# A rare extra-gonadal Germ Cell Tumour mimicking ischaemic heart pain.

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**Research Article**

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# Abstract

Background: Mediastinal germ cell tumours (GCT) are a heterogenous group of benign and malignant neoplasms that have their origin from primitive germ cells left in the mediastinum during the prime of embryogenesis that have failed to migrate completely. Whilst GCT themselves remain rare, accounting to only about 2-4% of all cancers in children and adolescents below the age of 20, the Extra-gonadal germ cell tumours (EGGCT) represent about 5-10% of all germ cell tumours. The incidence of EGGCT is around 1 in 1,000,000 population and coincidentally, the anterior mediastinum is the most common site for EGGCT (50-70%).

Case presentation: Here we detail the unusual case of a young gentleman who presented with symptoms of chest pain mimicking angina that not resolve with treatment, which turned out to be a large anterior mediastinal EGGCT. An en-bloc resection was done and the patient was thereafter subjected to adjuvant chemotherapy.

Conclusion: GCT, being a rare entity coupled with an unusual presentation cause a delay in subsequent appropriate treatment which affects the prognosis of the patient.


# Background

Mediastinal germ cell tumours (GCT) are a heterogenous group of benign and malignant neoplasms that have their origin from primitive germ cells left in the mediastinum during the prime of embryogenesis that have failed to migrate completely. Whilst GCT themselves remain rare, accounting to only about 2–4% of all cancers in children and adolescents below the age of 20, the Extra-gonadal germ cell tumours (EGGCT) represent about 5–10% of all germ cell tumours. The incidence of EGGCT is around 1 in 1,000,000 population and coincidentally, the anterior mediastinum is the most common site for EGGCT (50–70%). Here we detail the unusual case of a young gentleman who presented with symptoms of chest pain mimicking angina that not resolve with treatment, which turned out to be a large anterior mediastinal EGGCT. An en-bloc resection was done and the patient was thereafter subjected to adjuvant chemotherapy.

# Case Presentation

A 35 year old gentleman whom hitherto have been well presented to a district hospital with complaints of chest pain that radiated to his left shoulder. He was admitted and treated for unstable angina where he received anticoagulant therapy and initiated on Dual Antiplatelet Therapy.

A week thereafter he had presented to a tertiary centre for persistent chest pain associated with non-productive cough for about a week. He also complains of loss of weight for about 1 year duration, however is unable to quantify objectively. Otherwise, he has no history of fever, loss of appetite, prolonged cough, haemoptysis or family history of malignancy.

In the emergency department, point of care ultrasound was done which revealed a large pleural effusion with fibrin over the left hemithorax. Pleural tapping was performed and 1 litre of haemoserous fluid was drained, which were exudative in nature. The patient also had several episodes of tachycardic rate uncontrolled atrial fibrillation. Whilst initially responding to Amiodarone, eventually he required cardioversion twice at 50J and 100J's respectively.

Once stabilised a left pigtail catheter was inserted under radiological guidance and drained 1.5 litres of haemorrhagic effusion.

At this point the patient was subjected to a computed tomography (CT) of his thorax which revealed the presence of a large heterogenous mildly enhancing hypodense mass at the anterior mediastinum measuring about 20cm x 9.4cm x 11cm. There was contrast extravasation visible which may be indicative of active intratumoral haemorrhage. The mass appears to abut the pericardium forming an indentation with pericardial effusion raising the suspicion of tumour infiltration. Features of the imaging was suggestive of an anterior mediastinal mass with high suspicion of active intratumoural haemorrhage. Possibility of GCT, Lymphoma and invasive thymoma were all considered however Tumour markers for AFP (Alpha fetoprotein) were elevated at 818.1 IU/mL narrowing it down to GCT.

At this point the patient was referred to our cardiothoracic centre for further management. At our centre the patient was counselled and planned for sternotomy and resection of the tumour. Intraoperatively, it was noted that the tumour was closely abutting the sternal bone which caused some bleeding upon the sternotomy. The large tumour measuring 10x20cm was densely adhered to the medial portion of left lung parenchyma and loosely adhered to the pericardium. The sheer mass had pushed the heart infero-laterally to the right, which was restored to its original position post tumour excision. En-bloc resection of the tumour together with the portion of tumour attached to lung parenchyma was resected and sent for histopathological examination.

The histopathological examination for the anterior mediastinal mass came back as mixed germ cell tumour. It was predominantly a immature teratoma, and a minor component of yolk-sac tumour. The resected let lung parenchyma showed positivity of malignancy as it was infiltrated by the yolk-sac component. Tumour markers showed an elevated AFP 818.1 IU/ml and BHCG 23.9 IU/L reading. The patient was planned for completion of staging via CT scan and started on chemotherapy with Bleomycin, Etoposide, and Platinum (BEP Regime) for 4 cycles.

