

Case Report

Fifty five years old male with gastrointestinal stromal tumor: a case report

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Received: 02 May 2019

Accepted: 05 June 2019

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ABSTRACT

Esophageal Gastrointestinal Stromal Tumors (GISTs) were extremely rare, with an incidence of 0,2% of GIST cases. Majority of esophageal GISTs were classified as high-risk category (70.83%), thus, it was required more aggressive approaches, e.g. radical surgery, chemotherapy and radiotherapy. This was a case of a 55-year-old male patient who was referred to surgery department complained of atypical chest pain and swallowing difficulty. Based on thorax X-Ray, it was suspected mediastinal mass. In addition, based on chest CT scan, it was suspected a mass at distal esophagus with multiple nodular lesions in liver dd/fatty liver. X-ray Oesophagus Maag Duodenum (OMD): Esophageal mass at 1/3 middle-distal part. Upper endoscopy diagnosis showed a fragile circularly spreading tumor mass which it was easily bleeds. Biopsy also showed a mild esophageal dysplasia with non-specific inflammation and necrotic tissue which was difficult to assess. This patient underwent McKeown esophagectomy. Moreover, pathology report showed diagnosis of a malignant tumor which fitted with criteria of malignant GIST, with both ends tumor-free. Finally, the patient was administered for imatinib as adjuvant therapy 1x400mg daily. This report illustrated complexity in diagnosing and treating esophageal GIST. The tumor size and mitotic rate of tumor were associated with poor survival.

Keywords: Gastrointestinal stromal tumors, Esophagus, McKeown procedure

INTRODUCTION

Gastrointestinal stromal tumor or gastrointestinal stromal tumor (GIST) was one type of cancers in gastrointestinal tract that had potential to be malignant. GIST was a mesenchymal cancer in gastrointestinal tract. Recently, all GIST cases were considered as potentially malignant tumors and all GIST cases without metastasis had to be resected.¹

GIST was a rare type of tumor, especially in esophagus. The European Society for Medical Oncology (ESMO) estimated that GIST incidence rate was 1.5/100,000 per year.²⁻⁴ Geriatric was one of risks of GIST cases.⁴ Thus,

average age of GIST patients were ranges from 60-65 years, so GIST was rare in children.²⁻⁴ Esophageal GIST only represented <1% of all GIST cases recently, with an incidence of 0.1-0.3/1,000,000 people. However, esophageal GIST had a worse prognosis than gastric GIST and a high mortality rate than GIST in other locations.⁵

Most of GIST cases carried mutations inside KIT gene or PDGFR α gene, where both genes are located on chromosome 4 and made code of type III tyrosine kinase receptors.⁶ Patients with GIST usually complained of symptoms such as subsequent gastrointestinal bleeding and anemia, a feeling of fullness faster, abdominal

distension and discomfort with or without pain due to tumor compression. Sometimes GIST was asymptomatic until it was found in an advanced stage.^{3,7,8} Early diagnosis of GIST could be determined using endoscopy, endoscopic ultrasound (EUS) or computed tomography (CT). Management of GIST was generally a surgical therapy. In esophageal GIST, surgical technique with minimal invasion or minimally invasive esophagectomy (MIE), torakoabdominal technique, Ivor Lewis technique, transhiatal esophagectomy and McKeown esophagectomy used.⁹⁻¹²

CASE REPORT

Mr. H, a man, 55 years old, came with a complaint of difficulty of swallow for 8 months before being hospitalized, whether solid or liquid food, nausea, no vomiting, and a heart burn. There was a weight loss history, around 8 kilograms in the last 5 months. Family history of tumor was denied, however, there was a smoking history. Patient had a sufficient nutritional status. Based on physical examination, there was anemic conjunctiva but enlarged lymph nodes in the neck was not palpable. Based on laboratory examination, it was found anemia (Hb 8.8 g/dL), normal CEA (<0.5). Moreover, based on results of upper endoscopy, it was found tumor masses running circularly, brittle and easily bleeding. At 35 cm from the incisor (before LES), it appeared that tumor mass was protruded as it appeared from the stomach and there were signs of active bleeding. Based on biopsy, there was esophagus of moderate dysplasia, with non-specific inflammation+necrotic tissue that was difficult to evaluate. In addition, impressive thoracic CT scan, it was suspected distal esophageal mass, with multiple nodular lesions of liver, thus suspected to focal fatty liver and tumor metastasis to the liver. X-Ray Oesophagus Maag Duodenum (OMD) results also showed 1/3 middle esophageal mass to distal and thorax X-ray showed impression of mass on the mediastinum as well (Figure 1) (Figure 2) (Figure 3) (Figure 4) (Figure 5).



