Case Study

Pulmonary Hypertension Mimicking Acute Coronary Syndrome in Pediatric: A Rare Case Report

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Background: Pulmonary hypertension (PH) is a rare disease in children and chest pain as a presenting symptom is uncommon. Therefore, chest pain as presenting symptom of pulmonary hypertension in children makes the diagnosis challenging.

Aim: The aim of this case report is to highlight that pulmonary hypertension can be present similar to typical anginal pain mimic acute coronary syndrome and how can we establish diagnosis of pulmonary hypertension.

Case Description: We report a case, 14-years old boy who presented with typical chest pain as a main symptom. He had family history of primary pulmonary arterial hypertension. Laboratory examination at presentation, showed high level of troponin-T. Twelve leads electrocardiography (ECG) showed right ventricular strain pattern. Transthoracic echocardiogram showed dilatation of right atrium and right ventricle and computerized coronary tomographic angiography showed dilatation of pulmonary artery with no obstructive coronary artery disease.

Conclusion: Acute chest pain in pediatric population should be evaluated carefully, although it's uncommon symptom of pulmonary hypertension but must be evaluated carefully. Diagnosis of pulmonary hypertension in children can be done with ECG, echocardiography, and computerized tomographic angiography.

Keywords : Chest pain, hereditery, pulmonary hypertension, pediatric

INTRODUCTION

Pulmonary hypertension (PH) is an uncommon disease in children. This causes significant morbidity and mortality in the pediatric population. In Europe, incidence of Pulmonary hypertension (PH) was reported at 4-10 cases per million children per year with a prevalence of 20-40 cases per million while in USA it is 5-8 cases per million children per year and 25-33 cases per million children.[1] Survival rates of PH in pediatrics has improved significantly since the advancement of targeted PH therapies. From The Registry to Evaluate Early and Long-Term PAH (REVEAL) reported 1-, 3-, and 5-year estimated survival rates of 96 ± 4%, 84 ± 5%, and 74 ± 6% in each specific case.[2]

Pulmonary hypertension (PH) is a pathophysiological condition that defines as an increase in mean pulmonary arterial pressure (PAPm) ≥ 25 mmHg after 3 months of age which is assessed by right heart catherization (RHC).[3] It's still more challenging for clinicians, especially in remote areas where there are no advanced tools. PH is associated with various diseases like cardiac, pulmonary and systemic diseases that could happen at any age from infancy to adulthood. PH in pediatrics is divided into 5 groups, group 1 PAH especially heritable

pulmonary arterial hypertension (HPAH) has been identified in 20-30% of pediatric sporadic cases and 70-80% of familial PH cases, being very common form of PAH.[1]

Children with PH show non-spesific symptoms with initial symptoms induced by exercise. The most common presenting symptoms of PH in pediatric are dyspnea on exertion, fatigue and syncope and often misdiagnosed with general conditions such as asthma, laryngitis and angina.[2,3] In this case, chest pain is uncommon symptoms of PH, however the incidence of a cardiac-related cause for chest pain in the pediatric population is rare, ranging from 0.2% to 1 % of cases.[4] Therefore, It is a challenge to suspect and then establish the diagnosis of PH in pediatric.

A standardized approach to diagnostic testing has been recommended from World symposium pulmonary hypertension in 2018, although definite diagnosis of PH is done by cardiac catherization, the first diagnostic tool of suspected PH is commonly made with transthoracic echocardiogram.[5,6] Echocardiography not only shows cardiovascular anatomy but also helps in RV pressure elevation.

The aim of this case report is to highlight that pulmonary hypertension can be presented similar to typical angina in acute coronary syndrome and to show how the diagnosis of pulmonary hypertension is done.

CASE ILLUSTRATION

A 14 years old boy was admitted to emergency ward due to history of chest pain about 3 hours before admission. He got chest pain characteristically as heavy pressure around the left side spreading to his left arm, accompanied by vomiting and cold sweating. Chest pain increased progressively and did not reduce by rest. It was also accompanied by shortness of the breath. There was no history of persistent cough, fever, previous acute respiratory infection. Patient did not take any medicine before coming to hospital. Patient did not have any significant past medical problems like congenital heart disease, hypertension, coagulation disorder or metabolic disease such as diabetes melitus or dyslipidemia. He was born at term with normal birth weight. His father was diagnosed case of primary pulmonary hypertension and his sister died of pulmonary hypertension.

