**Scimitar syndrome a rare variant of partial anomalous pulmonary venous connection presenting as recurrent hemoptysis**

**Sir,**

Partial anomalous pulmonary venous connection (PAPVC), also called as partial anomalous pulmonary venous return, includes a group of congenital cardiovascular disorders that are caused by the abnormal return of one or more, but not all of the pulmonary veins directly to the right atrium or indirectly through a systemic vein. PAPVC commonly associated with other structural anomaly of heart, most often an atrial septal defect (ASD), ventricular septal defects (VSD), and patent ductus arteriosus (PDA).[1] Risk of developing PAPVC is increased in the certain form of chromosomal disorder such as Turner syndrome.[2] The overall incidence of PAPVC is estimated between 0.5% and 0.7% in the population. In the common form of PAPVC, the left upper pulmonary vein connects to the left innominate vein, which in turn drains into the superior vena cava. Other forms of PAPVC include anomalous pulmonary venous connections to the coronary sinus, azygous vein, or the inferior vena cava (IVC). Scimitar syndrome is a rare variant, accounting about 3%–6% of all PAPVC (1–3 cases per 100,000 live births) cases in which part or even the entire right lung is drained by right pulmonary veins that connect anomalously to the IVC.[3] The term scimitar syndrome comes from the characteristic appearance of pulmonary vein on chest X-ray as inverted crescent-shape shadow. Here, we report a rare case of PAPVC of right upper lobe pulmonary vein draining into supra hepatic IVC without any associated cardiac anomaly.

A 15-years-old female patient presented to our hospital with a complaint of recurrent streaky haemoptysis for the last 2 months which was not associated with fever, loss of weight, and decrease appetite. There was no history of cyanosis, breathlessness, or recurrent pneumonia in the past. General physical and cardiorespiratory examination was unremarkable. Sputum smear examination for acid-fast bacilli and Genexpert test was negative. Tuberculin skin test shows no induration after 72 h. Chest X-ray posteroanterior view shows haziness over the right lower zone with ill-defined diaphragmatic border and prominent descending pulmonary vessel [Figure 1]. Electrocardiogram and two-dimensional echocardiography were within the normal limits. In view of ruling out vascular disorder, the patient was advised computed tomography (CT)-pulmonary angiography which revealed thick enhancing intraparenchymal venous channel likely replacing to superior pulmonary vein measuring 11 mm in diameter is noted in the right lung field extending from the right upper lobe to lower lobe then passes along the right dome of diaphragm and open into suprahepatic IVC, suggestive of PAPVC [Figure 2]. The rest of bilateral lung field was normal in attenuation and morphology without significant mediastinal/hilar adenopathy. Complete blood cell count revealed hemoglobin 9.0 gm%, total leukocyte count 9700 cell/mm³ (polymorphs 63%, lymphocytes 32% eosinophils 3%, and monocytes 2%), and erythrocyte sedimentation rate was 38 mm/h in 1st h.

Scimitar syndrome is a rare variant PAPVC in which a part or even the entire right lung is drained by right...
features of cardiopulmonary involvement making it an unusual manifestation of scimitar syndrome. Systemic circulation to the right lung base, which had higher pressure than the pulmonary circulation, was thought to be the most likely reason for the hemoptysis, but in our case, there was no aberrant arterial supply over the right lung base. Long-standing pulmonary venous stenosis and the development of venous collaterals between pulmonary and bronchial circulation leads to the formation of venous varices. This could be the cause of haemoptysis in our case. The definitive treatment of scimitar syndrome is surgical separation of the pulmonary venous system from the systemic venous system through redirection of anomalous pulmonary vein (s) into the left atrium and pneumonectomy.\textsuperscript{5}

As haemoptysis is rarely seen in scimitar syndrome, it can be easily misdiagnosed and leads to increase morbidity and mortality. This case demonstrates the importance of systemic approach for early evaluation and diagnosis of a rare cause of hemoptysis.

Informed consent
All appropriate consent from the patient was obtained.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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