Abstract:
We report a rare case of extranodal histiocytic sarcoma with involvement of oesophagus, oesophago gastric junction and stomach which has not been documented in the literature so far. A provisional diagnosis of mitotic lesion was made based on the repeated upper g.i. endoscopy. Computed tomography scan revealed, circumferential gasoesophageal wall thickening involving the lower oesophagus, oesophago gastric junction and proximal stomach. Patient underwent sub total oesophagectomy and total gastrectomy with colonic interposition. The histiocytic origin of the tumor was confirmed by positive reaction of the tumor cells for CD 163. He is under regular follow up and receiving adjuvant radiotherapy.

Keyword: histiocytic sarcoma, oesophago gastric junction, gastrectomy

INTRODUCTION:

HISTIOCYTIC SARCOMA- OF OESOPHAGUS, OESOPHAGO GASTRIC JUNCTION & STOMACH-FIRST CASE REPORT
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Histiocytic tumors are rare comprising less than 1% of all nodal or extranodal lymphoid tissue tumors\textsuperscript{1,2}. The term histiocytic sarcoma was introduced in 1970 by Mathe et al\textsuperscript{3} Recently, World Health Organization (WHO) has defined histiocytic sarcoma as malignant proliferation of cells showing morphologic and immunophenotypic features of mature tissue histiocytes\textsuperscript{4}. The diagnosis of histiocytic sarcoma relies predominantly on the confirmation of its histiocytic lineage and the exclusion of other poorly differentiated tumors like lymphoma, carcinoma, sarcoma, and melanoma\textsuperscript{5}.

**CASE REPORT:**
A 41 year old gentleman has visited multiple centres with dysphagia for a period of three months. All the three endoscopists have made a diagnosis of mitotic disease involving the esophagus and stomach but the biopsy did not confirm the diagnosis of either squamous cell carcinoma or adenocarcinoma. CECT of chest and abdomen had picture suggestive of malignancy involving the esophagus, GE junction and stomach (fig 1, 2). A diagnostic laparoscopy was performed which showed serosal involvement at GE junction and proximal stomach. An omental nodule found was biopsied and was found to be non-mitotic. A multidisciplinary team consisting of Gastro Intestinal surgeons, Medical Gastroenterologist, Radiation Oncologist and Medical Oncologist decided on surgery for removing the disease, definitive diagnosis and relief of dysphagia. After obtaining informed consent, the patient underwent subtotal esophagectomy and total gastrectomy (fig 3, 4) with colon reconstruction with cervical anastomosis. Post operatively he had anastomotic leak at cervical region which was managed conservatively. The final pathology report was Histiocytic sarcoma. The patient is now on follow up radiotherapy.

**DISCUSSION:**
Histiocytic sarcoma is an extremely rare non-Langerhans histiocytic disorder of monocyte-macrophage lineage. It accounts for less than one percent of all nodal and extranodal lymphoid tissue tumours\textsuperscript{1,2}. Middle aged people (median age 46-55 years) are most commonly affected with no gender preponderance. The disease can be unifocal or multifocal with involvement of skin, soft tissues, bone, lymphnode, liver, spleen and central nervous system. Isolated involvement of Gastro intestinal tract involvement is common involving ilium, rectum, anus, stomach and esophagus. However the involvement of esophagus, Esophago-Gastro junction and stomach as a whole like our case was never seen before. Diagnosis is made by Histopathological examination which shows large polygonal or ovoid cells with abundant eosinophilic cytoplasm and irregularly folded eccentrically placed pleomorphic nuclei. Immuno-Histochemistry staining is positive for markers CD163, CD68, LYS and sometimes S-100 as in our case and are specifically negative for T and B cell markers (Fig 5)\textsuperscript{6,7}. Radical surgery followed by adjuvant Radiotherapy gives the best results while adjuvant chemotherapy is reserved for inoperable cases\textsuperscript{8}. We decided to proceed with surgery as the patient had symptomatic operable tumour with undiagnosed pathology. A preoperative definitive diagnosis of Histiocytic sarcoma, would not have altered the line of our management. The prognosis depends on the stage of the disease.
CONCLUSION:
This case is presented for its extremely rare diagnosis of Histiocytic sarcoma, and the first known case in the literature to the best of our knowledge to affect esophagus, Esophago Gastric junction and stomach in continuity. Pre op biopsy report:

Biopsy report:
Section shows marked thickening of gastric mucosa with extensive ulceration and inflammatory infiltrate, comprising predominantly eosinophils, lymphocytes, plasma cells and giant cells. Stroma showed congested vessels, inflammatory infiltration of entire thickness of wall up to serosa, section normal. All nodes show reactive hyperplasia.
Impression: eosinophilic gastritis

REFERENCES: