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Case Study

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A CASE REPORT OF INCIDENTAL LEIOMYOMA OF GE JUNCTION IN A CASE OF CHRONIC CHOLECYSTITIS

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ABSTRACT

Esophageal leiomyomas are rare tumors. They may be symptomatic or an incidental finding. Leiomyomas of gastroesophageal junction may mimic Gastrointestinal stromal tumors (GIST). Surgical excision followed by histological analysis gives the final diagnosis. We present a case of incidental esophageal leiomyoma which was diagnosed as GIST on imaging done for cholecystitis. A brief case report with review of literature is presented.

KEYWORDS: Case report, incidental, gastroesophageal junction, leiomyoma.

INTRODUCTION

Mesenchymal tumors of the gastrointestinal tract are seen less often than epithelial tumors. True leiomyomas of the muscularis propria are common in the esophagus and rare in the gastric body and antrum. About one-third of these neoplasms are located at the gastroesophageal junction and are often asymptomatic or minimally symptomatic.[1] Microscopically, they show hyper proliferation of smooth muscle cell well encapsulated by surrounding tissue or connective tissue capsule.

Leiomyomas are usually detected because of clinical symptoms that range from gastrointestinal bleeding to abdominal pain, dysphagia and palpable abdominal masses. However, they can be detected incidentally intraoperatively or at autopsy. Most of these incidental lesions are usually smaller hence being asymptomatic.

In 1981, Takubo et al[2] coined the term ‘‘seedling’’ leiomyomas to describe small (<7 mm) esophageal leiomyomas found incidentally during surgery or at autopsy, because of their resemblance to millet seeds. Among 342 patients, nearly 8% had leiomyomas, most of which clustered around the region of the esophagogastric junction (EGJ). An even higher frequency of leiomyomas (22%) was reported recently by Agaimy and Wunsch[3] in a study of 77 patients undergoing surgical resections for distal esophageal carcinomas. Diagnosis can be achieved with help of a barium swallow, esophagoscopy, computerized tomography (CT) scan or endoscopic ultrasound (EUS) Enucleation of leiomyoma is safe and effective procedure but whether to operate every patient of leiomyoma remains a controversy.

We present a case of obstructive jaundice secondary to CBD stones with incidental GE junction leiomyoma at a tertiary referral centre.

CASE REPORT

A 49 years old male presented in the outpatient department with pain in abdomen since10 days which was dull aching in the right hypochondrium and epigastric region. The patient had past history of obstructive jaundice and severe pain in abdomen in the right hypochondrium region 3 months back which on further investigations was found to be due to impacted CBD stones and multiple gall stones and cholecystitis. He had undergone a Computed Tomography (CT) scan of the abdomen then and there was an incidental mass seen at the Gastroesophageal junction. The patient underwent ERCP stenting for the CBD stones and was discharged. He did not follow up. He started experiencing similar pain again. Patient was non-diabetic, non-hypertensive, had no other co-morbidities. There was no previous history of any trauma or surgery. Patient also denied history of any addiction or drug abuse. His clinical examination was unremarkable. All haematological and biochemical investigations were within normal limits. Ultrasound abdomen revealed gall bladder with multiple calculi and wall thickening of 4 mm consistent with chronic calculous cholecystitis with a CBD stent in situ.

CT scan (Fig. 1) confirmed USG findings and was also suggestive of a well-defined heterogeneously enhancing mass measuring 7.8 x 3.5 x 5.0 cm at the gastro-esophageal junction with calcifications within. It was causing luminal compromise at GE junction with mild upstream dilatation of the esophagus. Most probable differential being gastrointestinal stromal tumor.

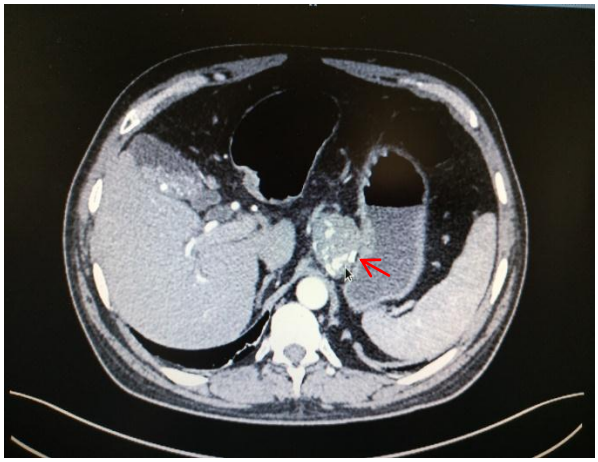

Fig. 1: CT scan showing a large GE mass with significant luminal compression and calcification. Also the gall bladder showed stones and slugde and was mildly thickened.

