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Case Report

Scimitar syndrome – An incidental diagnosis in a case of fibroadenoma ☆

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ABSTRACT

Scimitar syndrome is a rare congenital anomaly which is characterised by anomalous pulmonary venous drainage of the either entire right lung or part of it into the inferior vena cava or portal vein or hepatic vein or right atrium occasionally. This can be associated with hypoplasia of the right lung, dextroposition, underdevelopment of right pulmonary artery and anomalous systemic arterial supply from the descending aorta to the hypoplastic lung. A 36 year old female came with history of swelling in the right breast which turned up to be right breast fibroadenoma. Routine chest radiograph revealed scimitar syndrome which was confirmed on CECT chest.

Case report

A 36 year old female came with history of swelling in the right breast. Ultrasonographic findings revealed fibroadenoma in the right breast (Fig. 1). Routine pre-anaesthetic check up of chest radiograph revealed curvilinear radiopaque density at the right mid zone extending into the right hemidiaphragm for which contrast CT chest was performed. CECT Chest revealed hypoplastic right lung with ipsilateral mild mediastinal shift and anomalous vein draining into the right middle and lower lobes into subphrenic inferior vena cava which confirmed Scimitar syndrome.However there are no aberrant systemic arterial supply from the aorta. No other associated congenital disease are seen in our case (Fig. 2).

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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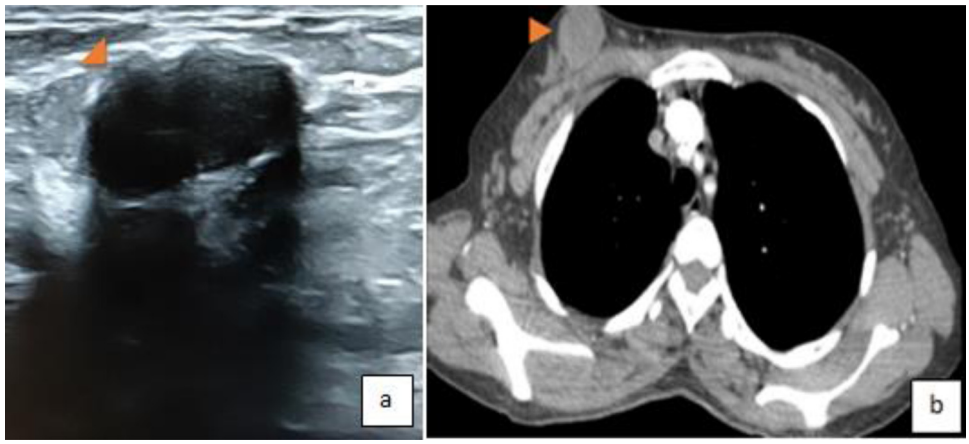

Fig. 1 – Thirty six year old female with lump in the right breast. Ultrasound image of right breast (A) shows well circumscribed wider than taller hypoechoic lesion (arrowhead) in the breast suggestive of fibroadenoma. CECT chest mediastinal window at the level of breasts (B) shows soft tissue density non-enhancing lesion (arrowhead) in the right breast suggestive of fibroadenoma.

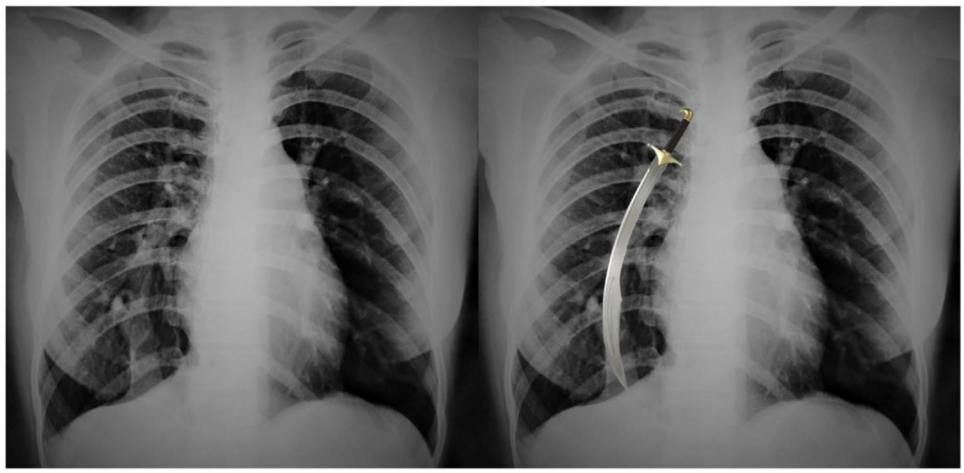

Fig. 2 – Thirty six year old female with lump in the right breast. Chest radiograph shows elongated radiopaque density in the right midzone extending into the right hemidiaphragm which represents Scimitar vein with graphic representation in the adjacent image.

Discussion

Scimitar syndrome is a congenital anomaly which accounts for 1–3/100,000 live births. It has female preponderance with 2:1 female to male incidence ratio [1]. This is otherwise known as 'Pulmonary venolobar syndrome' or 'Hypogenetic lung syndrome' or 'Epibronchial right pulmonary artery' or 'Mirror image lung syndrome'. It is characterised by anomalous pulmonary venous return of all or most of the right lung to the inferior vena cava just below or above the right hemidiaphragm or sometimes into the portal vein or hepatic vein or right atrium occasionally. There may be associated ipsilateral pulmonary hypoplasia, underdevelopment of right pulmonary artery and anomalous systemic arterial supply from the descending aorta to the hypoplastic lung [2–5].