On physical examination, he was pink in air, fully alert, moderately ill with height of 158 cm and body weight of 48 kg. His blood pressure in right arm was 109/65 mmHg, and was same in all extremities, heart rate was 84 bpm with regular rythm, respiratory rate 22/minute, temperature 36,7^{o} celcius, and oxygen saturation was 96% at room air. Clinically there was no evidence of conjunctival hemorrhages, yellowish cholesterol-filled plaque on or around the eyelid and petechie.The jugular veins was not distended and measured as 5+2 cm. On cardiac examination, first heart sound was normal, but pulmonary component of second heart sound was very loud in pulmonary area, grade 2/6 pan-systolic murmur at left lower sternal border (LLSB) that increases with inspiration. Respiratory and rest of physical examination was unremarkable.

Laboratory examination at admission showed elevated serial troponin-T in 3 hours (ΔTroponin T = 752 pg/mL) with normal D-dimer value (195 ug/mL). Twelve lead electrocardiogram showed sinus rhythm with

heart rate 80 bpm, right axis deviation, right venticular hyperthropy (RVH) with RV strain pattern, right atrial (RA) enlargement (P wave amplitude > 2.5 mm in the inferior lead and P wave amplitude > 1.5 mm in V1 and V2, figure 1).

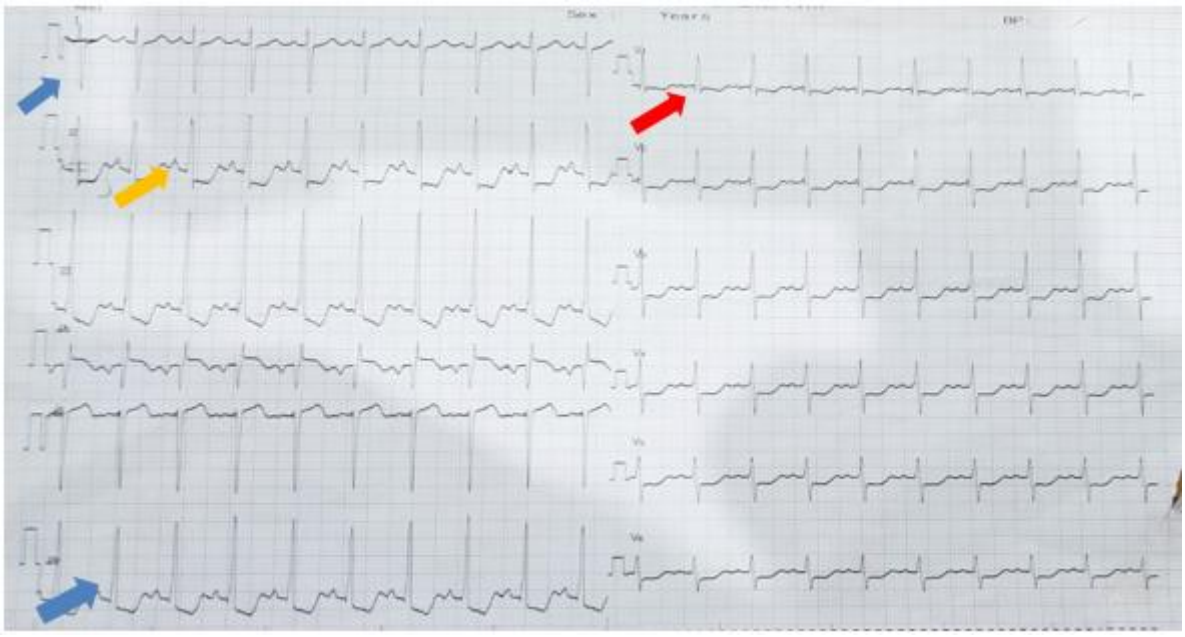

Figure 1. Electrocardiogram examination showed sinus rhythm with right axis deviation (blue arrow), right ventricular hypertrophy (RVH) with RV strain pattern (red arrow) and right atrial (RA) enlargement (yellow arrow).

Chest X-ray revealed cardiothoracic ratio 52%, increased PA segment (right descending pulmonary artery =27 mm (> 16 mm), elongated aortic segment, left descending pulmonary artery 33 mm (> 18 mm), convex/ bulging main pulmonary segment, right heart border enlargement (>44 mm from midline to the right heart border), flatten cardiac waist, and upward apex (Figure 2).