# Discussion & Conclusion

Mediastinal GCT are divided into benign and malignant neoplasms that originate from the primitive germ cells that fail to migrate during embryogenesis. The most common site of primary EGGCT is the anterior mediastinum. AFP and BHCG may be elevated in cases of malignant tumours. However, primary testicular or ovarian germ cell tumour should be excluded as the anterior mediastinum also remains as a possible site of mestastasis of Gonadal GCT.

EGGCT predominantly affect young males. 30–40% of these tumours are seminomas, and the remaining is accounted by Non-seminomatous-GCT. These include yolk-sac tumours, embryonal carcinomas, choriocarcinomas, teratomas and non-teratomatous combined GCT. Besides the mediastinum (50–70%), other areas these tumours may be found are the retroperitoneum (30–40%), pineal gland (5%) and sacrococcygeal area (< 5%). The only known risk factor for EGGCT is Klinefellter Syndrome (47XXY), which is found to be associated with Non-seminomatous-GCT, characterized by their location on the midline from the pineal gland to the coccyx.

The classification system developed by the International Germ Cell Collaborative Group (IGCCG) categorizes GCT on the basis of its' histological type (seminomas have better prognosis than non-seminomas), localization of metastasis (retroperitoneal and testicular offer a better prognosis than mediastinal and intracranial), as well as the initial AFP, HCG and LDH levels. Patients receiving intensive chemotherapy have a 5-year survival rate of 40–65%. The survival rate of Seminomatous EGGCT range from 88–100% and survival of non-seminomatous EGGCT is only around 40–45%. The reason for the poor prognosis is due to the Mediastinal GCT not being as sensitive to the chemotherapy, and the bulky disease increases the risk of poorer outcomes due to issues such as respiratory and cardiac compromise.

GCT remains as a rare entity, and in their presence, the anterior mediastinum appears to be the most common site it's located in. Whilst the usual presentation may be chest pain, dyspnoea, cough and constitutional symptoms, it rarely presents as a cardiac event, mimicking angina, which is further complicated with intratumoral bleed causing pleural effusion. Generally, the symptoms vary based on the site and size of the tumour and if the tumour arises from non-vital organs, it can reach large sizes before becoming symptomatic. These unfortunately cause a delay in presentation, which affects the prognosis of the patient.

# Lists Of Abbreviations

| | |
|---|---|
| **GCT** | Mediastinal germ cell tumours |
| **EGGCT** | Extra-gonadal germ cell tumours |
| **AFP** | Alpha fetoprotein |
| **CT** | Computed Tomography |
| **BEP** | Bleomycin, Etoposide, and Platinum |
| **BHCG** | Beta Human Chorionic Gonadotropin |
| **IGCCG** | International Germ Cell Collaborative Group |
| **LDH** | Lactate dehydrogenase |

# Declarations

**Ethics Approval and Consent to Participate:**

Not Applicable

**Consent for Publication:**

Written consent to publish the information was obtained and available.

**Availability of data and materials:**

Not Applicable as this is a case report and not a study involving recruitment of patients.

**Competing Interests:**

The authors declare that they have no competing interests

**Funding:**

No funding to declare.

**Code Availability:**

Not Applicable.

**Author Contribution:**

**KK :** Author of the manuscript, operating on the patient, managing the patient before, and after surgery.

**AZ:** The primary operating surgeon, managed the patient during hospital stay, and on follow up. Corresponding author, and reviewed manuscript prior to submission.

**KM:** Assisted in the surgery, involved in graphics creation, and contributed to the pictures and editing of the manuscript.

**FZ:** Assisted in the surgery, co-managed the patient during hospital admission.

**AF:** Assisted in the surgery, co-managed the patient during hospital admission.

All the authors are involved in the clinical management of the patient from this case report.

All authors have read and approved the manuscript.

