CASE REPORT

VASK Syndrome – Situs Inversus Totalis with Collapse of Anatomical Middle lobes of Both Lungs

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ABSTRACT

In our article, we present a unique case of situs inversus totalis (SIT)-associated collapse of anatomical middle lobes of both lungs, diagnosed on X-ray, and confirmed with computed tomography scan. The patients are generally asymptomatic and the diagnosis is incidental and easy to diagnose. These patients do not require any active management until associated with any other condition. Sometimes, this is seen associated with other syndrome like Kartagener syndrome. Such a case has never been published or reported in the literature. Hence, we would like to propose a new nomenclature: "VASK SYNDROME" based on the name of authors, that is, Vaibhav and Sushil Kachewar syndrome describing the unique finding of SIT coexistent with collapse of anatomical middle lobes of both lungs.

Key words: Collapse, inversus, situs, VASK syndrome

INTRODUCTION

Situs inversus (SI) is a rare positional anomaly in which there is complete transposition of abdominal and thoracic viscera. Situs abnormalities comprises group of congenital visceral and vascular anomalies with a varied radiological appearance.

Situs can be classified into three main groups:

1. Situs solitus: Organs of thorax and abdomen are present in normal configuration
2. SI: Complete inversion of the normal configuration
3. Situs ambiguus (heterotaxy): An intermediate configuration with duplication (isomerism).

In general, the patients are asymptomatic making it incidental findings on X-ray/ultrasonography, although definitive diagnosis is made by computed tomography scan which gives accurate anatomic location, relations of organs, cardiac apex, and vasculature.

CASE REPORT

A 26-year-old female presented with chronic cough for the past 6 months. The cough was dry in nature. There is no history of fever or weight loss and also no history ofcorrelation with variation of cough with change in season or position. Blood workup including complete blood count and erythrocyte sedimentation rate was within normal limit.

Chest X-ray frontal view [Figure 1] shows cardiac apex pointing towards right, that is, dextrocardia and fine reticular marking and fibrocavitary changes in the left perihilar region. Furthermore, prominent reticular markings were noted in the lingular region partially silhouetting the cardiac apex. The right-sided aortic arch was noted. The liver soft-tissue shadow was noted on the left while the stomach bubble was on the right side below the diaphragm.

As plain radiograph findings did not explain the patient's clinical presentation, plain high-resolution computed

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tomography thorax was carried out. It confirmed the chest X-ray findings and revealed dextrocardia [Figure 2] and complete transposition of the thoracic and abdominal organs [Figure 3]. Axial (thoracic level) & coronal Chest CT section shows collapse of the left middle lobe. The small left middle lobe bronchus is seen inside the collapsed lobe. The horizontal and longitudinal fissure outlines the collapsed left middle lobe [Figure 4a and b]. There were patchy opacities and underlying fibro-atelectatic and minimal bronchiectatic changes in left middle lobe. The lingula as well as the left middle lobe atelectasis was also seen [Figure 2].

Right sided aortic arch with transposition of all great vessels including IVC was demonstrated. Also there was evidence of tri-lobed left and bi-lobed right lung [Figure 5a-c].

On virtual bronchoscopy the right main bronchus showed 2 lobar bronchi and left main bronchus shows 3 lobar bronchi with collapsed middle lobe bronchus [Figure 6].

Final diagnosis was made as - A Case of Situs Inversus Totalis with Collapse of Anatomical Middle lobes of Both Lungs.

DISCUSSION

SI is an autosomal recessive genetic condition with a prevalence of 0.001–0.01%.[1] It is called SIT when there is a total transposition of abdominal and thoracic viscera (mirror image of normal visceral anatomy).[2]

During organogenesis, the primitive loops undergo fixed rotation which eventually leads to transposition of the