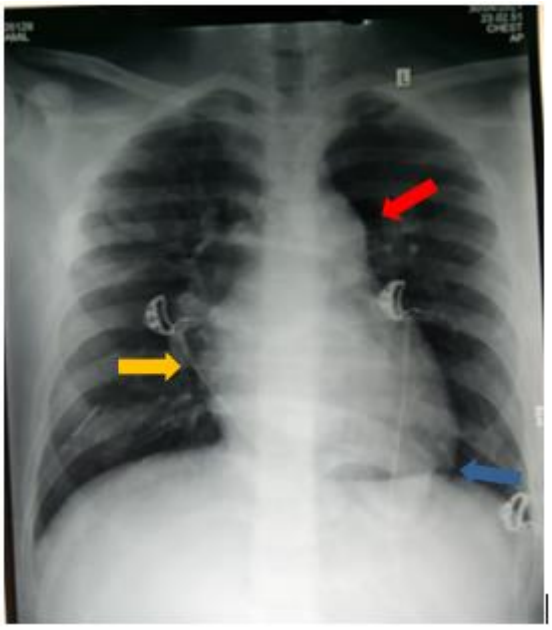

Figure 2. Chest X-ray showed cardiomegaly with bulging main pulmonary segment that makes flatten cardiac waist (red arrow), right heart border enlargement (yellow arrow), and upward apex (blue arrow).

Transthoracic echocardiography examination showed RV and RA dilatation, intact interventricular septum and intact interatrial septum (figure 3a), reduced right ventricular systolic function with tricuspid annular plane systolic excursion (TAPSE) of 15 mm, globally normo-kinetic and dilated pulmonary artery measuring 34 mm (figure 3b). Dominant RV with RV/LV basal diameter ratio=2.5 (figure 3c), LV eccentricity index >1.1 during systole and diastole, RA area (end systole) =20,1cm^2, inferior vena cava >21 mm with inspiratory collapse > 20% with quiet inspiration. Doppler examination showed mild tricuspid regurgitation with TR Vmax 5.3 m/sec and TVPG 111 mmHg, PV AccT = 60 ms (<105 ms), early diastolic pulmonary regurgitation >2.2 m/s.

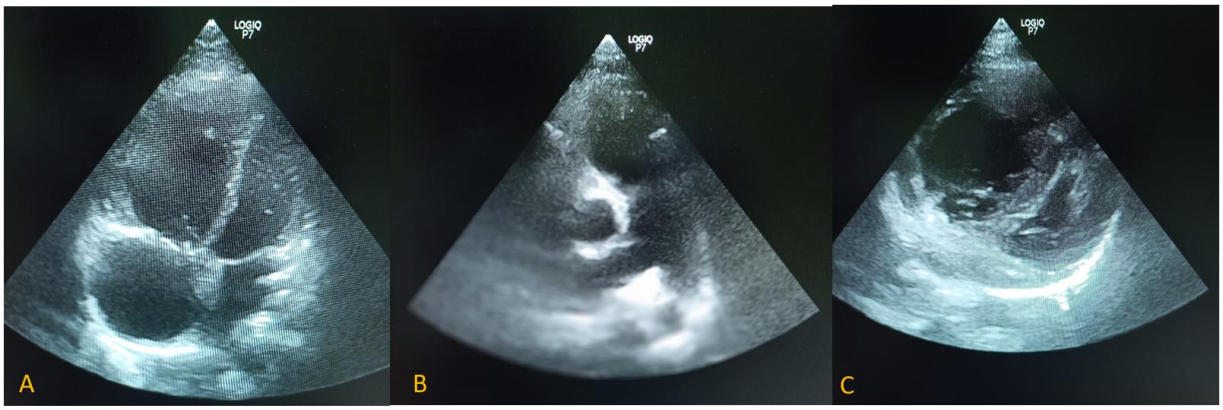

Figure 3. Echocardiography examination showed RV and RA dilatation, intact interventricular septum and intact interatrial septum (figure 3a), dilataion of pulmonary artery diameter (figure 3b) and RV dominant with RV/LV basal diameter ratio =2,5.