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# Figures

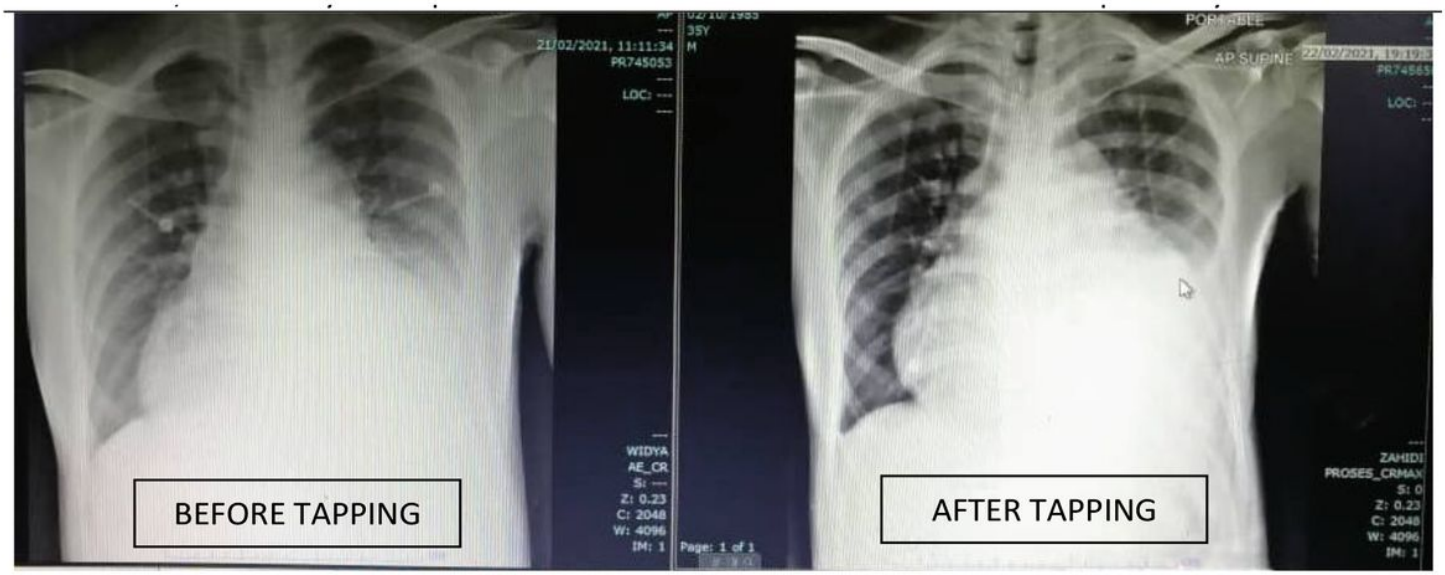
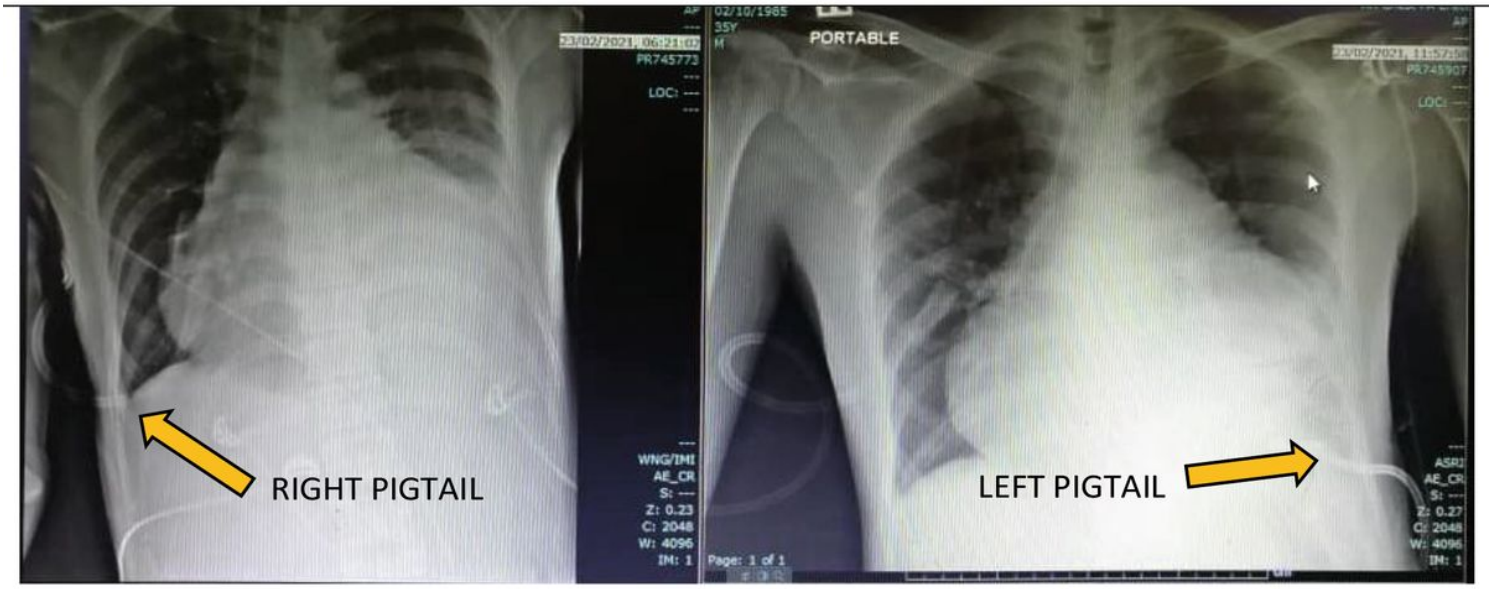