Figure 1: Thorax X Ray: impression of mass on the mediastinum.

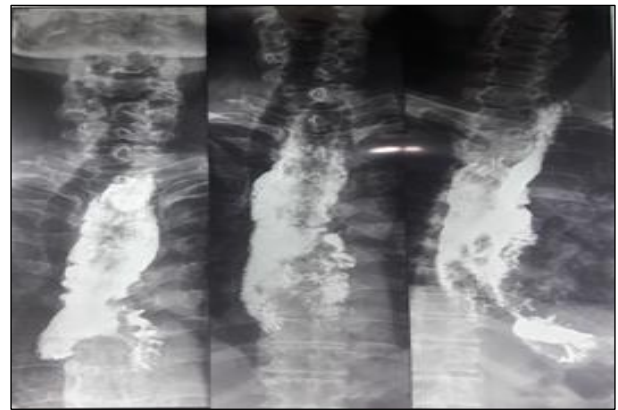


Figure 2: Esophageal mass 1/3 middle to distal.



Figure 3: CT imaging of the thorax: suspected distal esophageal mass, with multiple nodular lesions of liver, with suspected focal fatty liver and tumor metastasis to the liver.



Figure 4: CT scan of the thorax: suspected distal esophageal mass, with multiple nodular lesions of liver with suspected focal fatty liver and tumor metastasis to the liver.

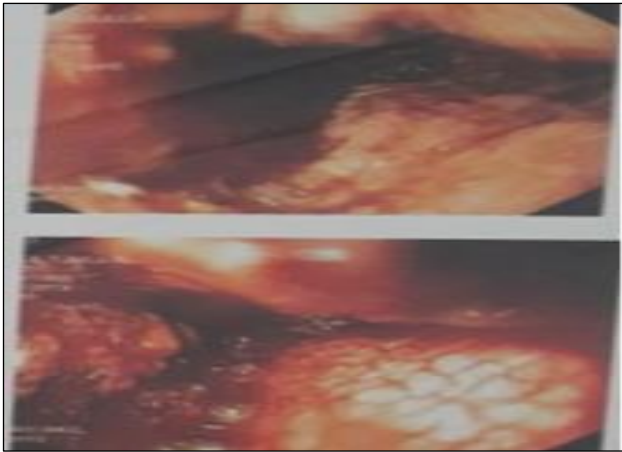


Figure 5: Results of upper endoscopy, it was found tumor mass running circular, brittle, easily bleeding. At 35 cm from the incisor (before LES) it appeared protruded tumor mass from the stomach and there were signs of active bleeding. Biopsy: Esophagus of moderate dysplasia, with non-specific inflammation and necrotic tissue that was difficult to evaluate.

Therefore, it was concluded that the patient was diagnosed with an esophageal tumor 1/3 middle suspected malignancy, T2N0M0 (Stadium IIA), Karnofsky score 70. Then, we performed a McKeow operation. During surgery process, it was found tumor mass reached the middle 1/3 of the esophagus, felt soft and indistinct and tumor diameter was approximately 5 cm. Then cervical esophageal resection was performed, gastric pull-up resection and end-to-end esophageal cervical and gastric anastomosis were also performed. 1/3 of middle esophagus and tumor were then examined by anatomical pathology. Duration of operation was around 4 hours, with bleeding of 500 cc, no transfusion during surgery and no intraoperative hemodynamic problems. After surgery the patient was treated in intensive unit for 25 days. After surgery, tumor mass was examined in anatomical pathology laboraroty and the examination results was a malignant tumor that was suitable for malignant GIST, with both ends of tumor-free resection. It also showed lymphadenopathy with a reactive picture of hyperplasia (Figure 6).



