CASE REPORT

KARTAGENER'S SYNDROME AND ACUTE APPENDICITIS

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Kartagener's syndrome is a combination of situs inversus totalis, sinusitis and bronchiectasis. Left sided appendicitis can present with problems in diagnosis and during surgery. We present here a case of Kartagener's syndrome and left sided appendicitis successfully managed in a peripheral hospital in Bahawalnagar, Pakistan.

Keywords: Kartagener's syndrome, Situs Inversus Totalis, Left sided appendicitis

INTRODUCTION

Situs Inversus Totalis is an uncommon genetically determined positional anomaly characterised by transposition of abdominal and thoracic viscera, i.e., the mirror image of normal.[1]

Kartegener's syndrome is a congenital autosomal recessive disorder that includes Situs Inversus Totalis, sinusitis and bronchiectasis. Anaesthetic concerns in a patient with Kartegener's syndrome include bronchiectasis, sinusitis and problems caused by cardiopulmonary inversion.[2,3]

Left sided appendicitis can occur in association with two congenital anomalies, i.e., Situs Inversus Totalis and intestinal malrotation. The abnormal location of appendix makes its diagnosis quite difficult.[4]

We present the case of a patient with Kartegener's syndrome and acute appendicitis who underwent appendectomy successfully.

CASE REPORT

A 24 year old housewife (wt. 48 kg, height 4 foot 10 inches) presented in surgical OPD of a peripheral hospital in Bahawalnagar, Pakistan. She had 2 days history of lower abdominal pain, nausea, vomiting along with low grade fever, malaise and anorexia. Abdominal examination revealed tenderness in both right and left iliac fossae. Rebound tenderness, guarding and rigidity were present only in left iliac fossa. There was no ascites and visceromegaly. Diagnosis of acute abdomen was made and later was confirmed as left sided acute appendicitis on abdominal ultrasound.

On pre-anaesthetic evaluation, her medical history included dextrocardia and chronic chest infections since childhood. She had a sister having identical complaints. On examination she her pulse rate was 104/min, BP 110/70 mmHg, Temp 100 ºF, and respiratory rate 18/min. On chest auscultation there were mild rhonchi and occasional bilateral crepitations. Her Haemoglobin was 12.8 g/dl and TLC 12.7×10^9/L. Ultrasonography abdomen and chest x-ray revealed situs inversus and dextrocardia respectively (Figure-1).

In the operation theatre, anaesthesia was induced with 1.5 mg/kg of propofol and atracurium 0.5 mg/kg was used for myorelaxation. Plain lignocaine 1 mg/kg was given intravenously and intubation was done in deep planes of anaesthesia. Anaesthesia was maintained with 1.5% isoflurane in 50% O_2 in N_2O. HME filter was used for humidification of inspired gases. Analgesia was obtained with 5 mg I/V morphine.

Incision was given at Mcburney's point in left iliac fossa. Inflammatory exudate was aspirated after opening the peritoneum and sample was sent for culture and sensitivity. Mesoappendix was ligated and cut and appendectomy was done. (Figure-2).

Local peritoneal toilet was done and wound was closed. At the end of operation, trachea was thoroughly suctioned and awake extubation was done. Patient had uneventful recovery and was discharged on 3rd post operative day. Stitches were removed on 9th post operative day.

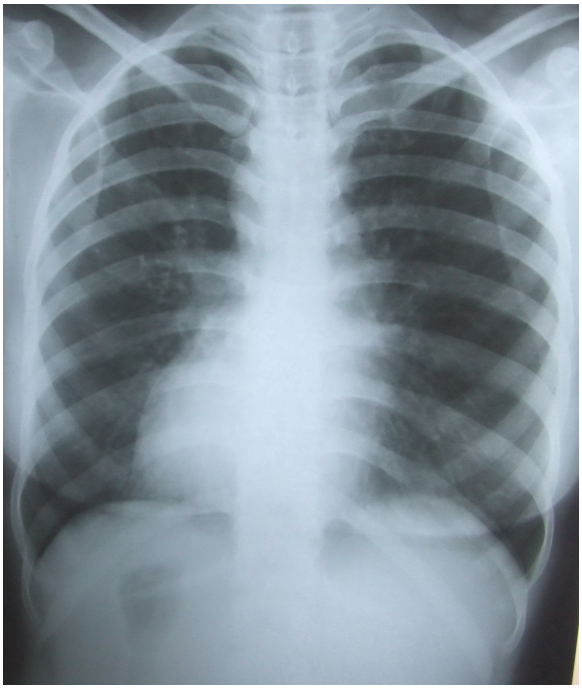

Figure-1: Dextrocardia showing on X-Ray Chest

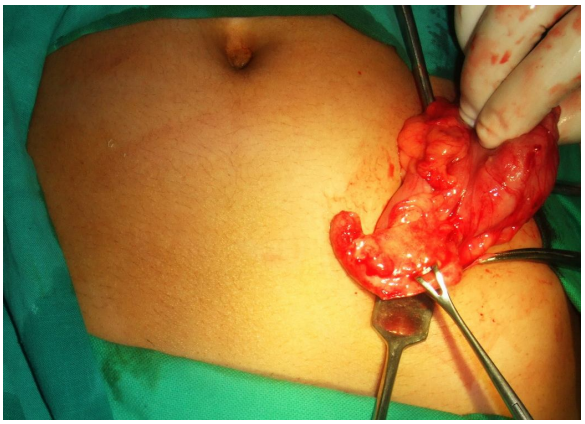

Figure-2: Left-sided appendix showing inflammation

DISCUSSION

Aristotle called visceral transposition, the punishment of gods, but now it is well established that it is an autosomal recessive disorder. It can coexist with other congenital anomalies, e.g., cardiovascular (Ventricular septal defect, Atrial septal defect, Tetrology of Fallot, Transposition of great arteries), respiratory (bronchiectasis, paranasal sinus deformity), digestive system (anal atresia, duodenal stenosis, absent appendix, megacolon) etc. When Situs Inversus Totalis, sinusitis and bronchiectasis appear together in a patient, it is called Kartegener's syndrome.[1]

In the anaesthetic management of such patients, following precautions can decrease the peri-operative complications:

- Pre-operative antibiotics and chest physiotherapy should be considered.
- Cough and respiratory depressants along with anti sialgogues should be avoided in pre-medication.
- Mainstem intubation can occur on left side and should be kept in mind while intubating the trachea. Nasal intubation and nasal airways should better be avoided. Frequent tracheal suctioning is often required.
- Inspired gases should be humidified.
- Parturients should have right sided uterine displacement.
- ECG electrodes and defibrillation pads should be placed in reverse manner.
- Regional anaesthesia is a preferred choice.[2]

In about 31% of patients with left sided acute appendicitis, first signs are pain and rebound tenderness in right iliac fossa. This is because, that although there is visceral transposition, it is without corresponding changes in nervous system. Therefore in 45% of cases, incisions at wrong sites have been given.[4]

Diagnosis of Situs Inversus Totalis is very important for surgeons because they have to take altogether different positions and work in a totally different fashion as whole of the anatomy is reversed. This different anatomical picture can lead to iatrogenic injuries.

With meticulous planning and care by the surgeon and anaesthetist, the Kartagener's syndrome can be successfully managed.

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