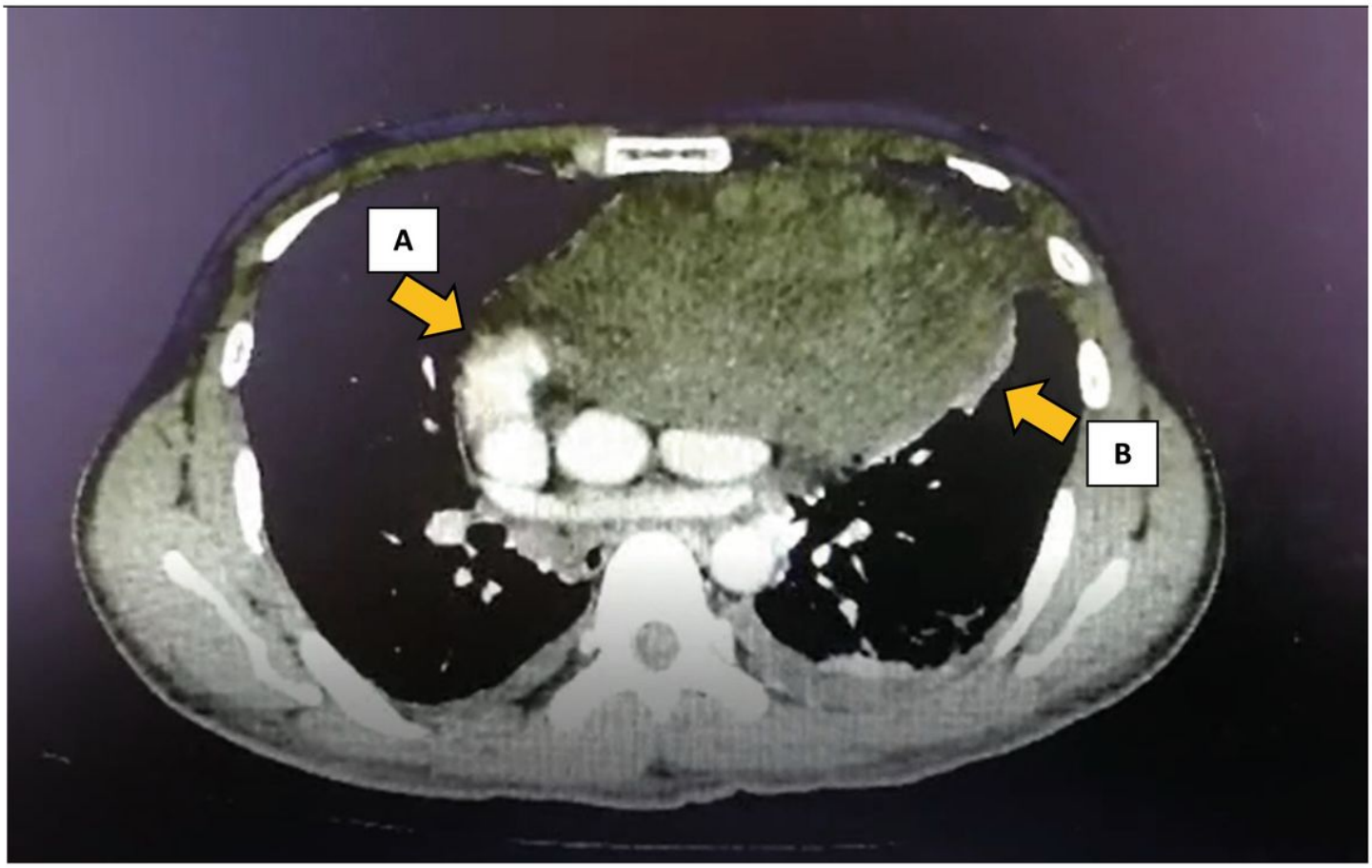

**Figure 1**

Chest Radiograph before (left) and after (right) the peritoneal tapping.




## Figure 2

Chest radiographs prior (left) and post (right) pleural pigtail insertion.




## Figure 3

CT Image showing the large heterogenous mass over the anterior mediastinum abutting great vessels.

