

# Heavily Calcified Gastrointestinal Stromal Tumour of Stomach: A Diagnostic Dilemma

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## Research Article

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# Abstract

Gastrointestinal stromal tumours (GIST) are the most common mesenchymal tumour of gastrointestinal tract and the stomach being the most commonly involved organ. Focal calcification may be seen in GIST but prominent or heavy calcification is rare. Gastric mass with prominent calcification on imaging may create a diagnostic dilemma. We present a rare case of gastric GIST with heavy calcification in a 55 years old female presenting with abdominal lump. Computed tomography (CT) showed a large heterogenous juxta gastric mass with solid-cystic component with heavy calcification. She underwent laparotomy and en-bloc gastric sleeve resection with the mass. Microscopic examination showed tumour with spindle cell and calcification with mitotic index of 6/50 High power field. Immunoreactivity with Vimentin, CD34 and DOG 1 confirmed diagnosis of GIST. Dystrophic calcification of necrotic or degenerative tissue is thought to be cause of calcification in GIST. Very few cases of heavily calcified GIST have been reported in literature, our case is of interest because presence of solid cystic component and a huge size ~ 14 cm (longest diameter).

## Introduction:

Gastrointestinal stromal tumours (GISTs) are unique neoplasms that occur throughout the gastrointestinal tract, mesentery, omentum or the retroperitoneum. They are the most common mesenchymal tumour of the gastrointestinal tract and are defined by the expression of the c-KIT, CD 117 & DOG 1. It is thought to be arising from the intestinal cells of Cajal throughout the gastrointestinal tract, mainly involving the stomach ~ 60–70 % cases, with small bowel (20–30%), colorectal (~ 5%) and oesophagus (< 1 %) in few cases.<sup>1,2</sup> They have an exophytic growth pattern and may also have cystic degeneration, necrosis, haemorrhage although calcifications were uncommon. Focal calcification within GIST has been reported, ranging from 10–50% in reported series.<sup>3,4,5</sup> However, extensive thick calcification is a rare phenomenon.<sup>1</sup>

## Case Presentation:

A 55-year-old woman presented to our outpatient department with an abdominal lump for 6 months duration associated with upper abdominal discomfort without any family history of malignancy. There was no history of any bleeding in form of hematemesis or melena or loss of appetite or weight. Her pre-referral ultrasound showed a large solid cystic mass reported to be arising from pancreatic. General examination was unremarkable. Abdominal examination revealed a large ~ 14 x 10 cm hard multilobulated mass in the epigastrium with restricted mobility. Computed Tomography showed a large juxta gastric (near lesser curvature) mass with solid and cystic component with heavy calcification (Fig. 1) compressing duodenum, head of pancreas, left lobe of liver; there was no evidence loco-regional lymph node enlargement. Endoscopic ultrasound showed a large heterogenous solid cystic mass probably arising from peripancreatic region (Fig. 2A). Cyst fluid analysis showed normal CEA and CA 19 – 9 level. Considering the large size of the mass, laparoscopic resection was not attempted and open approach was preferred. Intra-operatively, a large 14 x 12 cm exophytic mass was arising from stomach

wall (more towards lesser curvature) and it was resected en bloc with a sleeve of stomach. Cut specimen showed a variegated solid cystic mass with areas of sclerosis and dense calcification (Fig. 2B). Postoperative course was uneventful and she was started on oral diet on POD 2 and discharged on POD 5. Histopathology showed partially encapsulated spindle cells, arranged in short fascicles with nuclear palisading. Immunohistochemistry confirmed the diagnosis of gastric GIST (Fig. 3). It showed a positive reactivity with CD 34, DOG 1, vimentin and negative for CD 117, desmin, SMA and S100. Mitotic index was 6 per 50 High power field and Ki67 index was 6%. Considering size of the tumour & mitotic index, she was started on adjuvant imatinib therapy. On follow up at 2 years she is healthy and disease free on radiological evaluation.

## Discussion:

The stomach is the most common location of GIST but accounts 2–3 % of all gastric tumours. Usually presents as a mass arising from wall of stomach with extra-gastric extension. The extra-gastric location also makes it difficult to appreciate the origin of the tumour, especially with large tumours. Careful evaluation of gastric wall thickening may give clue to the diagnosis. A peripheral enhancement pattern was present in majority on CECT. Central areas of low attenuation correspond to haemorrhage, necrosis or cyst formation. Patchy calcifications may occur within the primary mass of large GISTs, and the reported series have indicated a wide variability among these (10–50%).<sup>19</sup> Heavy calcification is a rare feature, seen only in 3% cases.<sup>20</sup> Extensive calcified GIST has been reported only few cases in the literature. Indeed, only nine cases have been reported till now in our search, six cases arising from stomach<sup>6–13</sup> and three from colorectum.