Esophagogastroscopy (OGDscopy) revealed bulge at the GE junction at 38 cm from incisors suggesting extrinsic compression along with transverse elongation of the lumen (Fig. 2A). Patient's endoscopic ultrasound suggested large hypoechoic mass of size 7x4 cm in lower esophagus at 40 cm from incisors and arising from the 4th layer and with multiple calcifications. (Fig.2B).

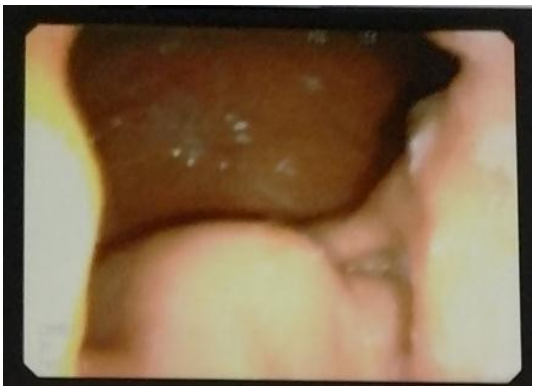

Fig. 2A: Esophagoscopy showing a bulge at GE junction with intact mucosa.

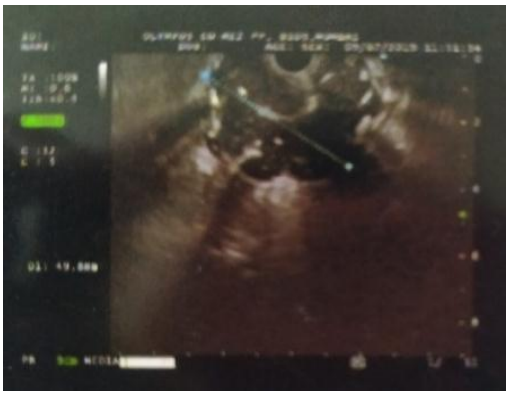

Fig. 2B: EUS showing the mass with calcifications.

After obtaining anaesthesia fitness and required consent, patient was taken up for Laparoscopic SOS open Cholecystectomy and Enucleation of the leiomyoma. However, due to frozen Calot's Triangle, it was converted to open laparotomy. Midline Abdominal Incision was taken and Subtotal Cholecystectomy was performed first. The gastroesophageal (GE) junction was mobilized. Intraoperatively a large elongated firm intramuscular well-defined tumour was noted at the GE junction with well-preserved planes and intact mucosa (Fig 3,4). Enucleation of tumour was performed. There was no evidence of mucosal breach as confirmed with intraoperative esophagoscopy and esophageal leak test. Muscular layer was approximated with intermittent sutures.

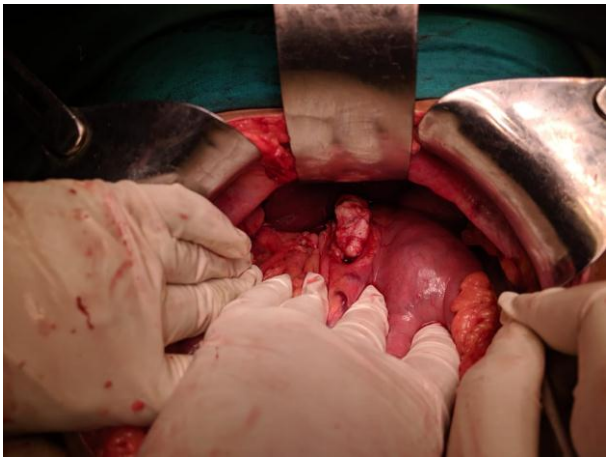

Fig. 3: Intraoperative image showing enucleating tumour at GE junction.