A: Compressed great vessels by mass

B: Large heterogenous mass

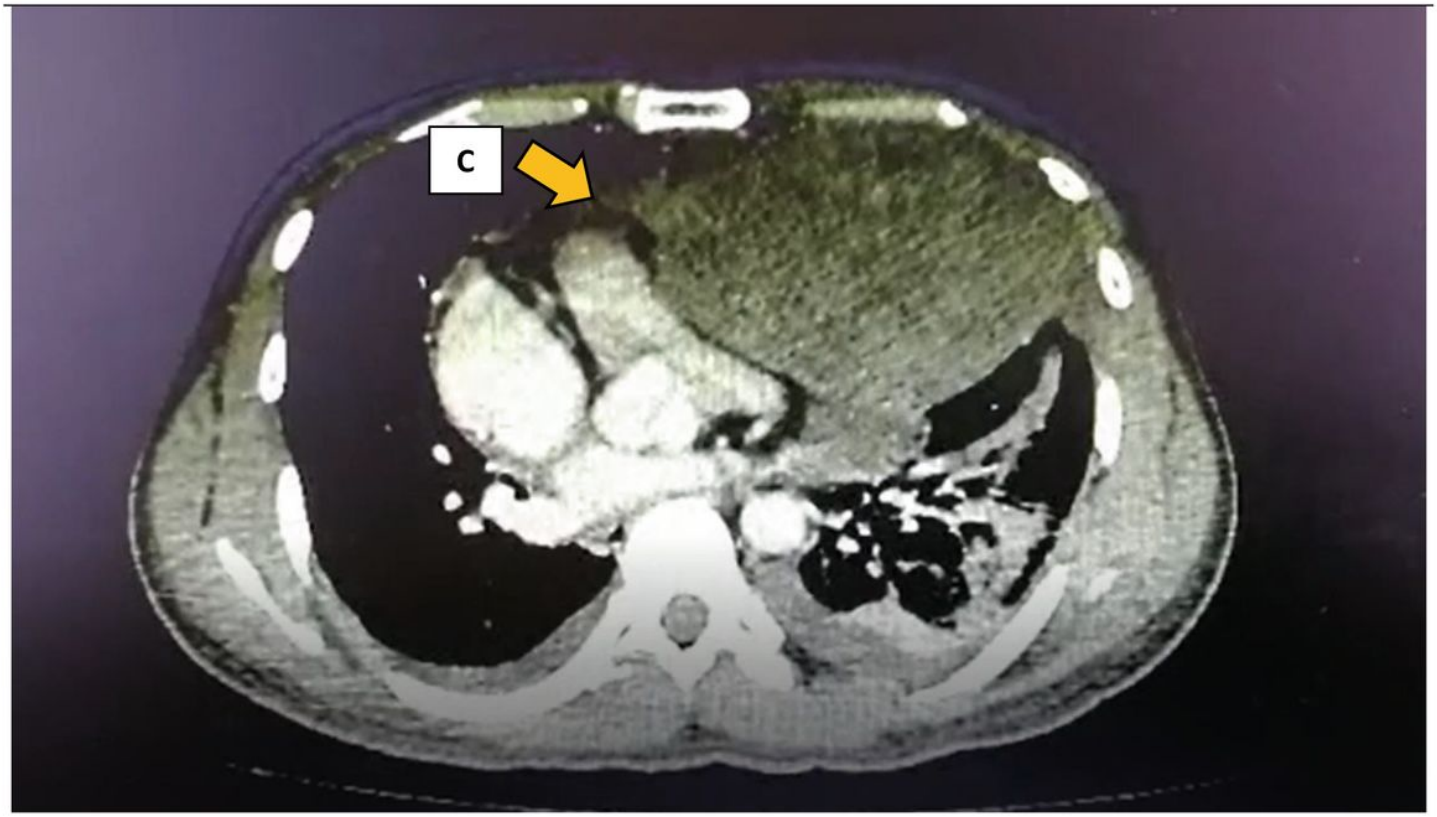
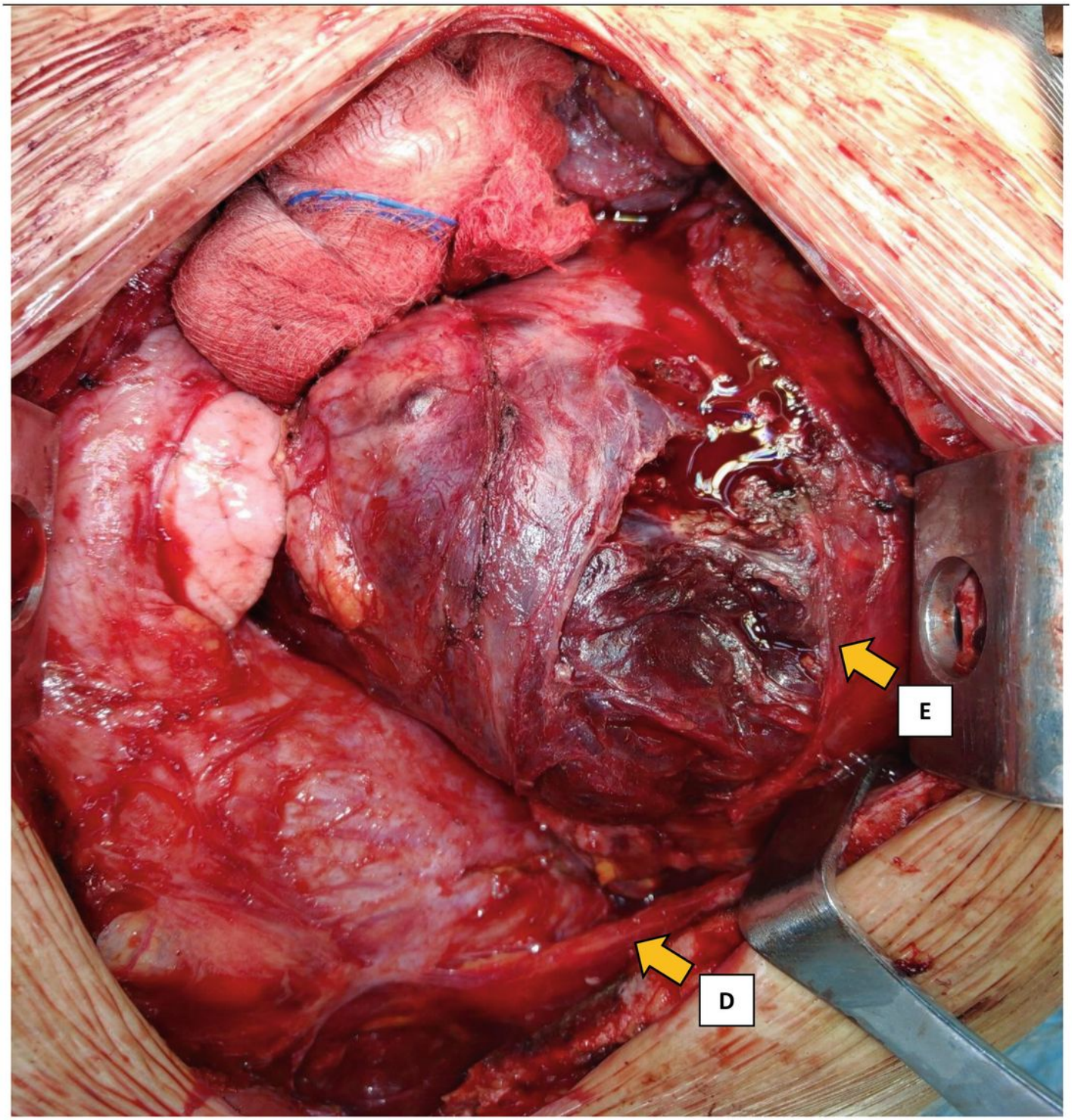



