Arch Gynecol Obstet (2014) 289:1371–1373
DOI 10.1007/s00404-014-3178-8

CASE REPORT

Scimitar syndrome and pregnancy, complicated with severe preeclampsia

Mehmet Aytac Yuksel · Metehan Imamoglu · Burcu Dincgez Cakmak · Mahmut Oncul · Riza Madazli

Received: 15 January 2014 / Accepted: 4 February 2014 / Published online: 19 February 2014

Abstract Scimitar syndrome (pulmonary venolobar syndrome) is a rare anomaly of venous return to the heart, most commonly consisting of partial or total anomalous pulmonary venous return from the right lung. This is the report of a case of a 29-year-old woman at 31 weeks of gestation of pregnancy who was previously diagnosed with scimitar syndrome. MR angiography and PET–CT results which were obtained before pregnancy demonstrated vascular malformation in the inferior part of the right lung. No specific treatment was planned throughout the pregnancy due to the absence of any symptoms. The patient's first physical examination was unremarkable except mild hypertension. In her follow-up, severe preeclampsia was developed and the patient had undergone a cesarean section of a live birth at 34 weeks and 2 days of gestation. This is the first case of scimitar syndrome with pregnancy in which the cardiac status of the patient deteriorated coincidentally due to the development of another manifestation such as severe preeclampsia besides the syndrome itself.

Keywords Scimitar syndrome · Pulmonary venolobar syndrome · Pregnancy · Preeclampsia

Introduction

Scimitar syndrome (pulmonary venolobar syndrome) is a rare, complex, and variable malformation with the abnormal pulmonary venous return of all or most of the right lung to the inferior vena cava at a location just below or above the right hemidiaphragm [1, 2]. It was first described in 1836 by George Cooper during an autopsy of a 10-month-old infant [3]. This rare anomaly has an incidence of approximately 1–3 per 1,00,000 live births, but its actual incidence is believed to be higher because of asymptomatic and non-diagnosed cases [4]. The term "Scimitar" is derived from the shadow created by the anomalous vein on the chest radiograph. This shadow extends from the lateral superior of the right lung to a relatively medial location and increases in caliber as it descends toward the cardiophrenic angle. The appearance closely resembles that of a curved Turkish sword or scimitar [4]. Here, we reported the clinical course of a pregnancy with Scimitar syndrome, complicated with severe preeclampsia.

Case

A 29-year-old woman at 31 weeks of gestation with a former diagnose of Scimitar syndrome was admitted to the antenatal care unit of a tertiary care/referral center for a routine visit. In her medical history, she reported no shortness of breath, paroxysmal nocturnal dyspnea, orthopnea, wheezing, fever, cough or any known cardiopulmonary symptoms. She had no history of syncope, lightheadedness or diaphoresis. Her family history was unremarkable and exclusively negative for cardiopulmonary anomalies.

On physical examination, she was in good general condition, pulse was 86/min and blood pressure was 140/90 mmHg. Her pulmonary examination was normal, and she had no cardiac murmur. Other system findings were unremarkable as well. Laboratory findings were as follows: hemoglobin 12.2 g/dl, hematocrit 35.4 %, White blood cell (WBC) 9,400/mm^3, platelets 228,000/mm^3.

M. A. Yuksel (✉) · M. Imamoglu · B. Dincgez Cakmak · M. Oncul · R. Madazli
Department of Obstetrics and Gynecology, Cerrahpasa School of Medicine, Istanbul University, Fatih, 34098 Istanbul, Turkey
e-mail: maytacyuksel@gmail.com

Other biochemical parameters were within normal limits. 12-lead electrocardiogram demonstrated normal sinus rhythm, normal PR, QRS, and QTc intervals, while *P* wave length was 3.5 mm in V1 lead indicating right atrial dilation. The chest radiography before pregnancy demonstrated a scimitar sign in the right hemithorax, obscuring the contours of the right atrium. Echocardiography revealed the dilatation of the right cardiac cavities and mild tricuspid and mitral regurgitation without any cardiac abnormality. PET-CT imaging before pregnancy demonstrated vascular malformation in the inferior part of the right lung (Figs. 1, 2). No specific treatment was planned throughout the pregnancy due to the absence of any symptoms. The optic fundoscopic examination was normal. In the obstetric ultrasonography, umbilical and uterine artery Doppler velocimetry findings were normal. The amniotic fluid index was between normal ranges. No protein was detected on urine dipstick test. However, the result of 24-h urine sample was 0.42 g. The patient was diagnosed with mild preeclampsia and follow-up of the patient was planned with routine weekly visits to antenatal care department. Cardiology and pneumology consultations were also scheduled.