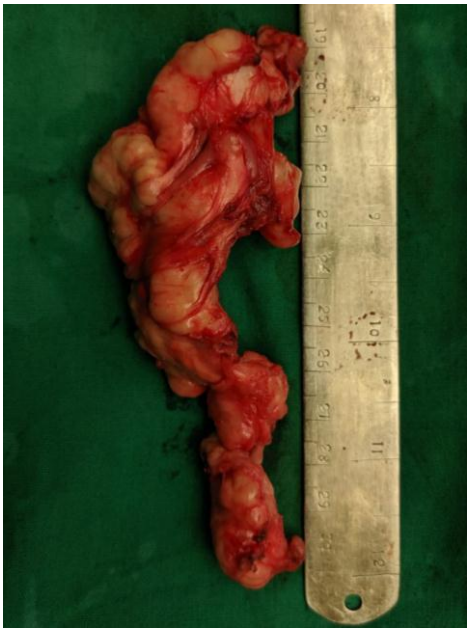

Fig. 4: Enucleated Surgical Specimen.

Tumour was sent for histopathological examination which confirmed it to be leiomyoma. Histopathologically, sections revealed benign spindle cell tumour with tumour cells arranged in interlacing fascicles and showing abundant eosinophilic cytoplasm with a few foci of calcification. (Fig.5A,5B). On

immunohistochemistry, tumour cells are diffusely and strongly positive for Desmin and SMA. Focal positivity for Ckit, Dog 1 was also seen, however expression was seen in less than 10% tumor cells.

Post operatively, the patient developed wound dehiscence and underwent serial Negative pressure dressing for the same and healing by secondary intention. He was discharged on POD 19. He underwent ERCP and removal of CBD stent after confirmation of clear CBD. Follow up at 2 months was symptom free.

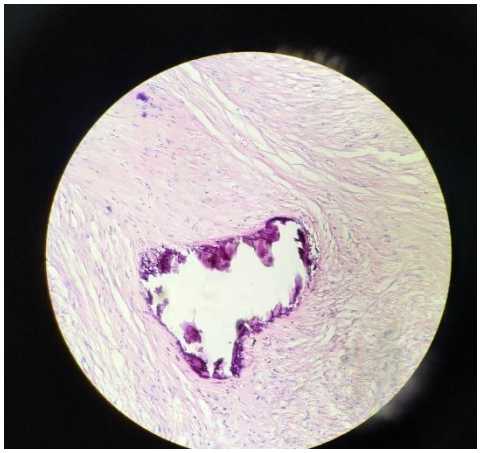

Fig. 5A: Microscopic images showing spindle cells with area of calcification. (20 x Haematoxylin & Eosin).

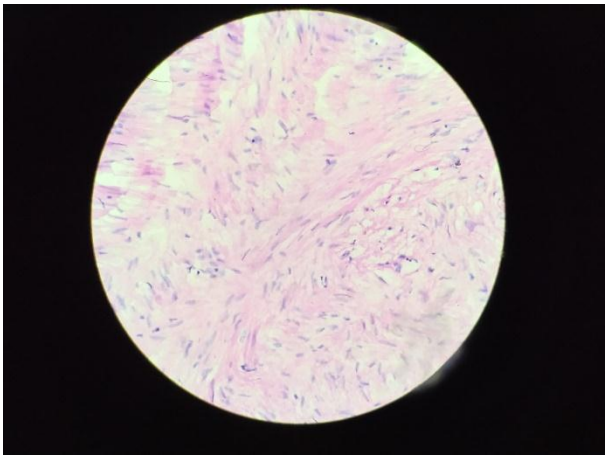

Fig. 5B: Microscopic image showing elongated nuclei in spindle cells arranged as fascicles. (40 x Haematoxylin & Eosin Stained).

DISCUSSION

Esophageal leiomyoma is most common benign esophageal tumour and accounts for about two third of all esophageal benign tumours. Studies using detailed histologic examinations have reported the frequency of esophageal leiomyoma to be higher, at 7.9%[3] Leiomyoma can occur at any age but 90% of cases occur in patients between the ages of 20 and 69 years, with peak incidence in the third to fifth decades. The age at which our patient presented was 49. The male-to female ratio is approximately 2:1 (in children it is reversed) and there are no apparent racial or geographic predilections for the disease.[4] Approximately 80% of leiomyomas are found intramurally and originate in the muscularis propria as seen in our case. Leiomyoma is a slow growing tumor and the size of the lesion may not change for many years. Approximately one-half of all tumors are smaller than 5 cm. Larger tumors are often elongated in a dumbbell-like shape.