**Figure 4**

CT Image showing the large anterior mediastinal mass compressing the left chambers of the heart causing inferior displacement.

C: Compressed left heart chambers with inferior displacement

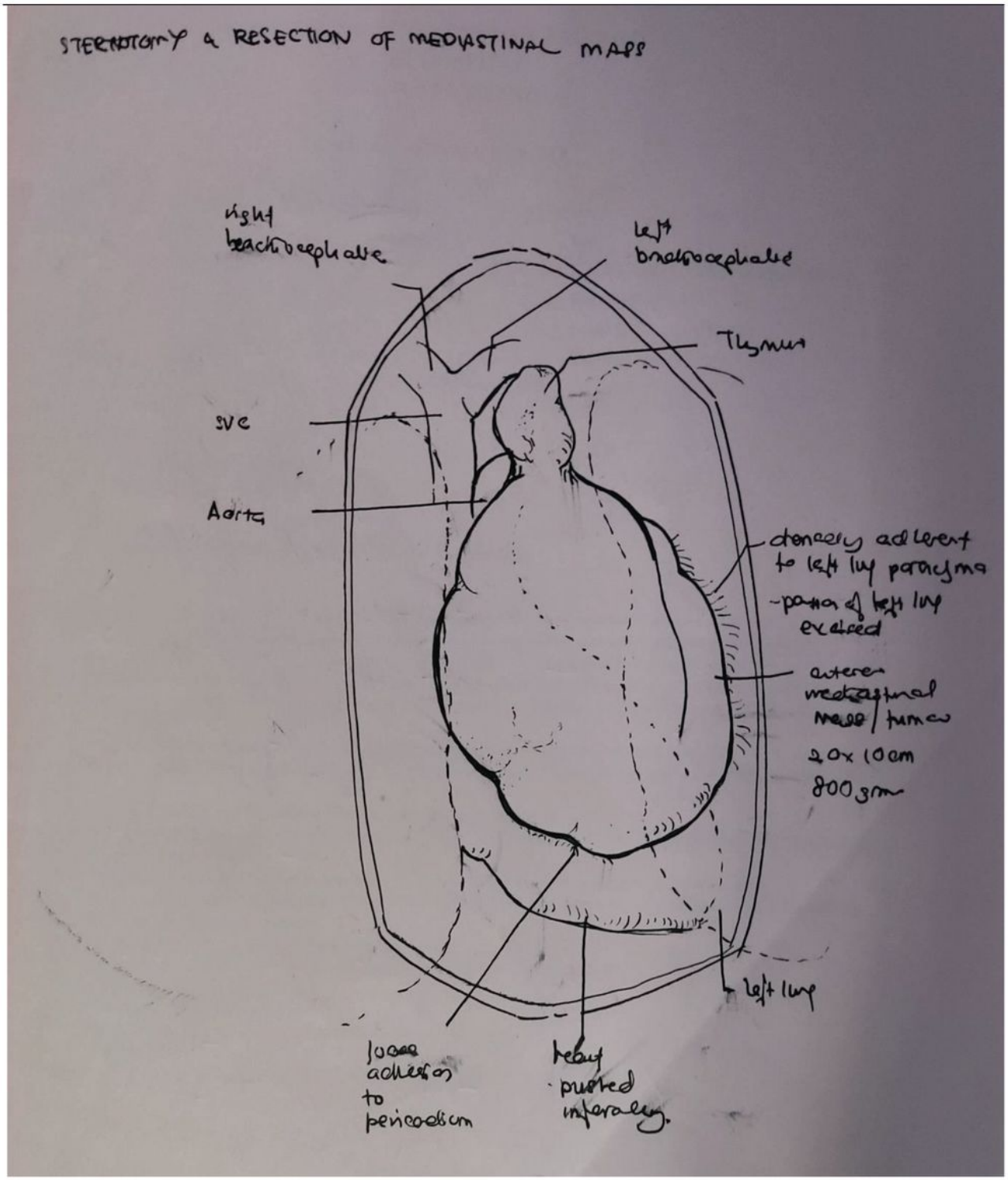