Clinically, a large peri-gastric mass with prominent calcification includes various differentials from pancreas, solid pseudopapillary tumour or mucinous neoplasms which may be associated with peripheral calcifications (egg shell calcification). Serous cyst adenoma will have central calcifications (sunburst calcification). This cystic neoplasm of pancreas may also present with calcification. Diffuse calcification may be associated with mucin producing adenocarcinoma stomach. Production of PDGF (platelet derived growth factor) and BMP (bone morphogenic protein) has been proposed as a stimulator of osteoblastic lineage, which also play important role in regulation of bone formation.<sup>12</sup> Pathologically, dystrophic calcification is most accepted theory.<sup>13</sup> It usually occurs in degenerated tissues like necrosis or haemorrhage. In alkaline environment, the binding of denatured protein to both phosphate and calcium ions ultimately forms calcium phosphate precipitates.<sup>16</sup> Several reports of calcification in metastatic sites of GIST after starting treatment with imatinib, perhaps follows a different mechanism of calcification.<sup>14,15</sup> The biological behaviour of calcification in GIST also has been evaluated by researchers. Kim et al,<sup>17</sup> showed no CT features other than size correlated with the biological behaviour. However, case reports have shown GIST with prominent calcification carries a less aggressive behaviour with indolent course because of their low mitotic index.<sup>18</sup>

Our case a rarity of clinical presentation with such a heavily calcified GIST with solid cystic component and a largest size reported in literature. In conclusion, gastric GIST may also present with calcification and should be consider as differential diagnosis in patients presenting as calcified solid cystic mass in epigastrium.

## Abbreviations

GIST: gastrointestinal stromal tumour, CECT: contrast enhanced computed tomography, EUS: Endoscopic ultrasound

## Declarations

**Conflict of Interest/Competing interest:** The authors declare that they have no competing interests.

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### Compliance with Ethical Standards

**Authors' Contributions:** B Har wrote the manuscript and reviewed the literature. Vinay K Kapoor and Anu Behari critically revised the manuscript for important intellectual content. Vinay K Kapoor supervised the process and approved the final draft. Bappaditya Har is the corresponding author and is the article guarantor. All the authors have read and approved the final paper to be submitted for publication.

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**Consent to Participate:** Obtained

**Code Availability:** Not Applicable

**Consent for Publication:** Written informed consent for publication of their clinical details and clinical images was obtained from the patient

**Availability of data and material:** All data generated or analyzed during this study are included in this published article

**Patient's consent:** Informed consent obtained for publication of this case All authors helped in writing, editing and critically evaluating the final manuscript