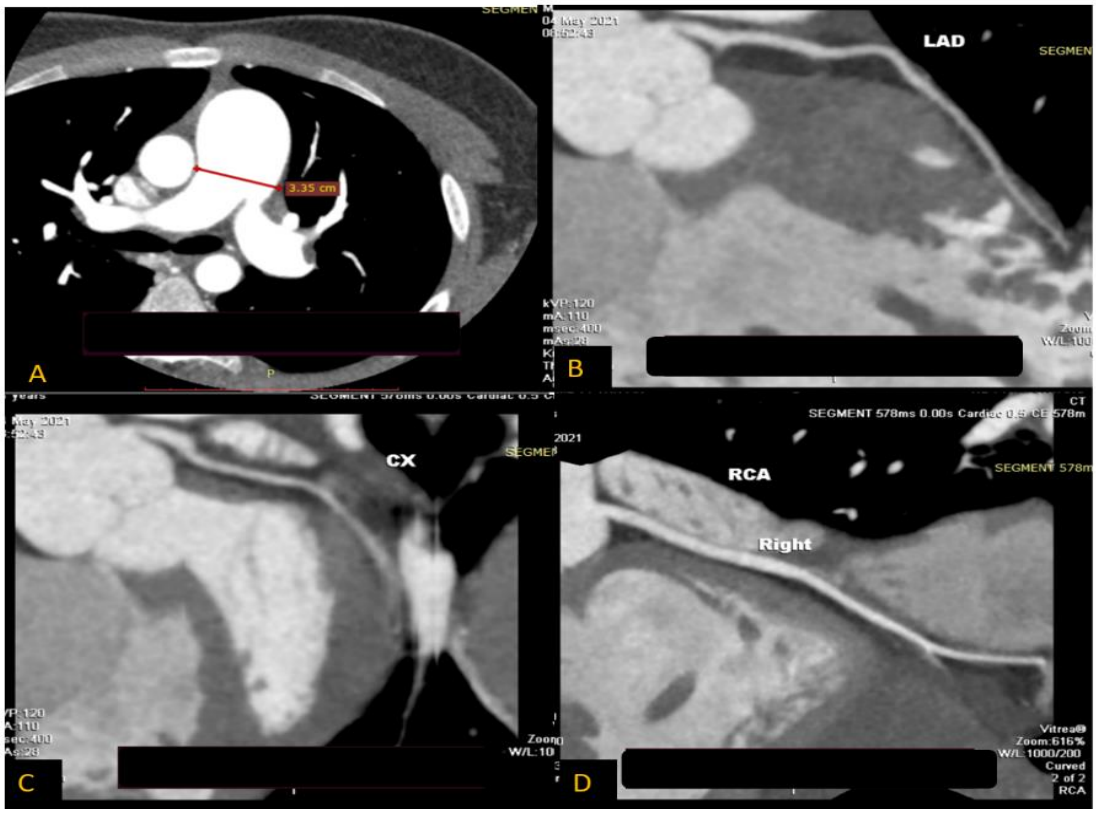

Figure 4. CT angiography showed dilatation of pulmonary artery and right pulmonary artery looks stenosed (figure 4a), and no obstructive coronary artery disease in left main – left anterior descending (figure 4b), left circumflex (figure 4c), and right coronary artery (figure 4d).

Computed tomography angiography (CTA) showed dilatation of pulmonary artery (figure 4a) and dilated right ventricle (RV), compatible to pulmonary hypertension (pulmonary artery diameter 30 mm), PA/aorta ratio = 1:3 (normal PA/Ao >0,9), RV free wall thickness was >4 mm, lumen ration for RV/LV = 3 (normal ratio for RV/LV >1). Scan did not show any soft or calcified plaque or significant luminal stenosis in left main – left anterior descending (figure 4b) , left circumflex (figure 4c) and even in right coronary artery (figure 4d).

He was treated with oral aspirin, sildenafil, beta blocker, and fondaparinux. During hospitalization his clinical condition improved, chest pain settled and patient was discharged on day 5.

DISCUSSION

Chest pain in children is often caused by different non-cardic conditions but infrequently cardiac conditions can lead to this presentation[7]. Chest pain can be a clinical presentation of pulmonary hypertension rarely. In this case, A 14-years old boy showed angina-typical chest pain, accompanied by dyspnea 3 hours prior to admission. The pain spread to his left arm accompanied by sweating. On physical examination, we found accentuated second heart sound with grade 2/6 pan-systolic murmur at left lower sternal border (punctum maximum) that increased with inspiration known as carvallo sign.

Based on Proceeding book of 2018 World Symposium Pulmonary Hypertension (WSPH), patients with suspected PH should do undergo several investigational steps starting from assessing the symptoms and signs suggesting PH (figure 5).

