SITUS INVERSUS vis-à-vis SUDDEN DEATH

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ABSTRACT

Situs Inversus, a rare congenital anatomical abnormality although is not directly related to Sudden Death. However, 3-5% of people with Situs Inversus have associated functional heart defect & 25% have an underlying condition i.e. Primary Ciliary Dyskinesis presenting as Kartagener’s Syndrome; are prone to Sudden Death.

Two cases of Situs Inversus on which medico-legal autopsy was conducted are reported. In first case, death was attributed to cardiac component (massive cardiomegaly, narrowing of coronaries and an old, healed apical scar) and in the second, due to respiratory complications (infection of lungs with areas of pus formation) with Blood Alcohol Concentration as 241.50 mg % ; being another significant finding. Apart from discussing the various aspects of the condition, vital medico-legal issues varying from 'Negligence' to 'Assessment for Insurance purpose' are being discussed.

Key Words: Situs Inversus, Sudden Cardiac Death, Primary Ciliary Dyskinesis, Medicolegal Issues.

INTRODUCTION

Situs Inversus is a rare congenital anomaly in which there occurs reversal placement of various thoracic and abdominal organs to their normal anatomical location. In Situs Inversus totalis, which is a recessively inherited condition, there, occurs mirror image of the anatomic location of all the viscera while in Dextrocardia only the apex of the heart points to right and is present in the right hemithorax. Sometimes, when Situs Inversus is accompanied by sinusitis and bronchiectasis, it constitutes Kartagener Syndrome[1]. In all these conditions, person is apparently normal and average life spans have been reported. However, owing to susceptibility of these individuals to catch various infections, it contributes to morbidity and mortality in a person having pre-existing cardiovascular or respiratory lesions. Thus, the individuals having these lesions are prone to sudden death.

Sudden Death can be defined as one, which is not preceded or only preceded for a short time by morbid symptoms[2]. The WHO defines it as someone who dies within 24 hours of appearance of symptoms while currently it is defined as "Unexpected Death occurring within one hour of new symptoms" [3]. The incidence of Sudden Death is about 10% of all causes of death and of these, almost half are due to cardiovascular causes and 15-23% is due to respiratory involvement[2,4]. Before the age of 35 years, the congenital and hereditary cardiovascular diseases are main causative factor for Sudden Death3.

Sudden Cardiac Death (SCD) is defined as "death as a result of cardiac cause within 6 hours of onset of symptoms[5]". In 5-10% of SCD, hearts are apparently normal. Cardiomegaly is a frequent finding in cases that died suddenly from cardiovascular pathology[3]. 3-5% of people with SITUS INVERSUS have associated functional heart defect & 25% have primary ciliary dyskinesis presenting as Kartagener Syndrome.

Two such cases of SITUS INVERSUS on whom medico-legal autopsy was conducted owing to suspicious circumstances are being reported and

Case Report No. 1

An apparently healthy, 56 years old, male allegedly died suddenly on 1st January. Earlier, about 4 months back, he met with a road traffic accident, treated and later developed loss of memory for which he was diagnosed as having
A middle-aged rickshaw puller was found dead under mysterious circumstances in his hut (jhuggi). Alleged history as given by nephew of the deceased was: the deceased, a known smoker & alcoholic, married but issueless; was seen alive last on 9th January, 1999 at about 9 P.M. He used to carry passengers to and from a lodge on his rickshaw and meet his nephew and other relatives in night after finishing this work. On the night of 10th January, he did not meet them. In the morning of 11th January, when his nephew went to see at his residence, he saw the door of jhuggi was not locked. As he pushed the door, it opened and he saw a dead body lying on cot with disfigured face.

**Autopsy findings**

Average built and nourished dead body of an old male without any marks of external injury. On opening the body cavities, right lung weighed 400 gms and had two lobes while left lung had three lobes and weighed 550 gms. Heart was present on right side of chest cavity with apex pointing towards right side i.e. dextrocardia (photograph No. 1 & 2). The heart weighed 740 gms and had measurements as 17x14x7.5 cms (Photograph No. 3). Stomach was present on right side and contained 50 cc of whitish fluid without any suspicious smell and having normal mucosa. Liver and caecum were present on left side of abdominal cavity while spleen was present on right side.

