A Challenging Case of Bilateral Pulmonary Emboli Requiring Emergent Thrombectomy in a Pediatric Patient

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Background

Pediatric Pulmonary Emboli (PE) are rare, with a reported incidence of 0.14 - 0.9 in every 100,000 children per year [1]. PE in pediatric populations appears to have a bimodal age distribution, affecting infants/toddlers (ages 0 - 3) and teenagers [1]. Pediatric PE can present with very subtle or no abnormal physical exam or objective findings, often leading to a missed diagnosis. For this reason, pediatric PE carry a high patient mortality rate estimated to be 20.6 - 30.5% in patients ages 0-9 and 11.9 - 16.2% for ages 10-19 [2].

Evidence based guidelines for the management of PE in children are limited, and most recommendations are derived from pooled data of studies involving adult populations. Decisions regarding pursuing systemic thrombolysis, surgical embolectomy, catheter-based interventions, or medical management are often made on a case-by-case basis by a multidisciplinary team after careful consideration of short- and long-term benefits.

We present the case of a 17-year-old female who was successfully treated with mechanical thrombectomy for bilateral sub-massive PE after recent initiation of combined oral contraceptives (OCP).

Methods

A comprehensive review of the patient's medical record was performed. Diagnostic modalities such as Transthoracic Echocardiogram (TTE), Computed Tomography Angiography Pulmonary Embolism (CTA PE), and Right Heart Catheterization(RHC) with Pulmonary Angiography were performed. The patient's care involved a multidisciplinary approach including specialists in Adult Interventional Cardiology, Pediatric Critical Care, Pediatric Cardiology, and Pediatric Hematology.

References

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Case Summary

17 year-old Female with past medical history of anxiety and recent initiation of combined oral contraceptives presented to the ED, for the evaluation of shortness of breath. Patient mentioned that the shortness of breath had been progressively worsening over the last week, especially with physical exertion, and this morning she developed significant pleuritic chest pain with associated lightheadedness and dizziness.

- Vitals: HR = 115, BP = 103/76, RR = 27, SpO2 = 93% on 2 L NC.
- EKG: Sinus tachycardia, new right bundle branch block, and right axis deviation
- D-Dimer = 4.36 mg/L
- High sensitivity Troponin = 648 ng/L
- NT proBNP = 5,664 pg/mL
- CTA PE: Prominent bilateral pulmonary emboli involving the distal main, lobar, and segmental branches of all lobes of the lungs with greatest prominence in the lower lobes, with evidence of right heart strain (Figures 1, 2). Patchy nodular opacities noted in the left upper lobe possibly representing pulmonary infarcts (Figure 3).
- TTE was performed showed a severely dilated Right Ventricle (RV) with reduced systolic function, interventricular septal bowing, tricuspid regurgitation (Figures 4, 5), and right ventricle strain (Figure 6).
- Duplex ultrasound of bilateral upper and lower extremities was negative for DVT.

Patient was diagnosed with sub-massive PE, started on heparin infusion, and admitted to Pediatric Intensive Care Unit due to increasing oxygen requirements. A multidisciplinary discussion was had amongst Pediatric Cardiology, Hematology, Critical Care, and Adult Interventional Cardiology who determined that she would benefit from thrombectomy given the severity of her symptoms. Patient's family elected to proceed & she was transferred to our institution.

Pulmonary angiography confirmed CT findings (Figure 7), and emergent bilateral thrombectomy was successfully performed. Post-thrombectomy the mean Pulmonary Artery Pressure improved from 36 to 20 mmHg, with complete resolution of tachycardia, hypotension, and hypoxia.

Repeat TTE showed normal RV size, systolic function (Figure 8), resolution of tricuspid regurgitation (Figure 9), as well as RV strain (Figure 10).