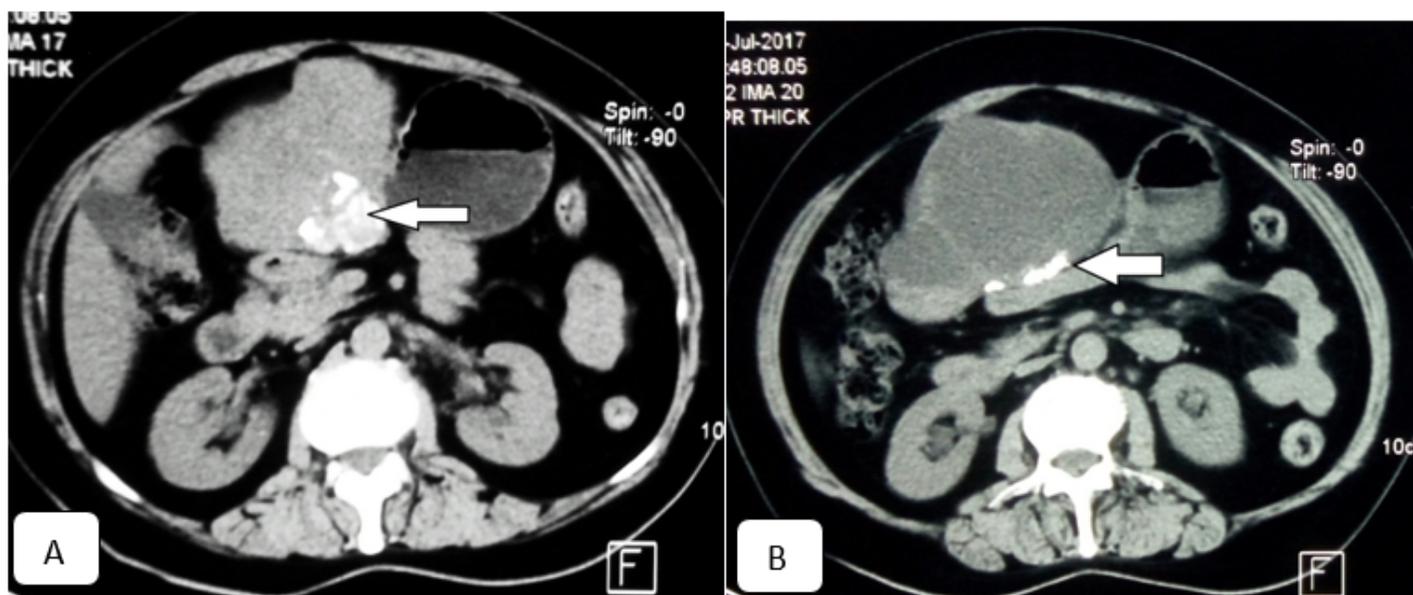
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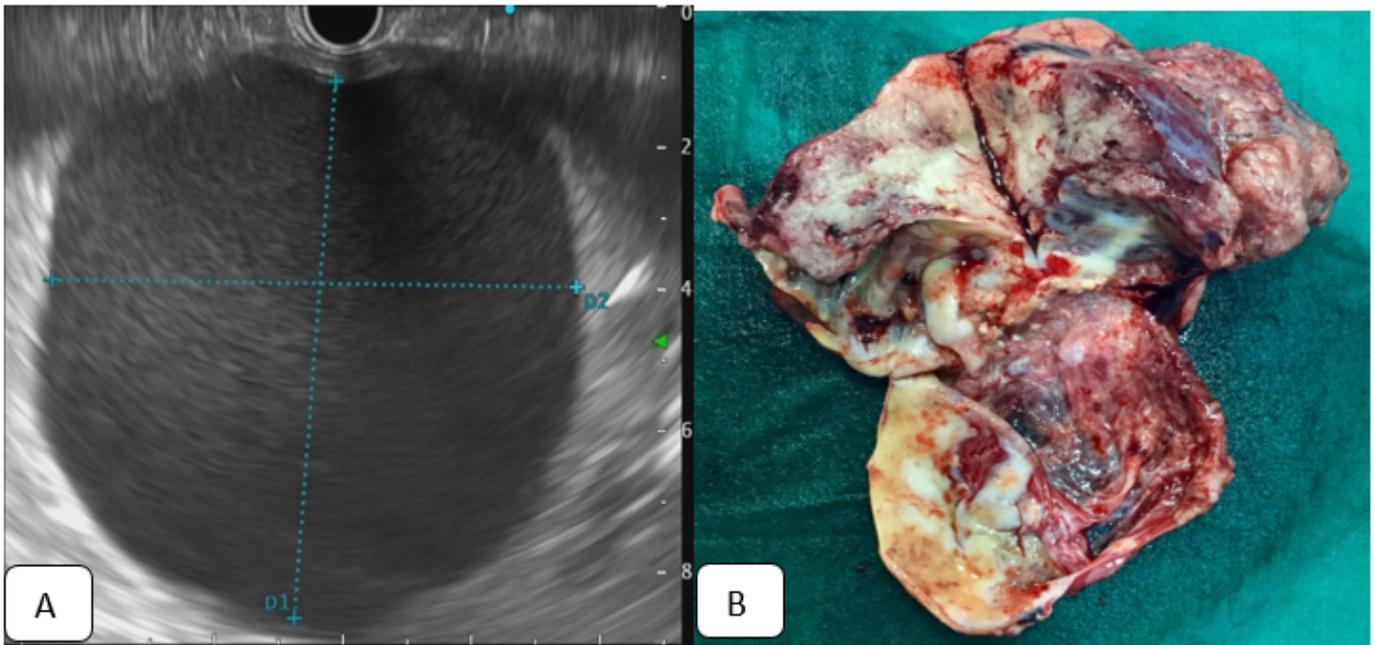
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## Figures