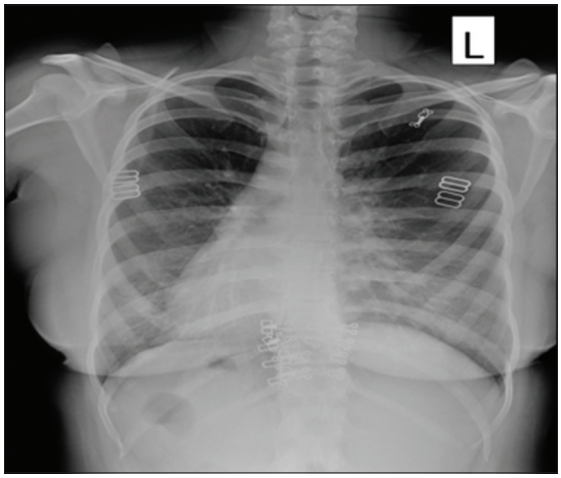

Figure 1: Chest X-ray posteroanterior view shows dextrocardia and fine reticular and fibrocavitary marking in the left perihilar region. The right-sided aortic arch is seen. Stomach bubble is noted on the right side below diaphragm, the left dome of diaphragm is higher as compared to right

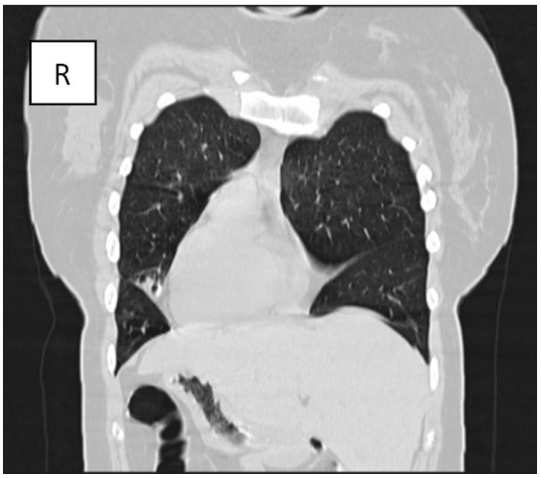

Figure 2: Coronal chest computed tomography scan images reveals cardiac apex toward the right side. Collapse of the lingular lobe is well seen. The liver soft-tissue density is noted on the left side with stomach and splenic flexure on the left side

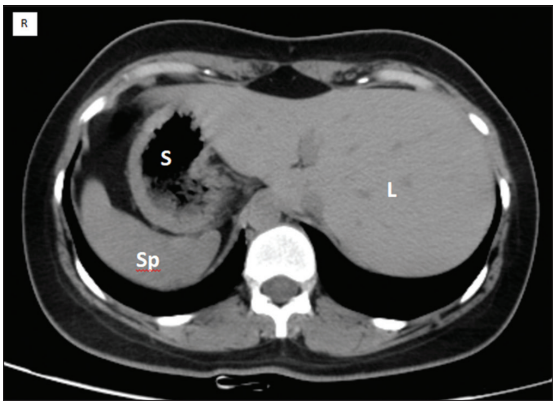

Figure 3: Axial computed tomography scan section at the abdominal level shows liver (L) on the left side and stomach (S) and spleen (Sp) on the right side. Aorta and inferior vena cava (IVC) also show total transposition, that is, the aorta is noted on the right and IVC on the left side, respectively. Neither asplenia nor polysplenia was noted

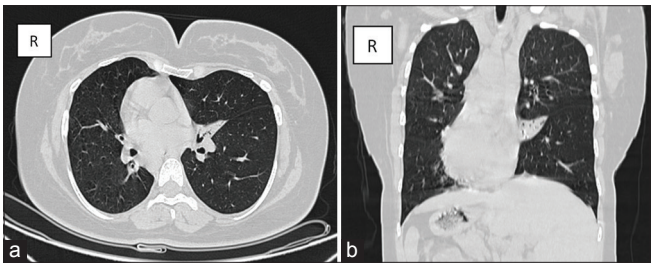

Figure 4: (a and b) Axial (thoracic level) and coronal chest computed tomography section shows collapse of the left middle lobe. The small left middle lobe bronchus is seen inside the collapsed lobe. The horizontal and longitudinal fissure outlines the collapsed left middle lobe