Figure 6: Image of removed tumor, 5 cm in diameter.

DISCUSSION

The esophageal GIST as previous mentioned was a rare case, the incidence was less than 1%, with incidence of men was similar in women, occurred at the age of 50-70 years, thus, this case was appropriated. Some sources stated that location of 1/3 distal was the most common (70.83%), then 1/3 Middle (11.40%), however the most rare cases was in 1/3 proximal (1.76%). This case was about 1/3 center to distal. The diagnosis and management of GIST in the esophagus had its own dilemma, such as endoscopic biopsy which was sometimes not representative for a GIST, and the same thing we encountered in this case.

The results of endoscopic biopsy only showed a non-specific inflammation. Other sources said that EUS-FNAB or core biopsy was recommended, although this was still debated, because this action could damage the GIST capsule, then resulted to a difficult enucleation process. Another source stated that preoperative biopsy was indicated if tumor size was more than 2cm and/or planned for neoadjuvant chemotherapy. Whereas according to NCCN preoperative biopsy was not necessary if the tumor was resectable.

An indication of tumor surgery conducted if tumor was suspected to be a GIST due to potential for high GIST malignancies. Thus, surgery was chosen based on GIST localization or potential GIST to be resected. In gastric GIST, resection was recommended when tumors measuring > 2 cm or tumors enlarged or showed signs of malignancy such as uneven edges, ulcers, bleeding, changes in consistency, necrosis or heterogeneous echogenicity on endoscopy or EUS^{8,13}

Tumors which were smaller (< 2 cm) without signs of malignancy could be actively monitored. However, small tumor size did not rule out of possibility of malignancy in the GIST. In non-gastric GIST, resection was highly recommended regardless of tumor size or morphology.¹³

In esophageal GIST, surgical techniques with minimal invasion or minimally invasive esophagectomy (MIE) had been developed to reduce complications and mortality. The MIE technique included a hybrid technique minimally invasive esophagectomy (HMIE) or totally minimally invasive esophagectomy (TMIE) which was more superior compared with an open esophagectomy technique as this was associated with perioperative results without reducing oncological safety.⁹

The most common approach to esophageal surgery was a torakoabdominal technique that opened abdomen and thoracic cavity together and used Ivor Lewis technique where abdomen was opened earlier than thorax¹⁰ (Figure 7).

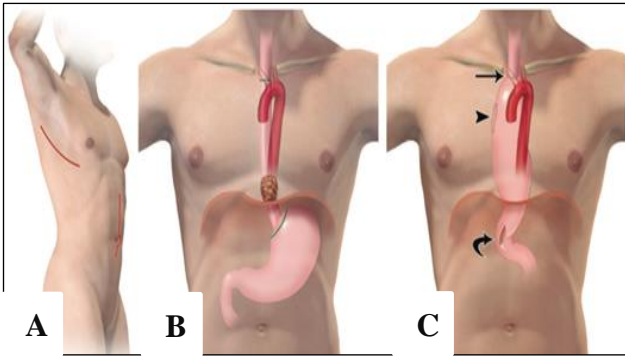


Figure 7: Esophagectomy with Ivor Lewis. Skin incisions (red line) for upper abdominal laparotomy and right thoracotomy (A), resection line (green) and tumor in distal esophagus (B), and post surgery anatomy after esophagus resection and right intrathoracic anastomosis was made (straight arrow line) in thorax between the remnants of esophagus and gastric tract (C).¹⁴

Another surgical technique that can be used was transhiatal esophagectomy which was commonly used in tumors located in lower third of esophagus and gastric cardia and other benign cases.¹⁰

The advantage of this transhiatal technique was respiratory system complications could be reduced by avoiding thoracotomy and intrathoracic anastomosis as intrathoracic anastomosis leakage could be occurred¹⁰ (Figure 8).