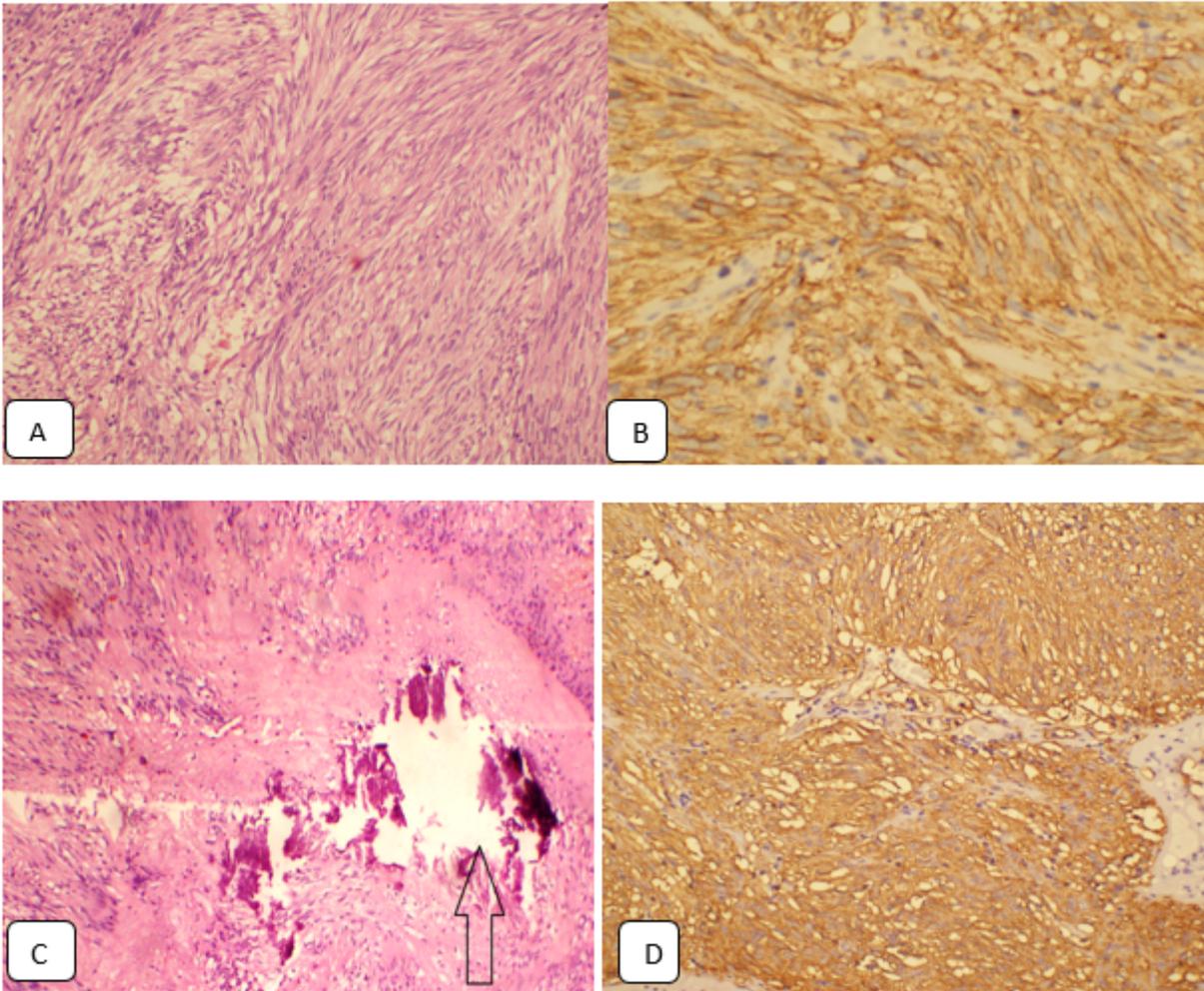
**Figure 1**

(A) Non contrast abdominal tomography axial sections showing juxta gastric mass with dense calcifications (arrow); (B) Non contrast abdominal tomography axial section showing juxta gastric mass with solid-cystic component with calcifications (arrow). Note mass stretching the lesser curvature of the stomach.



**Figure 2**

(A) Endoscopic ultrasound showing cystic component within the mass; (B) Specimen (cut section) showing solid cystic mass with variegated appearance and areas of necrosis.



**Figure 3**

(A) Histopathology (H & E stain) showing encapsulated spindle cells, arranged in short fascicles with nuclear palisading (B) Immunohistochemistry with DOG 1 showing positivity; (C) Histopathology (H & E stain) showing intratumoral calcification (arrow); (D) Immunohistochemistry with CD 34 showing positivity