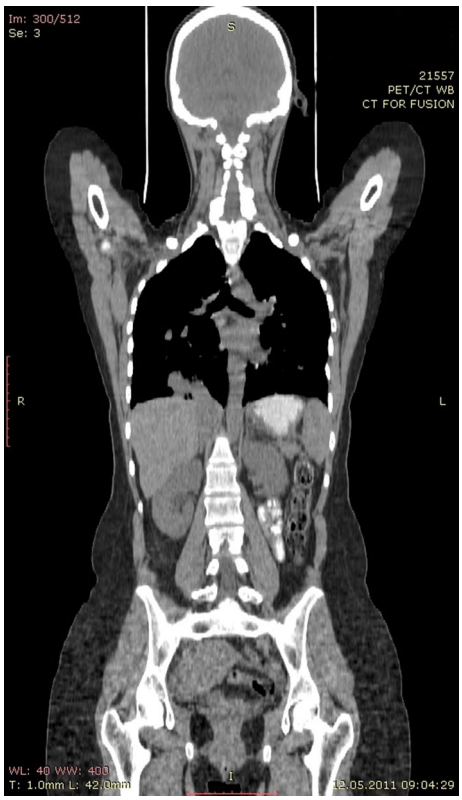

Fig. 1 Contrast-enhanced PET/CT images (maximal intensity projection, and 3D volumetric images) show an anomalous right lower lobe pulmonary vein descending vertically, draining the right lower lobe and entering the inferior vena cava

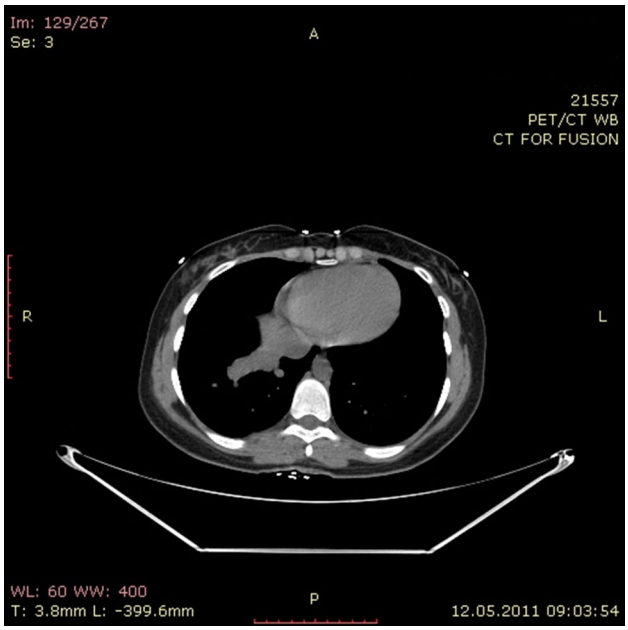

Fig. 2 Contrast-enhanced PET/CT images (maximal intensity projection, and 3D volumetric images) show an anomalous right lower lobe pulmonary vein descending vertically, draining the right lower lobe and entering the inferior vena cava

On her routine follow-up at 34 weeks of gestation, she complained about blurred vision and epigastric pain. Her blood pressure was 170/110 mmHg. Patient was diagnosed with severe preeclampsia and undergone a cesarean delivery of an healthy male neonate weighing 2,020*g* and APGAR scores of 8 at 1st min and 9 at 5th min. She had an uneventful postpartum period and was discharged at fifth day of delivery from the hospital.

