ORIGINAL RESEARCH PAPER

Anaesthesiology

ANAESTHETIC MANAGEMENT OF EPIGASTRIC HERNIA REPAIR IN PATIENT WITH KARTAGENER'S SYNDROME–A CASE REPORT

KEY WORDS: Kartagener's syndrome, situs inversus totalis, spinal anesthesia, epigastric hernia,bronchiectasis.

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ABSTRACT

Kartagener's syndrome is a hereditary syndrome involving a combination of dextrocardia (situs inversus), bronchiectasis and sinusitis, transmitted as an autosomal recessive trait. Situs inversus is featured by the switching of the organs to the opposite side of the body. When associated with dextrocardia, it is known as situs inversus totalis. This disease is a uncommon congenital anomaly and can lead to a diagnostic misperception for surgeons because of infrequent location of symptoms resulting in late diagnosis of known pathologies and operative complications because of changed anatomy.The incidence rate of this syndrome is 1:30,000 live births. Here we present the anaesthetic management in a patient with Kartagener's syndrome posted for Epigastric hernia repair which we managed successfully under spinal anaesthesia.

Case Report –

A 31Year old male moderately build and nourished with history of Kartagener's syndrome presented to our institution with swelling in the epigastric region since 3 months which is insidious in onset, progressive in nature and associated with pain.The patient was diagnosed as epigastric hernia with history of recurrent respiratory tract infections since child hood and with primary infertility.On General examination patient had clubbing. Airway examination revealed adequate mouth opening with mallampati grade-1,spine was normal.His pulse rate was 86/min and blood pressure was 110/70 mmHg in right arm supine posture.His respiratory rate was16/min, thoraco abdominal type. Auscultation of his chest revealed bilateral normal vesicular breath sounds. On cardiovascular system examination, apex beat was located at right 5th intercostal space 1.5 cm medial to mid clavicular line. On auscultation, heart sounds were heard on the right side of the chest; no murmurs heard.His routine blood investigations along with serum electrolytes were within normal limits.Chest X-ray revealed fibrosis and bronchiectatic changes noted in left upper mid and lower zones and dextrocardia[Fig-1].Pulmonary function test showed moderate obstructive and restrictive pattern[Fig-2].His ECG showed marked right axis deviation with negative "p" wave in lead aVL and I with T wave inversion in V1 to V6 [Fig-3].Two-dimensional echocardiogram showed dextrocardia with no shunting with normal valves and chambers with ejection fraction of 60%.USG abdomen revealed situs inversus [Fig-4].

Patient was accepted for anesthesia with ASA grade II and was given nebulization with Duolin and Budecort night before and morning on the day of surgery,inj.cefixime 1gm I.V and inj.Hydrocortisone 100mg I.V morning on the day of surgery.patient was shifted to OT with 18G I.V cannula in the right arm. One point RL was connected.Monitors including pulse oximetry, ECG, and NIBP were connected.Under strict aseptic conditions, Lumbar puncture was performed at L3-L4 space with 23-G QBS needle and 3ml of 0.5% Hyperbaric Bupivacaine with 25mcg fentanyl was given.Adequate analgesia was obtained up to T4.Operative procedure lasted for about 1h and30min.Intraoperative urine output was 170 mL; 1,200 mL of crystalloids were used intraoperatively. Patient was hemodynamically stable throughout the procedure and was shifted to postoperative ward. Adviced analgesic-Inj.Diclofenac, nebulization,antibiotics,chest physiotherapy and spirometry postoperatively and his postoperative course was uneventful.

DISCUSSION

Kartagener's syndrome is a variant of the immotile cilia syndrome.[1] This has also been called primary ciliary dyskinesia and is a result of an autosomal recessive disorder of the microtubules of ciliated cells. Symptoms include male sterility, chronic or recurrent respiratory tract infection, and bronchiectasis because of the absence of mucociliary clearance. In 50% of patients situs inversus occurs and hence Kartagener's syndrome.

It may become possible to link the occurrence of abnormal ciliary function and abnormal position of the body organs. It seems that genes determine the structure,function, and time of appearance of proteins in the embryo that influence normal development and siting of the internal organs asymmetrically (situs solitus). On the left side of the embryo, near Hensen's node, a variety of proteins are secreted (Sonic Hedgehog, Nodal, Lefty and Pitx2). On the right side are others (Activin betaB, Snail and Fibroblast Growth factor-8).[2] These normally influence rotations, that lead to a left-sided heart and the usual position and structure of the lungs and abdominal organs. Abnormal genes and so abnormal proteins would lead to malpositions. Normal ciliary motion in the mouse node is anticlockwise and conceivably influences the flow of the proteins to their correct sites in the embryo. In the immotile cilia syndrome it is possible that abnormal distribution of the proteins occurs.