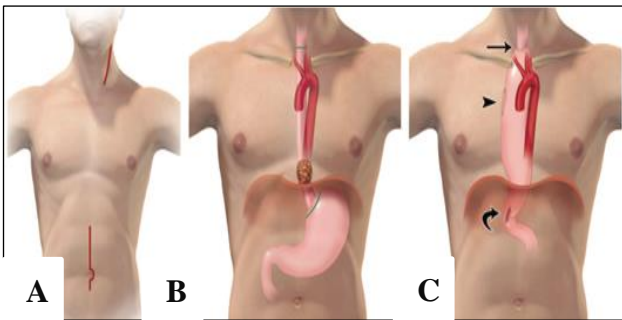


Figure 8: Transhiatal esophagectomy. skin incision (red line) for upper abdominal laparotomy and cervical anastomosis (left neck) (A), resection line (green) and tumor in distal esophagus (B), and postoperative anatomy after esophageal resection and right intra-thoracic anastomosis was made (straight arrow) on the thorax between the remnants of esophagus and the gastric tract (C).¹⁴

One of the techniques which was most often used in treating esophageal carcinoma was the McKeown technique. The McKeown technique was usually used in surgery for proximal GIST tumors, where one third of incision was made in neck for cervical anastomosis.¹⁰ The McKeown procedure was one of surgical techniques used with minimally invasive resection of esophageal

carcinoma. The McKeown technique was defined by thoracic esophageal mobilization; lymph node dissection, ligation of thoracic duct (using thoracoscopic and open thoracostomy techniques); abdominal exploration (using laparoscopic and open laparotomy techniques); gastric mobilization; lymph node dissection; feeding jejunostomy (feeding through jejunum); left cervical incision for anastomosis.^{11,12}

A previous study conducted by D'Amico explained that the McKeown technique had a better potential advantage than other techniques which included lower local recurrence rates, easier management of neck anastomosis and less expansion of thoracic incisions due to anastomosis conducted in neck, not in chest¹⁴ (Figure 9); (Figure 10).

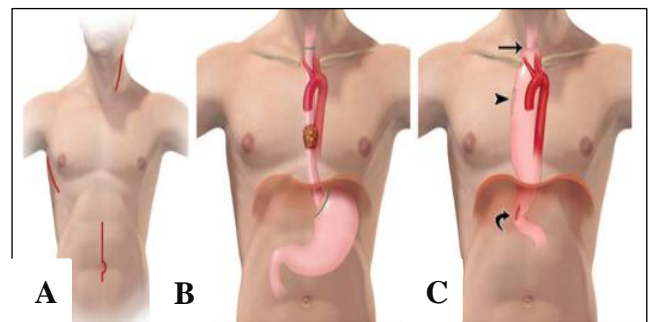


Figure 9: McKeown esophagectomy. Skin incision (red line) for right thoracotomy, upper abdominal laparotomy and cervical anastomosis (left neck) (A), resection line (green) and tumor in mid-esophagus (B) and postoperative anatomy after esophageal resection and right intra-maxillary anastomosis was made (straight arrow) on thorax between the remnants of esophagus and gastric tract (C).¹⁴

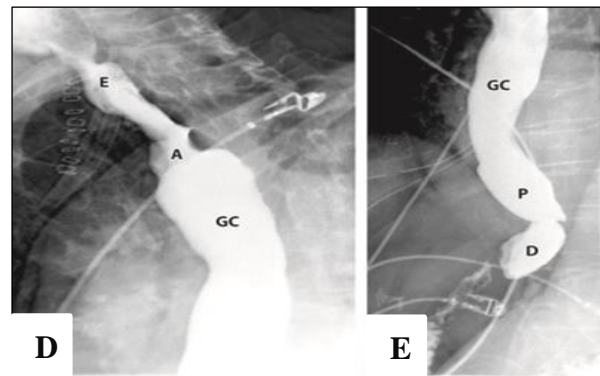


Figure 10: Esophagrams of upper thorax (D) and lower (E) in posterior side of right oblique in patients with ESCC in distal esophagus showed anatomy after McKeown esophagectomy. A = cervical anastomosis, D = duodenum, E = leftover esophagus, GC = gastric tract, P = pylorus.¹⁴

One potential advantage of McKeown method was better exposure treatment and cervical lymphadenectomy. In

addition, cervical anastomosis was relatively simpler and easier to do so the technique was more comfortable and easier to understand for beginners.¹⁴

Pulmonary complication was one of the most common postoperative complications as two-thirds of postoperative deaths were caused by pulmonary complications. There were 30% of postoperative pulmonary complications included pleural effusion, atelectasis, kilotorax, pneumonia, pulmonary embolism and respiratory failure.¹⁴⁻¹⁶