The mean age of diagnosis is 7 months. This malformation is classically divided into infants and adult forms [6]. Clinical presentation vary from asymptomatic individuals to severe cases. Severe cases present in infancy with dyspnea, cyanosis, respiratory distress and cardiac failure. Mild cases present in the adulthood with recurrent respiratory tract infections.There can be other associations including congenital heart disease, congenital cystic lung disease, congenital diaphragmatic hernias and vertebral anomalies.

Chest radiography reveals an elongated radiopaque density in the right midzone extending into the right hemidiaphragm. This radiopaque density represents 'Scimitar vein'. Scimitar vein is the anomalous pulmonary venous drainage most commonly from the right middle and lower lobes into the inferior vena cava. This resembles the silhouette of Turkish sword known as shamshir [7].

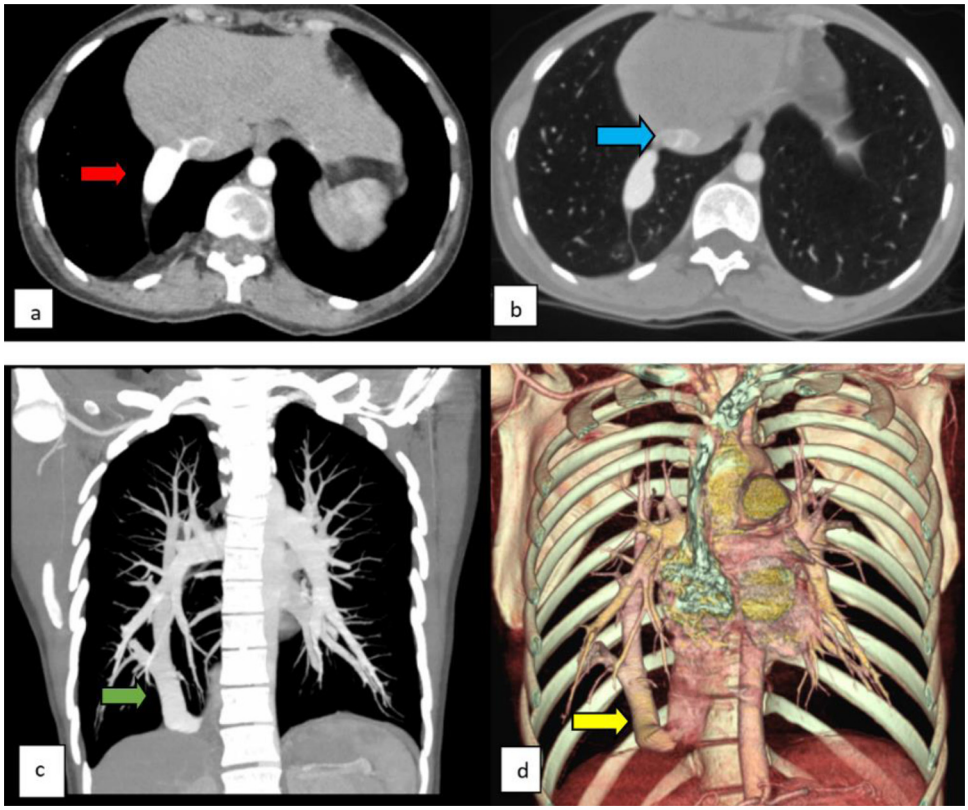

Fig. 3 – Thirty six year old female with lump in the right breast. CT angiography axial images shows scimitar vein (A) draining into the inferior vein cava (B) supplying the right lower lobe. Coronal reformatted image of CT angiography (C) and VRT image (D) shows anomalous vein draining right middle and lower lobes.

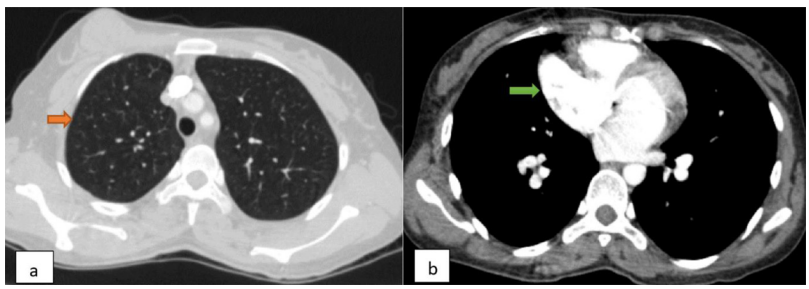

Fig. 4 – Thirty six year old female with lump in the right breast. CECT chest shows decrease in the antero-posterior diameter of the right lung (orange arrow in A) without any respiratory symptoms suggestive of mild hypoplasia. CECT chest mediastinal window shows mild midline shift of mediastinum toward right side (green arrow in B).

CT angiography is the investigation of choice [8,9]. Contrast images shows anatomic variations and vascular pathologies (Figs. 3 and 4).

In our case, Scimitar vein is associated with right pulmonary hypoplasia and dextroposition of heart with mediastinal shift to right (Fig. 4).

Conclusion

Scimitar syndrome can occur undiagnosed and have good prognosis.

Scimitar syndrome should be differentiated from other scimitar variants.

Patient consent

Patient consent form has been obtained.

The patient is aware that his data will be used in the publication of the case report.

Allowing the patient's information will not be charged additional costs in his treatment.

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