Dextrocardia with complete situs inversus occurs in approximately 2 per 10 000 births. The incidence of congenital heart disease is low being about 3%.Kartagener's syndrome will occur in about 20% of patients.[3]Thus, the incidence being approximately 1 in 50 000 births.In contrast, dextrocardia with situs solitus or situs ambiguus is less common (1 per 20 000 births) and the incidence of congenital heart disease is extremely high, probably 90% or greater. Dextrocardia with situs solitus usually, although not invariably, associates with severe complex cardiac abnormalities. They are most commonly transposition of the great arteries, double outlet right ventricle, ventricular septal defect, single ventricle and pulmonary stenosis or artresia.[3]

The anaesthetic implications of Kartagener's syndrome are varied. The anaesthetist might be involved with patients who have sinus surgery, pulmonary surgery, infertility investigations or possibly cardiac surgery. Of primary importance will be assessment of pulmonary and cardiac structure and function, and also prevention of pulmonary

complications in the bronchiectatic patient. The challenging aspect for anesthesiologist in Kartagener's syndrome is induction of anesthesia and intubation as they are more predisposed to airway problems and pulmonary infections. So, regional anaesthesia is preferred unless absolutely contraindicated. Physiotherapy, postural drainage, antibiotics, bronchodilators and incentive spirometry all have a role perioperatively..In thoracic surgery, the anatomy of the bronchi should be considered before selecting a double lumen tube. Knowledge of the position of the abdominal organs and of the branching pattern of the main stem bronchi is important in categorizing malpositions; and these must be borne in mind. Opposite direction application of the ECG electrodes is must as the altered surface electric polarity may result in pseudo-scenario of the perioperative ischemia. Lung separation in cardiothoracic surgery forms a difficult task owing to switching of the thoracic viscera.In scenarios of cardiac arrhythmias and cardiac arrest, one needs to be extra cautious in applying direct current with defibrillator pads on the right side. A fruitful revival of such patients warrants anesthesiologists and intensivists with a complete knowledge and skills. The aforementioned consequences and deliberations in a case of situs inversus totalis reveal without doubt that regional anesthesia is the ultimate option for any infraumbilical surgery when compared with the usage of general anesthesia, as long as there is no spinal defect.[4]

CONCLUSION

The accurate diagnosis of kartagener's syndrome with situs inversus totalis and a complete preoperative examination can reduce, greatly, the complications and several other potential challenges associated with its anesthetic management.

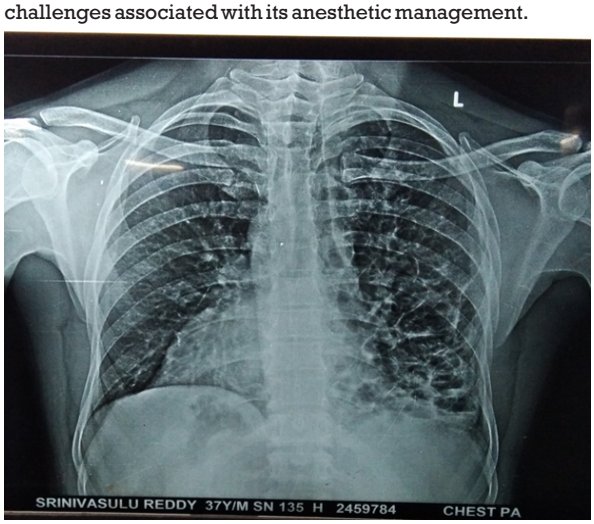

FIGURE-1 : CHEST X-RAY PA VIEW

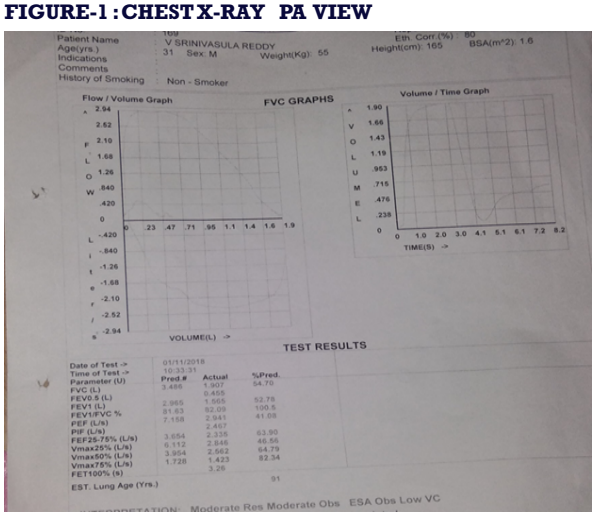

FIGURE -2 : PULMONARY FUNCTION TESTS

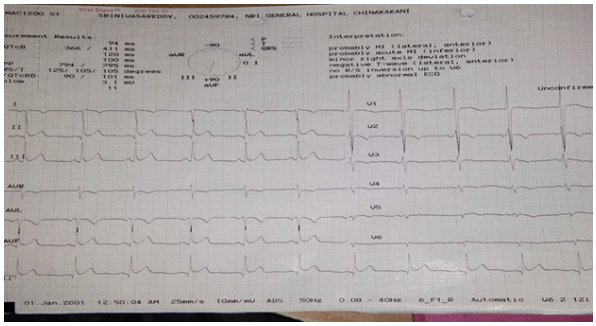

FIGURE-3 : ECG

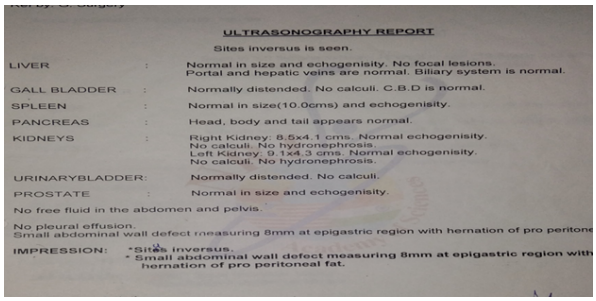

FIGURE-4 : USG ABDOMEN

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