**Photograph 1:** Showing reverse anatomical placement of various viscera. Heart present on the right side and apex pointing towards right side. Liver is present on left side.

On histopathological examination of heart, right and left ventricular walls measured 0.3cm and 0.8 cm respectively. A grey white area of 0.5x0.4cm was found at apex of heart corresponding to old, healed myocardial infarct. Cut section of coronaries showed atheromatous plaque causing narrowing of lumen. (Right coronary artery - 50%, Left anterior descending - 50%, Left circumflex - 25-30%). Chronic venous congestion was found in both liver and lungs, while no pathological change was found in brain.

**Photograph 2:** Right lung showing two lobes instead of three. Left lung showing three lobes instead of two.

**Photograph 3:** Enlarged heart (cardiomegaly) weight 740 grams. Measuring 17 x 14 x 7.5 cms.

On 31st December and further advised CT scan. However, he died the next day. On further investigation, it was revealed that in the month of June, he was clinically diagnosed as a case of Situs Inversus with Dextrocardia with global hypokinesia, Left Ventricular systolic and Left Ventricular diastolic dysfunction with hypertension.

**Case Report No. 2**

A middle-aged rickshaw puller was found dead under mysterious circumstances in his hut (jhuggi). Alleged history as given by nephew of the deceased was: the deceased, a known smoker & alcoholic, married but issueless; was seen alive last on 9th January, 1999 at about 9 P.M. He used to carry passengers to and from a lodge on his rickshaw and meet his nephew and other relatives in night after finishing this work. On the night of 10th January, he did not meet them. In the morning of 11th January, when his nephew went to see at his residence, he saw the door of jhuggi was not locked. As he pushed the door, it opened and he saw a dead body lying on cot with disfigured face. With the help of clothing, body structure and configuration, he recognized the body as of his uncle. He informed other relatives and police and...
it was alleged that his uncle has been killed by his neighbour due to an old enmity. Also, he narrated an incidence of his uncle having a scuffle with the same neighbour 3-4 years back in which he suffered a fracture of right arm. Further, he correlated the statement of neighbour when that same neighbour threatened to eliminate his uncle, by killing and throwing against dogs. Autopsy could only be conducted after 3 days of recovery of dead body due to political intervention, owing to sensitive nature and constitution of 'Board of Doctors' for conducting postmortem examination.

**Autopsy Findings**

Average built and poorly nourished dead body of a middle aged male having put on a lot of clothing like shirt, jersey, pullover and covered with quilt and blanket. A 17x15 cm, U-shaped tissue defect with irregular margins involving front of neck and upper part of chest and without any vital reaction. Face was devoid of skin and underlying tissue, had crenated margins but without any evidence of antemortem hemorrhage (Photograph 4). Trachea absent, all fingernails showed bluish discoloration and body was in a state of advanced decomposition.

**Photograph 4: Tissue defect in form of crenated margins evident over forehead without any signs of antemortem hemorrhage.**

Entire body was subjected for radiological examination. Chest x-ray revealed the heart shadow and gas shadow of fundus of stomach on right side, but it was reported as normal by radiologist. The 'marker' for left side was not properly placed and in fact, it was placed almost in the middle (Roentgenogram). On opening the chest cavity, the heart was present on right side but apex pointed towards left. An anomalous placement of aorta in form of origin i.e. anterior to pulmonary trunk was observed. Left lung had three lobes while right lung had two lobes and on cut section, foci of pus were present in both lungs. All other abdominal viscera were placed opposite to their normal anatomical location. Toxicological examination revealed a Blood Alcohol Concentration of 241.50 mg% while autolytic changes were found on histopathological examination of various viscera.

**Photograph 5: X-ray chest AP view. Heart shadow on right side, gas shadow of stomach on right side. Note the presence of 'marker', which is almost in the middle.**

**DISCUSSION**

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. The incidence of Kartagener Syndrome is about 1:20,000. 3-5% of people with Situs Inversus have associated functional heart defect, which is higher than rate of heart defect in general, population. Although Situs Inversus alone does not increase the risk of cardiovascular disease but in cases with associated cardiac anomaly, it may lead to Sudden Death.