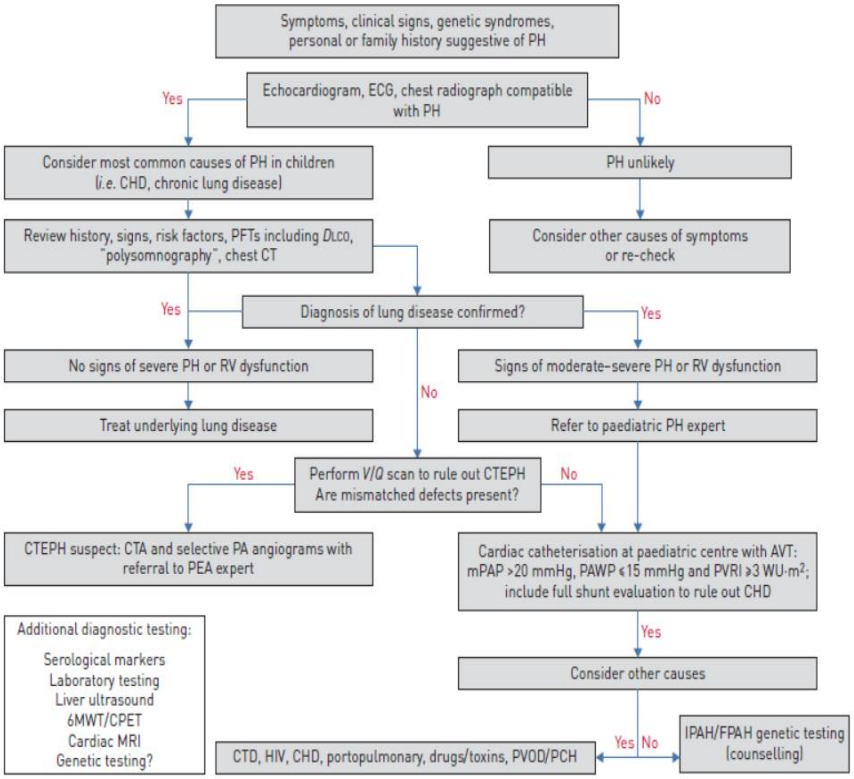

Figure 5. Diagnostic algorithm for pulmonary hypertension in children.[1]

In ECG findings, we found RAD, RAE, RVH with RV strain. It had been known that several ECG findings alone were not sufficient for diagnosing PH even though QRS right-axis deviation was the best discriminator and was highly suggestive of RV enlargement.[8]

Chest X-ray showed cardiomegaly with increased PA segment, RAE and RV enlargement, generally associated with elevated pulmonary artery (PA) pressure.[9] Elevated PA pressures are associated with enlargement of the right descending pulmonary artery (RDPA) and left descending pulmonary artery (LDPA).[9] If both LDPA and RDPA were enlarged on chest radiography the positive predictive value of detecting PH is 93%.[9]

Based on guideline, transthoracic echocardiography should be performed as the first line modality to stratify the probability of PH. There are several echocardiographic signs of PH endorsed by the European Association of Cardiovascular Imaging.[3] From echocardiographic finding, We found RV/LV basal diameter >1.0, IVC diameter >21 mm with decreased insipiratory collapse, left ventricular eccentricity index > 1.1, right atrial area (end-systole) > 18 cm2, early diastolic pulmonary regurgitation velocity >2.2 m/sec, PA diameter > 25 mm. Patient was classified into high probability of pulmonary hypertension. Another recommended scan is V/Q scan to detect any mismatch perfusion defects.

In CT angiography we found dilatation of pulmonary artery and no obstructive coronary artery disease in left main – left anterior descending, left circumflex and right coronary artery. After discharged, patient was referred to cardiovascular center in Jakarta for having further work up, in order to find the etiology if any and exclude other possible causes of pulmonary hypertension.

CONCLUSION

Pulmonary artery hypertension in 14-years old boy may present similar to acute coronary syndrome. The uncommon symptom like chest pain in pediatric should be assumed as cardiac if patient had family history of cardiac disease such as pulmonary arterial hypertension group 1. Baseline investigations like ECG, echocardiogram and chest X-ray should be performed to establish the diagnosis and to establish the cause of pulmonary hypertension. Further scans or invasive studies may be needed to find etiology of pulmonary hypertension.

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