Hemitruncus Arteriosus in a 10-Day-Old Neonate with Patent Ductus Arteriosus and Thrombocytopenia

Abstract: Hemitruncus arteriosus refers to an uncommon congenital cardiovascular abnormality. It usually presents in infancy and leads to the development of pulmonary hypertension, heart failure, and in severe cases maybe death. Herein, we reported hemitruncus arteriosus in a 10-day-old neonate with respiratory distress, patent ductus arteriosus, and thrombocytopenia.

Keywords: pulmonary artery, cardiac anomaly, Hemitruncus arteriosus, patent ductus arteriosus, thrombocytopenia

Introduction
In 1868, Fraenkel described an abnormal origin of one pulmonary artery in a 25-year-old woman as hemitruncus arteriosus for the first time. Hemitruncus arteriosus (HA) is an uncommon congenital cardiovascular abnormality which refers to the abnormal originating of one branch of the pulmonary artery from the aorta with the other branch arising normally from the right ventricle. Mostly the condition is diagnosed in infancy but rarely it may be undiagnosed until adulthood. Commonly, in most HA cases, the right pulmonary artery (RPA) originates from the ascending aorta next to the aortic valve. In most patients, the right pulmonary artery originates from the posterior side of the ascending aorta next to the aortic valve. In some patients, the left pulmonary artery originates from the ascending aorta, which is usually associated with the right aortic arch. The persistent flow of systemic high-pressure blood causes progressive diseases like pulmonary hypertension, and death in severe cases. The 3-month and first-year mortality rates of HA are 30% and 70% respectively which the condition can be corrected by early cardiac surgery.

Case Presentation
A 10-day-old male neonate with birth bodyweight of 3700 g, firstborn of non-relative parents in which the pregnancy was induced by intrauterine insemination (IUI) and the baby was born by cesarian section (C/S) with a gestational age of 40 weeks and good Apgar score was referred to the pediatric emergency department of Motahari hospital of Urmia. According to the mother’s statements, the chief complaints were shortness of breath, rapid breathing, and vomiting. Also, the patient had a history of NICU admission on 2nd day after birth because of...
dehydration and poor feeding for 1st day. The patient could not take enough oral feeding for the last 2 days and vomiting was progressive.

On physical examinations, the anthropometric measures were normal, the patient was non-dysmorphic and chest X-ray showed mild to moderate cardiomegaly with bilateral perihilar infiltrates. Vital signs were as follows; PR: 162/min, RR: 62/min, BT: 36.5°C, and oxygen saturation of 94% (room air). Mucous membranes were dry but skin turgor and fontanels were normal. The chest shape was normal and symmetric with subcostal retraction and tachypnea. Cardiac auscultation revealed a grade II/VI continuous murmur. Peripheral pulses were palpable. On abdominal examination, the liver span was increased on palpitation and generalized edema was obvious. Primitive neonatal reflexes such as Moro, sucking, and rooting were normal.

The urgent ABG evaluation revealed PH of 7.14, PaCO2 of 29 mmHg, HCO3 of 9 mEq/L, and PaO2 of 43 mmHg. Serum electrolytes, BUN, creatinine, serum albumin, and blood sugar were in normal ranges. Primary complete blood count (CBC) and coagulation tests were normal except for the platelet count and INR. The patient had thrombocytopenia (platelet count of 95×10^3/mm^3) and an INR of 1.19.

Due to severe pulmonary distress, the patient was intubated and admitted to NICU. After stabilizing the vital signs and hydration, a pediatric cardiology consultation was done. Echocardiography demonstrated no visible RPA, PDA with right to left flow, mild supravalvular aortic stenosis, mild aortic insufficiency, a severe reversal flow in aortic arch, secundum type ASD, dilated right atrium, right ventricle, and main pulmonary artery, severe tricuspid regurgitation and severe pulmonary hypertension, moderate mitral regurgitation and pulmonary insufficiency, dilated inferior vena cava, mild bilateral pleural effusion, and severe ascites. Due to the findings, close cardiological follow up and CT-angiography was performed.

