

# A Rare Case of Situs Inversus Totalis (Mirror- Image) with Sudden Death

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## Abstract

Situs inversus(SI) is a rare congenital anomaly in which the visceral organs are placed reversely. This report describes the first ever case of SI with sudden death by cardiac tamponade. A 75 years old lady was admitted to the hospital with the history of fall onto the ground at home. Although resuscitation was done in emergency unit, hospital, she could not be revived and succumbed to death. The cause of death was shock due to flail chest, given by the medical officer in-charge. Autopsy revealed the situs inversus totalis and the cause of death was cardiac tamponade due to ruptured-myxoma.

**Key words:** *situs inversus, sudden death, ruptured-myxoma.*

## Introduction

Situs inversus (SI) is a congenital condition in which the major organs are reversed or mirrored image from their normal positions<sup>1</sup>. One literature expressed that Dextrocardia and Kartagener Syndrome is situs inversus as well and it is accompanied by sinusitis and bronchiectasis<sup>2</sup>. This report describes the rare case of sudden death by ruptured- myxoma with situs inversus totalis.

## Case report

An apparently healthy, 75-year-old lady was admitted to the hospital with the history of fall onto the ground, she died awhile after admission and the cause of death was given as “Shock due to the flail chest”.

## Autopsy findings

General appearance revealed an average built and

nourished female body without any marks of external injury. On opening the body cavities, the first unexpected finding was an appendix at left iliac fossa (figure 1). Liver was also situated on the left side of abdominal cavity while the spleen was present on the right side (figure 2). Heart was present on the right side of the chest cavity with apex pointing towards right side (figure 3). The block of thoracic and abdominal organs were removed in “ en-bloc”. Pericardium was incised in-situ and inspected the content, which revealed huge blood clot (hemopericardium), but the heart was seemed to be normal and no features of ruptured-myocardial infarct (figure 4) were seen. There was a remnant of ruptured-myxoma at the left atrio-ventricular groove which was the source of hemopericardium (figure 5). Other organs were unremarkable apart from the reversed sides of normal positions.

Histological examination of tumour showed abundant eosinophilic amorphous loose matrix containing stellate shaped myxoma cells. No features of malignancy are seen.

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Figure 1;

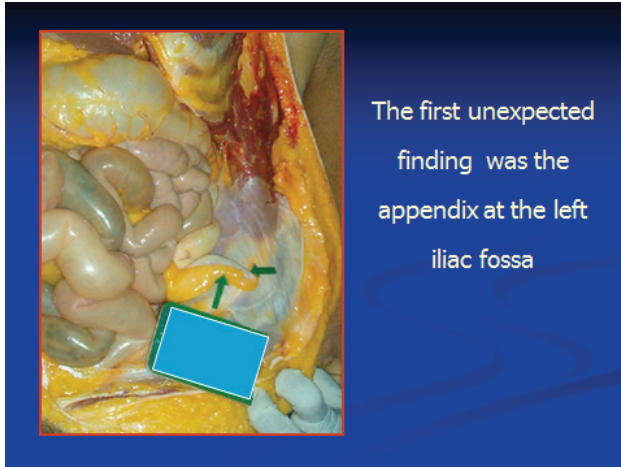


Figure 2;

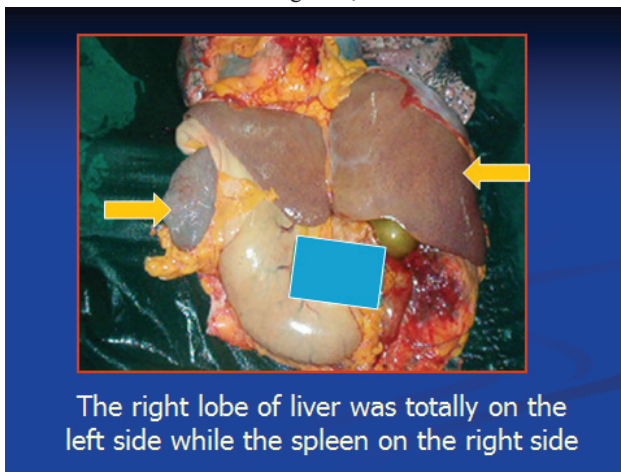


Figure 3;

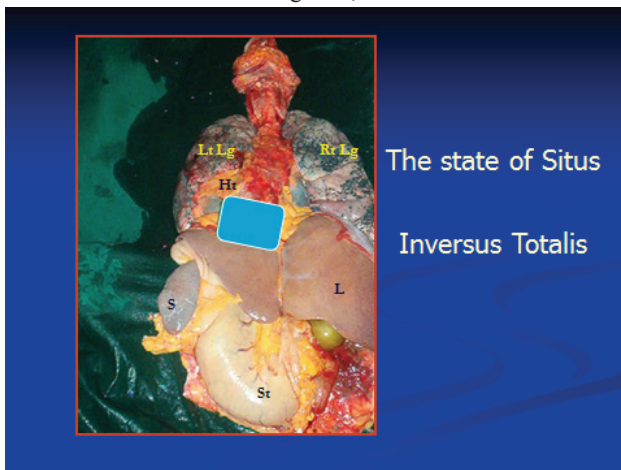


Figure 4;