Anastomosis leakage was also one of esophagectomy complications that could occur in postoperative. A study data showed anastomotic leakage occurred in 10% -40% of postoperative patients and accounted for as much as 40% of postoperative patient mortality.¹⁴⁻¹⁶

Anastomosis leakage usually took place within the first 10 days after surgery and could be caused by stress, either too much tension or less on anastomosis. Lack of stress could affect to weak tissue appositions with inhibited extravasation and healing while excessive stress could affect to ischemia and necrosis¹⁴ (Figure 11).

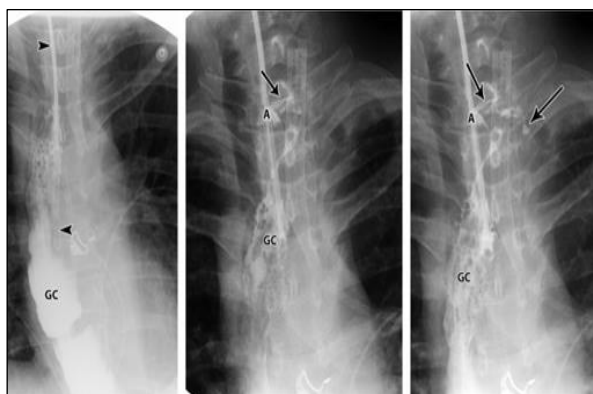


Figure 11: Cervical anastomosis leakage.
GC = gastric conduit.¹⁴

Some technical complications that could occurred related to esophagectomy procedures were injury to recurrent laryngeal nerve, kilothorax, bleeding, tracheobronchial injury, and diaphragmatic herniation. Injuries to recurrent laryngeal nerve usually occurred at the time of cervical dissection and had an incidence rate of 10% -20% when cervical anastomosis was formed. This could affect to patient's voice and also risk of aspiration of patient.¹⁴

Kilotorax was a rare complication. Kilotorax incidence was due to injury to thoracic duct and had a recurrence rate of 1% -5%. The loss of chyle continuously could result in a decrease in a number of lymphocytes, nutritional deficiencies, low body immunity which could lead to systemic infections.¹⁴

Despite of earlier mentioned complications, one of challenges that had to be faced after esophagectomy was

recovery of body functions as functional complications could be occurred such as late gastric emptying, dumping syndrome, and esophageal reflux.¹⁴

In patients who had approximately 5 cm tumors with no metastases to other organs, it was decided to use McKeown procedure. This technique was chosen because it was easier than other techniques which included lower local recurrence rates, easier management of neck anastomosis and less expansion of thoracic incision because anastomosis was performed in neck, not in chest.

Pathological Anatomy results showed both tumor-free ends, spindle cells, and many mitoses. Immunohistochemical results: CD 117: +++. Based on the National Institute of Health (NIH) on Risk Classification Modification for Primary GIST, this case was high risk category. Based on Guideline GISTs, this patient was given imatinib 1x400mg for 2 years, CT scan control for 3 months in the first 2 years, followed by 6 months for the next 2 years, then once a year.

CONCLUSION

Esophageal Gastrointestinal Stromal Tumor (GIST) was one type of malignancy that was quite interesting for digestive surgeons because it is one of rare diseases. Esophageal GIST had a worse prognosis and higher mortality than other types of malignancy. The most common treatment for esophageal GIST was based on stage of tumor, generally if it was a resectable tumor , then it would be performed surgery.

McKeown esophagectomy technique was the most common technique used in GIST cases. In McKeown esophagectomy technique, there were three incisions conducted such as right thoracotomy, laparotomy and left neck incision to make cervical anastomosis. This technique was easier for digestive surgeons, especially for beginners, because the technique was simple and sufficient exposure. However, common complications of McKeown esophagectomy were pleural effusion, atelectasis, kilothorax, pneumonia, pulmonary embolism and respiratory failure.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Laura SF, Warsingih W. Kusuma MI, Faruk M. Fifty five years old male with gastrointestinal stromal tumor: a case report. *Int J Res Med Sci* 2019;7:2823-8.