscopically which is many a times done after Sleeve Gastrectomy also in our centre.

Both IMT & duplication cyst present similarly, however, final diagnosis is confirmed on histopathology and immunohistochemistry after surgical excision.Present case highlights the importance of the same. IMT have a spindle cell morphology but they lack the cellular atypia of sarcomas.They are typically reactive to vimentin and Smooth Muscle actin on IHC[9]whereas duplication cyst mimic the histopathological appearance of organ of origin.

4.Conclusion

This report brings forth a case of cystic lesion arising from the stomach where the patient presented to us with complaints of epigastric pain with no positive examination findings. Preoperatively cystic lesions of the stomach can be confused as duplication cyst like in present case. In such cases malignant potential is low as compared to GIST or myofibroblastictumors and other cystic lesions. Surgical management is indicated in symptomatic gastric cyst. However, a GIST or myofibroblastictumors has to be surgically excised even if it is asymptomatic. EUS guided Fine needle aspiration biopsy is controversial in such cases as evidence suggest significant risk of rupture and infective potential of the procedure. Combination of intraoperative endoscopy along with laparoscopic leak test helps in preventing complications.

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