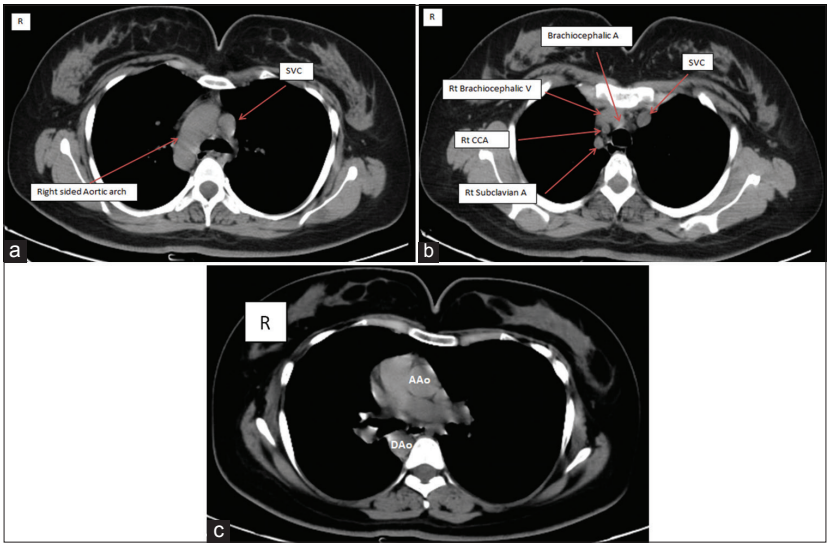

Figure 5: (a-c) Axial computed tomography scan sections at the thoracic and arch of aorta level shows right-sided aortic arch with the left-sided ascending aorta (AAo) and right-sided descending aorta (DAo). Furthermore, complete reversal of origin of great vessels is seen

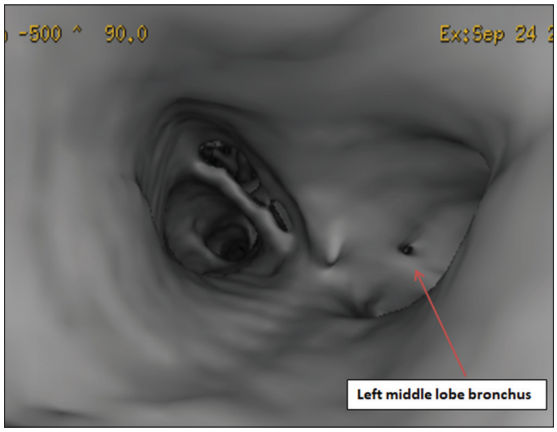

Figure 6: Virtual bronchoscopy image in the left main bronchus shows small collapsed left middle lobe bronchus

organs.[3] In SIT, the abdominal organs and lungs are transposed such that left and right lung is trilobed and bilobed, respectively. The liver, ileum, and ascending colon are on the left and stomach, jejunum, descending colon, and spleen are on the right side of midline.

In general, the patients are asymptomatic,[4] however, when it occurs in association with other conditions such as Kartagener syndrome (~ 20–25% cases) or primary ciliary dyskinesia, patients present with associated symptoms.[5]

Usually, these patients do not require any specific management or treatment; however, diagnosis is crucial before any thoracic or abdominal procedures or before surgical intervention for other causes.

This is a unique case where SIT is associated with collapse of anatomical middle lobes of both lungs. As bronchial buds of these collapsed lobes are poorly developed (demonstrated on virtual bronchoscopy), lobar collapse is likely congenital. Rest of the lungs showing proportionate compensatory hypertrophy.

Differential diagnosis list is as follows:

- Kartagener syndrome
- SI with dextrocardia
- Heterotaxy syndrome.

CONCLUSION

This was a case of SIT with collapse of anatomical middle lobes of both lungs.

To the best of our knowledge, such a case has never been published or reported in the available worldwide literature. Sarkar *et al*.'s "SI with unilateral pulmonary hypoplasia"[6] and Abdullah Simsek's "SI with Pulmonary Atelectasis"[7] are the only two cases close to our case.

Hence, the authors would like to propose a new nomenclature: "VASK SYNDROME" based on the name of authors, that is, VASK syndrome describing the unique finding of SIT coexistent with collapse of anatomical middle lobes of both lungs.

Such naming is permissible and is in better tradition of scientific nomenclature.[8]

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