## Figure 5

Tumour depicted here with bleeding from torn part adhered to sternal wall and dislocating the heart infero-laterally to the right.

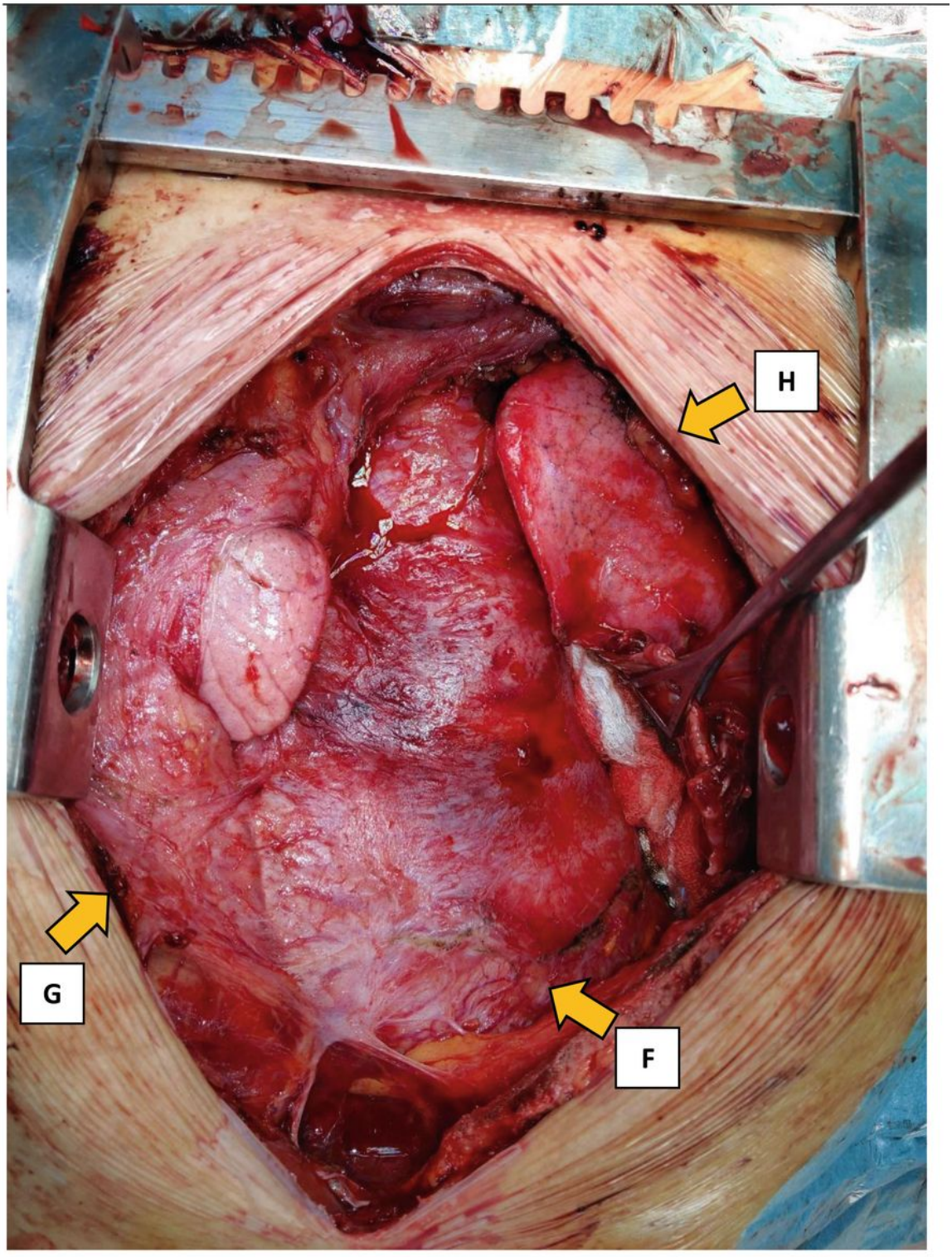
D: Heart displacement inferolaterally to the right.