About half of the patients with leiomyoma are asymptomatic; if symptomatic, the symptoms comprise of long standing retrosternal pain and dysphagia. In large tumours patient may also present with esophageal obstruction, regurgitation, weight loss and muscle wasting. Also quite rarely bleeding. Some studies suggest that there is no correlation between size and symptoms[5,6], however some suggest a correlation that a tumour size of more than 5.3 cm can cause symptoms.[7] In present case, the tumor was incidental finding.

Radiologically, if leiomyoma is large enough, a lesion will appear as a lobulated homogeneous mass. Barium swallow is the most common preoperative investigation. They will demonstrate a smooth filling defect like any other benign tumor of esophagus. Endoscopy is a valuable asset in determining the presence and location of the tumor and in assessing the condition of the mucosa. Postlethwait[8] cites four findings characteristic of leiomyoma: 1) the overlying mucosa appears normal and intact; 2) the tumor projects into the lumen to varying degrees; 3) the tumor is freely movable, with the overlying mucosa easily sliding over the lesion; and 4) narrowing of the lumen is common, but stenosis and obstruction to passage are not. Determining the layer of origin of a tumor is essential for the diagnosis and treatment of leiomyoma. The strength of endoscopic ultrasonography (EUS) lies in its ability to depict the five layers of the esophageal wall, making it possible to ascertain the layer of origin to differentiate between intramural and extrinsic lesions. On EUS leiomyoma appears as a well circumscribed, homogeneous, hypoechoic mass with a regular borders, and arise from the muscularis mucosa, submucosa, or muscularis propria. EUS has become instrumental in improving the diagnostic and staging accuracy of esophageal tumors.

On CT scans, most leiomyomas appear as eccentric, focal esophageal wall thickening. Larger leiomyomas usually appears homogeneously low- or isoattenuated masses. The surrounding mediastinal fat is usually not disrupted.

Histologically, on gross inspection leiomyomas are firm, well encapsulated, and often have a smooth or nodular surface. In section, they appear white to gray with a whorled surface. These tumors have low overall cellularity, appear strongly eosinophilic on hematoxylin-eosin stain, and are comprised of interlaced smooth-muscle cells with hypovascularity and absent mitoses. Typically leiomyoma is positive for desmin and smooth-

muscle actin and negative for CD34 and CD117. In present case too, the HPE report showed similar findings and was positive for Desmin and SMA.

Once a clinical diagnosis of leiomyoma is established many factors must be taken into consideration when determining an appropriate course of treatment. Of particular importance are tumor size, location, and morphology and the patient's symptoms and overall condition. Other indications for resection include uncertainty of diagnosis, mucosal erosion, regional lymph node enlargement, facilitation of other esophageal procedures and increase in size. There is a general consensus in the literature that esophageal leiomyoma should be surgically removed in symptomatic patients. Open resection has been the standard approach to resecting leiomyomas, but it has been recently challenged by minimally invasive and endoscopic methods. Many experts advocate the resection of asymptomatic tumors based on the following: 1) the possibility of malignant transformation; 2) the possibility of symptomatic transformation; 3) the need to obtain a definitive histologic diagnosis; and 4) the exclusion of malignancy only by removal.[5]

Tumour removal can be done either by thoracotomy with esophageal resection or enucleation of tumour by thoracoscopic approach. Tumours at gastroesophageal junction can be addressed through upper midline laparotomy as in present case. Esophageal resection is recommended for tumour of size more than 8 cm, when tumour is adherent to mucosa or if there is extensive mucosal damage during dissection.[9]

In present case, leiomyoma was more than 5 cm and patient was symptomatic and was already planned for another procedure under general anaesthesia, hence excision was planned.

CONCLUSION

Esophageal leiomyoma can present as an incidentaloma with a synchronous disease.

In view of large size and diagnostic dilemma, it is better to do surgically excision. Histopathology augmented with histochemistry confirms the diagnosis.

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