ABSTRACT
Background: Gastrointestinal stromal tumors (GISTs) are the most common mesenchymal neoplasms of the gastrointestinal tract believed to originate from interstitial cells of Cajal. Development of GISTs outside the gastrointestinal system, called as Extra-intestinal gastrointestinal stromal tumor (EGISTs) is rare. Incidence of primary GIST in the greater omentum has been reported to be <1%. Case report: A rare case of EGIST in an 89 year old woman, arising from the greater omentum is presented here. In this patient EGIST was primary and of epithelioid type with prominent myxoid change.
Conclusion: Recognition of myxoid epithelioid variant of EGIST is important as it is defined as a separate entity with availability of molecular targeted therapy.

Key words: Myxoid epithelioid EGIST, Greater omentum, Primary.

CASE REPORT

An 89 year old woman presented with abdominal fullness for duration of two months. A physical examination revealed a firm, large abdominal mass, extending from the left hypogastrum to the umbilicus. An endoscopy of the GI tract showed a normal mucosal study. A computed tomography (CT) scan of the abdomen revealed a large, ill-defined and heterogeneous enhancing soft tissue exophytic mass arising from the omentum. (Fig 1) The mass was not adherent to any intra-abdominal structure. At laparotomy, a well encapsulated, large mass measuring 20 x 15 cm was found in the greater omentum. The tumour was completely resected and anorectum turned up for follow up since her discharge. A diagnosis of Myxoid Epithelioid GIST of the greater omentum was made based on the histological and immuno-histo-chemical findings. Patient has not turned up for follow up since her discharge.
DISCUSSION

Gastrointestinal stromal tumors (GISTs), the mesenchymal tumors arising commonly in the gastrointestinal tract and occasionally outside the GIT constitutes 1% of all GI malignancies. These tumors are derived from interstitial cells of Cajal, the primitive stem cells that can differentiate into both interstitial cells of Cajal and smooth muscle cells and these tumors are defined by expression of CD 117 immune reactivity. It is now accepted that mutations in the kit receptor tyrosine kinase protein play a central role in the pathogenesis of GIST and likewise these lesions are designated as specific ckit expressing tumors and kit signaling driven mesenchymal tumors.

Extra-intestinal Gastrointestinal stromal tumors (EGISTs) arise outside the GIT but share a similar morphological, immuno-phenotype and molecular genetic characteristics with GISTs. EGISTS are rare and more aggressive compared to GISTs.

Gastrointestinal stromal tumors in the omentum apparently arises from the normal CD 117/CD 34 positive mesenchymal cells, like the cells of Cajal, as reported by Sakurai ET al. Gastrointestinal stromal tumors have been reported in all age groups. However, they occur predominantly in adults, the median age being 60 years and shows a male predilection. A literature survey by Fagkrezos et al states that the median diagnosis of omental GISTS is 65 years with an even male to female ratio. The authors further state that there is no difference in the occurrence of GIST between greater and lesser omentum.

Most of the patients with omental GISTS show symptoms as these tumors are large. Miettinen et a and Reith et al state that only large tumors produce symptoms. In our case, since the tumor was large, patient presented with symptoms. Since the tumors are large at initial diagnosis, the clinical symptom is often delayed. Imaging diagnostic modalities such as CT and MRI plays an important role in pre-operative diagnosis of EGISTS and in addition helps in obtaining material for guided aspiration/biopsies. Ortiz-Ryet et al underlines the importance of this imaging diagnostic modality in EGIST, especially in obtaining material for fine needle aspiration.

Omental GISTS seem to be morphologically and immuno-histo-chemically identical to their gastric counter parts. Histologically they can be either spindle cell, epithelioid cell or show a mixed pattern. Our case showed predominantly an epithelioid pattern. Generally EGISTS like GISTs shows a strong expression for CD117 and CD 34 immuno-histo-chemically. At times, expression of CD 117 may be weak or absent. Among the different types of GISTS, epithelioid GISTS are known to be...
more often negative for CD 117 protein. However, our case showed positivity to both CD 117 and CD 34. As suggested by all workers, we are also of the opinion that a combination of immuno-histo-chemical markers for c-kit and CD 34 are the most reliable markers for the diagnosis of EGISTS and GISTS.

One distinct feature about the present case is that the epithelioid variant showed a prominent myxoid change. Myxoid change in the stroma is a rare occurrence. Suster et al suggested that GIST with a myxoid stroma is a distinct morphological variant of myogenic GIST and needs to be differentiated from benign schwannoma of the stomach and Gastrointestinal Autonomic Neural Tumors (GANT). The authors further mention that the myxoid change observed in these tumors probably represents a secondary, non-specific reaction pattern of the tumor cells to some noxious stimulus or it may be a form of degenerative phenomenon. We concur with the statements made by Suster et al that myxoid change represents a degenerative phenomenon.

Another interesting fact about myxoid epithelioid GISTs is that some of the workers have suggested it to be a distinct subtype of GIST that shows PDGFRA gene mutation. The authors further mention that recognition of such subtle histological feature is necessary for molecular sub classification of GISTs that are important for molecular targeting therapy by imitanib mesylate, inspite of weak or negative expression of CD 117 on IHC. We stress that the need for PDGFRA gene mutation studies in all cases of myxoid epithelioid EGISTs. Unfortunately PDGFRA gene mutation analysis could not be done in our patient.

CONCLUSION
Recognition of myxoid epithelioid variant of EGIST is important for two reasons: one, for weak/negative expression of CD 117 protein immuno-histo-chemically which may prevent its recognition; two, for the presence of PDGFRA gene mutation which enables the oncologist for use of imitanib mesylate as a molecular targeted therapy.

REFERENCES