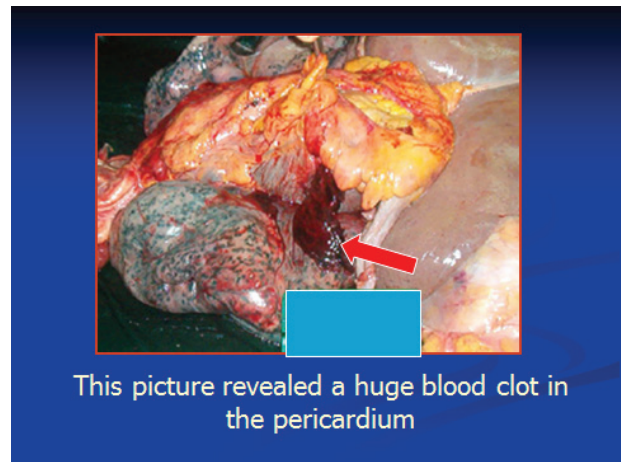
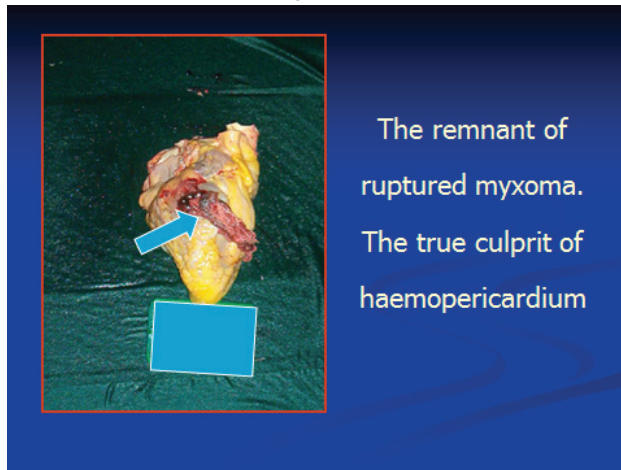


Figure 5;



## Discussion

The normal arrangement of internal organs is situs solitus while situs inversus is generally the mirror image of situs solitus<sup>1</sup>. Although cardiac problems are more common in people with situs inversus than in the general population, there is no symptom or complications of the condition until the advent of modern medicine it was usually undiagnosed<sup>1</sup>. The exact incidence of Situs Inversus is not known because persons remain asymptomatic; however, an incidence of 6-8 per 1000 live births is reported for congenital anomalies of heart<sup>2</sup>. Situs inversus totalis is a rare congenital anomaly in which general transposition of viscera while dextrocardia indicates the heart is mainly on the right and its apex points to the right<sup>3</sup>. Isolated dextrocardia usually displays situs solitus as stomach on left and liver on the right. There have been numerous reports describing the presence of rheumatic mitral stenosis with dextrocardia and/or situs inversus<sup>4</sup>, and some reports of Lutembacher's syndrome with dextrocardia<sup>4</sup>.

Cardiac myxoma, the primary cardiac tumours (PCT) are rare, accounting for 0.0017-0.03% in autopsy series<sup>5</sup>, in contrast to metastatic tumours of the heart which are 30 times more frequent<sup>4</sup>. Nearly 75% of PCT are benign and most often represented by a myxoma in 50% of cases in the adult population<sup>6,7,8</sup>.

Cardiac myxoma in dextrocardia with situs inversus is an uncommon condition. On review of literature, we could find one result, J Gabriel et al presented rare case of left atrial myxoma in dextrocardia with situs inversus totalis and the tumour was successfully excised with a trans-septal approach.<sup>4</sup>

Our case described the presence of cardiac myxoma with situs inversus totalis. The lady lived her whole life without any symptoms of situs inversus and it was incidentally found in autopsy as a rare finding. Our first impression was cardiac tamponade which was due to ruptured myocardial infarct as it was usual finding of sudden cardiac death. However, the ruptured myxoma was revealed when the heart was removed. The coronary arteries were free of occlusion and there were no features of recent or old myocardial infarct. The heart was grossly normal apart from the malposition as dextrocardia. There was the remnant of ruptured myxoma, 7x2 cm in size at the atrio-ventricular groove on the epicardium. The other internal organs were normal. Thus, with the presence of abovementioned findings, the cause of death was opined as cardiac tamponade due to ruptured myxoma, precipitated by the flail chest because of fall onto the ground.

### Conclusion

Our aim to of this report is to let the young generation of professional colleague knows that “things are not always as they seem at first glance” that the first impression might not always correct. It is unwise to give the cause of death before a complete and thorough post-mortem examination.

If any case of situs inversus is encountered at autopsy, the family should be counseled and advised to

undergo a possible screening as they are prone to have various cardiac abnormalities.

### Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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Ethical clearance

Necessary ethical approval was obtained from General Hospital Committee of Mandalay General Hospital, Myanmar.

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