Discussion

Scimitar syndrome is a rare malformation of the abnormal pulmonary venous return of all or most of the right lung to the inferior vena cava at a location just below or above the right hemidiaphragm, and this abnormal structure is occasionally called as the Scimitar vein [1]. It occurs more commonly in females and in the right lung [4]. In this case, the malformation was also in the right lung. This malformation is classically classified as infant and adult forms [5]. The syndrome can present itself in infancy/childhood or adulthood and bears a variety of presentation based on the age at which the diagnosis is made [5]. Clinical manifestations in infancy are more severe compared to those in adults [5]. The presence of Scimitar vein is essential for the diagnosis of this syndrome. Other anomalies are: abnormal right lung lobation and hypoplasia (almost 100 %, with widely varying degrees of hypoplasia); dextroposition of the heart;

hypoplasia of the right pulmonary artery (60 %); systemic arterial blood supply to the right lower lung from the infradiaphragmatic aorta (60 %); ASD of secundum type [6]. In this case, the diagnosis was established before the onset of pregnancy and Scimitar vein was detected solely. According to the degrees of pulmonary hypoplasia and pulmonary artery hypertension, about half of the patients with Scimitar syndrome are asymptomatic or mildly symptomatic when the diagnosis is made [1, 7]. In cases which have no significant left to right shunt and any concomitant cardiac and/or respiratory abnormality and have normal blood pressure, routine follow-up of the patient without any treatment is recommended [5].

As far as we could find, two cases of Scimitar syndrome with pregnancy were reported in the literature [8, 9]. First case was an adult form in which the pregnant woman was asymptomatic until 34 weeks of pregnancy. At 34 weeks of gestation, her condition deteriorated, heath failure was developed and the patient had to be delivered [8]. Other case was an infantile form in which two subsequent pregnancies of the patient were complicated with worsened respiratory functions in 36 and 33 weeks of gestation, respectively [9]. In our case, the patient was asymptomatic and obstetric follow-up without any treatment was recommended by cardiology and pneumology consultants. We suggest that in this case, the pregnancy was coincidentally complicated with severe preeclampsia at 34 weeks of gestation. All cases which were reported previously presented the complications of pregnancies due to the syndrome itself. This is the first case in which the pregnancy was complicated coincidentally with another manifestation such as severe preeclampsia besides the syndrome itself.

Conflict of interest No conflict of interest, financial or other, exists.

References

1. Khalilzadeh S, Hassanzad M, Khodayari AA (2009) Scimitar syndrome. Arch Iran Med 12:79–81
2. Ramirez-Marrero MA, de Mora-Martin M (2012) Scimitar syndrome in an asymptomatic adult: fortuitous diagnosis by imaging technique. Case Rep Vasc Med 2012:138541. doi:10.1155/2012/138541
3. Cooper G (1836) Case of malformation of the thoracic viscera consisting of imperfect development of the right lung and transposition of the heart. Lond Med Gazette 18:600–601
4. Rajaii-Khorasani A, Kahrom M, Mottaghi H, Kahrom H (2009) Scimitar syndrome: report of a case and its surgical management. Ann Saudi Med 29:50–52
5. Wang CC, Wu ET, Chen SJ, Lu F, Huang SC, Wang JK, Chang CI, Wu MH (2008) Scimitar syndrome: incidence, treatment, and prognosis. Eur J Pediatr 167:155–160. doi:10.1007/s00431-007-0441-z
6. Yehia BR, Bachmann JM, Traill TA (2010) Scimitar syndrome: a rare cause of dyspnea in adults. South Med J 103:578–580. doi:10.1097/SMJ.0b013e3181dfd59e
7. Takeda S, Imachi T, Arimitsu K, Minami M, Hayakawa M (1994) Two cases of scimitar variant. Chest 105:292–293
8. Miyake M, Katayama S, Masaki K, Kubo H, Hirakawa S, Saji T (2001) Scimitar syndrome and pregnancy: a case report. In: The Japanese teratology society abstract book, vol 41. p 243
9. Bell JD, Awonuga AO, Thompson ME, Ray CB, Devoe LD (2009) Infantile scimitar syndrome complicating pregnancy. Int J Gynaecol Obstet 106:259–260. doi:10.1016/j.ijgo.2009.03.054