In case report I, the weight of heart, which was taken after removing, blood and clots from chambers, was 740 gms. According to Reddy, the persons having their heart weight in excess of 420 gms, are prone to Sudden Death[4]. Durigaon in
his study on 77 cases has stated that persons dying due to cardiovascular disease had a mean heart weight as 452gms[3]. There is a definite relationship between Body Mass i.e. both body height, weight, and heart mass and must be considered before labeling any heart as abnormal. Hitosugi et al has devised a simplified normal heart weight scale after taking into consideration Body Mass Index7. Kitzman et al devised formula for estimating heart mass from body mass based on sex and found it to be a better predictor of normal heart mass than either body surface area or body height8. Also the individuals with large heart weight are at risk for developing a dysrhythmia and hence die unexpectedly[8]. Heart weight in normal subject is also said to increase with age and physical activity, hypertension and alcoholism as well as cocaine abuse [8,9].

In the instant case, the body was of an average built and nourished old male, having a length of 5'3"; heart weight of 740 gms is definitely higher than normal expected weight. In addition, various coronary arteries showed 30-50% narrowing of their lumen, which collectively had their affect over the heart. More than 75% stenosis of at least one segment of a major epicardial coronary artery without any other cardiac lesion can cause sudden death[3]. In older adults i.e. age >45 yrs, 80-90% of sudden cardiac death have significant coronary artery disease while it is associated with 58-70% of sudden cardiac death in young adult population5. Congenital conditions comprise about 14% of structurally abnormal heart and next in occurrence to coronary artery disease i.e. about 65% cases[5]. Congenital and hereditary cardiovascular diseases are main etiologies of Sudden Death before age of 35 years[3]. Both obesity and epilepsy are co-morbid conditions in sudden cardiac death with structurally normal hearts.

No evidence of any gross or microscopic finding in brain was found which could substantiate the clinical diagnosis of subdural hemorrhage and supplemented the charge of Road Traffic Accident as cause for sudden death of this individual. Thus, due to presence of abovementioned findings with diagnosed hypertension during lifetime, it was opined that person was prone for Sudden Death and declared as a case of Sudden Cardiac Death.

In case Report 2, the external findings were declared as Postmortem Injuries. The presence of pus foci in lungs on gross examination favored the diagnosis of bronchiectasis although no histological comment was possible due to autolysis of viscera including lungs. As reported by Kartagener there occur Situs Inversus with chronic otitis media, chronic sinusitis and abnormalities leading to bronchiectasis due to loss of cilia in various cells leading to abnormal muco-ciliary clearance from middle ears, sinus cavities and airways. It is also seen in men with infertility secondary to sperm immobility due to same reason. Since blood alcohol concentration was 241.50 mg%, the person was prone to develop aspiration; also because of absence of ciliary movements in such a case. On further investigation, it was revealed that deceased was childless thereby favoring the diagnosis of Kartagener Syndrome. Thus in the absence of any antemortem traumatic pathology, taking due consideration of all findings; cause of death was opined as respiratory complications in presence of alcohol in a case of Situs Inversus/ Kartagener Syndrome.

CONCLUSION

If any case of Situs Inversus is encountered at autopsy, the family of deceased should be counseled and first-degree relatives may be advised by autopsy surgeon to undergo a possible screening, as they are prone to have various cardiac abnormalities[3].

If it is diagnosed during lifetime, as these persons are extra-susceptible to catch various respiratory infections; the dangers of smoking and exposure to industrial fumes should be explained by treating physician. An appropriate vocational guidance may also be supplied to them.

Failure to diagnose these cases by Physicians, Anesthetists and Radiologists may lead to charge of negligence against them[10]. The inattention to proper labeling of side i.e. Right or Left on X-ray films can lead to non-recognition or missed diagnosis of Situs Inversus. If the condition is missed by Anesthetist during pre-anesthetic check-up, surgical mishaps may result.

Situs Inversus with cardiac abnormality or respiratory complications decreases the life span and thus increases risk for sudden death. This
requires focusing while assessing these individuals for insurance purposes.

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References

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