E: Tumour with sternal wall adherence separated




## Figure 6

Intraoperative Findings of the Tumour Visualized




## Figure 7

Restoration of normal cardiac position post tumour resection.

F: Heart return to normal anatomical position

G: Right Lung, fully able to expand

H: Left Lung, fully able to expand

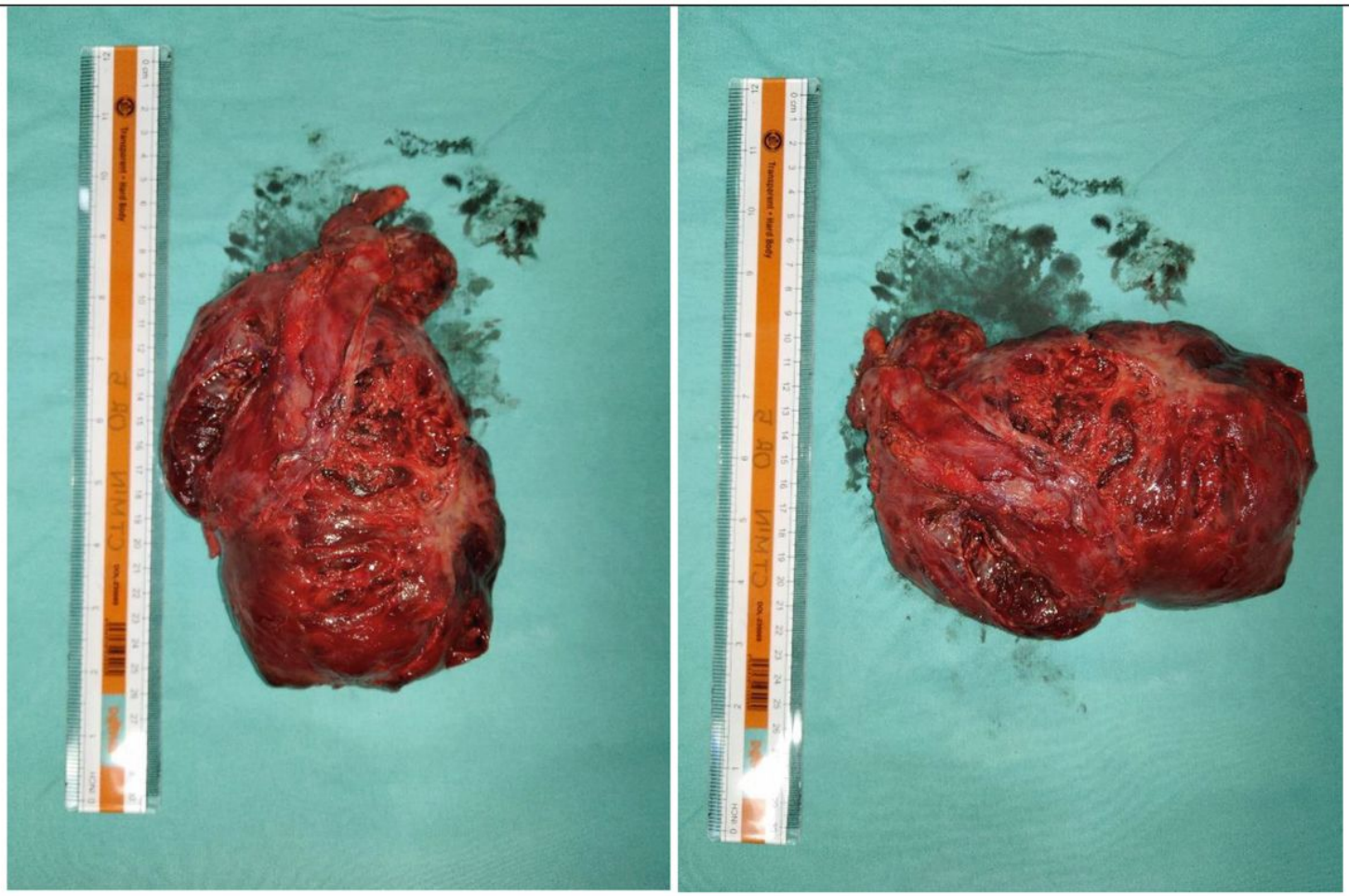


**Figure 8**

Tumour Specimen sent for Histopathological examination

## Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- CAREchecklistEnglish2013.pdf