Multi-slice computed tomography of the heart and major vasculature with special reconstructed views (dynamic and delayed images with contrast medium) revealed; anomalous arising of RPA from ascending aorta (hemitruncus), PDA 3.5 mm in size, stretched PFO (6 mm), atelectasis and congestion in both lungs with pleural effusion (Figure 1).

Serious efforts to perform cardiac surgery in addition to medical therapy such as inotropes and diuretics began. However, pediatric cardiac surgery was not available in the health care center or nearby cities. So, the patient was maintained on medical therapy and supportive care until cardiac surgery was a possibility.

Gradually, the patient’s respiratory distress increased and liver function tests were more impaired and eventually cardiopulmonary arrest occurred on the 24th day of admission in the NICU. Immediate CPR was begun by cardiac massage, intravenous epinephrine, and positive pressure ventilation. After 30 mins the patient had no vital response and CPR was stopped. The last blood tests revealed metabolic and respiratory acidosis. The last blood test values were in Table 1.

**Discussion**

Hemitruncus arteriosus is an unusual cardiac structural abnormality that, as Agati et al described, “results from failure of development of the left sixth arch and persistence of the left fifth arch”. In the disease, the pulmonary pressure increases with time and the mortality rate varies from 30% to 70% depending on case severity and receiving cardiac surgery as soon as possible.1–8 Early surgical repair can prevent pulmonary vascular disease progression.4 The surgical repair should be done within the first six months to prevent severe pulmonary vasculature disease. In our case, the severity of the disease, refractory acidosis, and unavailability of immediate surgery and coagulopathy and thrombocytopenia led to death.
The anomalous origin of the left pulmonary artery results from the failure of development of the left sixth arch and persistence of the left fifth arch. If left untreated, the pulmonary bed is vulnerable to early onset of pulmonary vascular obstructive disease owing to the large blood supply to both lungs. It is important to recognize this rare congenital cardiac anomaly early and contemplate surgical repair for a better prognosis.

Table 1 The Last Blood Tests of the Patient on 24th Day of NICU Admission

| Test     | Results | Unit   | Reference Value |
|----------|---------|--------|-----------------|
| PH       | 7.18    | –      | 7.35–7.45       |
| PsCO₂    | 51      | mmHg   | 35–45           |
| HCO₃     | 18.6    | mEq/L  | 22–26           |
| PaO₂     | 85      | mmHg   | 80–100          |
| BUN      | 16      | mg/dL  | 7–20            |
| Creatinine | 1   | mg/dL  | 0.5–1.3         |
| AST      | 41      | U/L    | 5–40            |
| ALT      | 7       | U/L    | 5–40            |
| Blood sugar | <20 | mg/dL  | 70–100          |
| Serum Na | 137     | mEq/L  | 135–148         |
| Serum K  | 3.6     | mEq/L  | 3.5–5.5         |
| Serum Ca | 10.1    | mg/dL  | 8.6–10.3        |
| WBC      | 7.2     | 10⁹/mm³| 4–12            |
| RBC      | 3.08    | 10⁹/mm³| 4.5–6.3         |
| Hb       | 9.8     | g/dL   | 14–18           |
| Hct      | 29.9    | %      | 39–52           |
| Platelet | 63      | 10¹¹/mm³| 140–440       |
| MCV      | 97.08   | fl     | 77–97           |
| MCH      | 31.82   | Pgm    | 26–32           |
| MCHC     | 32.78   | %      | 32–36           |
| PT       | 25      | Sec    | 10.1–12.9       |
| INR      | 3.2     | –      | 0.9–1           |
| PTT      | 39      | Sec    | 25.4–38.4       |

Conclusion

The anomalous origin of the left pulmonary artery terms results from the failure of development of the left sixth arch and persistence of the left fifth arch. If left untreated, the pulmonary bed is vulnerable to early onset of pulmonary vascular obstructive disease owing to the large blood supply to both lungs. It is important to recognize this rare congenital cardiac anomaly early and contemplate surgical repair for a better prognosis.

Ethics and Consent Statement

The patient’s parents had the opportunity not to accept the study. The patient’s personal information remained confidential. There was no cost or harm to the patient through the study. All of the stages of the study were conducted under supervision and confirmation of the ethical committee of Taleghani hospital and Urmia medical sciences university.

Disclosure

The authors report no conflicts of interest in this work.

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