Pediatric Hematology recommended 5-day loading with Lovenox 1 mg/kg BID followed by oral DOAC for a 3-month course. Hypercoagulable evaluation included: Prothrombin genetic testing, Lupus Anti-coagulant & mixing study, β2 glycoprotein, and anti-cardiolipin antibody. Anti-cardiolipin IgG was elevated (11 u/mL; ULN = < 10 u/mL), and all other studies were negative.

Post hospital Follow Up:

CTA PE 3-months post-thrombectomy demonstrated complete resolution of pulmonary emboli, left upper lobe densities (Figure 11), and RV dilation (Figure 12).
Patient discontinued use of all combined OCP indefinitely, completed a 3-month course of DOAC, and is now being monitored off anticoagulation.

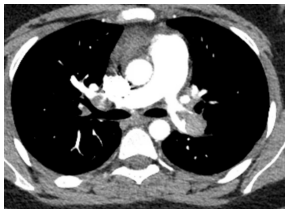

Figure 1: CTA PE demonstrating prominent bilateral pulmonary emboli involving the distal main, lobar, and segmental branches of all lobes of the lungs.

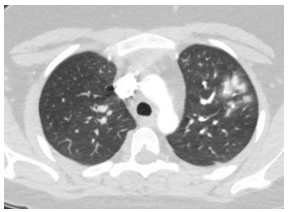

Figure 3: CTA PE highlighting the greatest prominence of thromboembolism burden in the bilateral lower lobes. Enlargement of the right ventricle with evidence of right heart strain demonstrated.

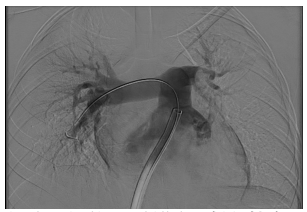

Figure 7: Pulmonary Angiogram during emergent mechanical thrombectomy confirming CTA PE findings of extensive bilateral pulmonary emboli involving distal main, lobar, and segmental branches of all lung lobes.

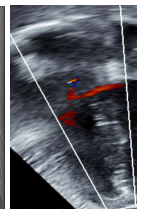

Figure 9: TTE post-thrombectomy demonstrating resolution of tricuspid regurgitation.

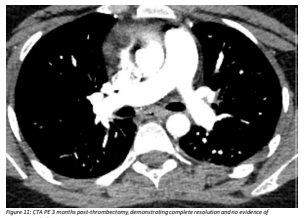

Figure 11: CTA PE 3 months post-thrombectomy, demonstrating complete resolution and no evidence of recurrent embolism.

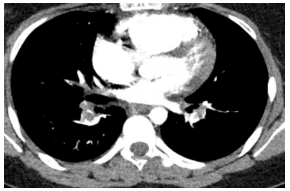

Figure 2: CTA PE demonstrating the left upper lobe opacities suggestive of punctate pulmonary infarcts.

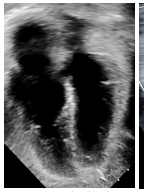

Figure 4: TTE prior to mechanical thrombectomy demonstrating interventricular septal bowing, right ventricular enlargement, and impaired systolic function.

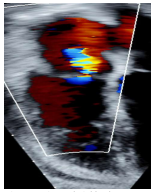

Figure 5: TTE prior to mechanical thrombectomy demonstrating moderate tricuspid regurgitation.

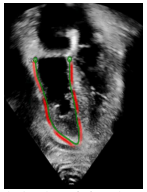

Figure 6: TTE prior to procedure demonstrating right ventricle free wall strain -12.1% and septal strain -11.9%.

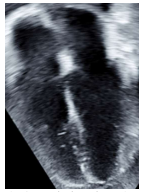

Figure 8: TTE post-thrombectomy demonstrating improved right ventricle systolic function, size reduction, and resolution of interventricular septal bowing.

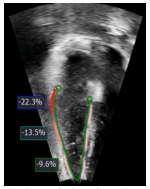

Figure 10: TTE post-thrombectomy demonstrating improved right ventricle free wall strain (-14.8%) and septal strain (-15.2%).

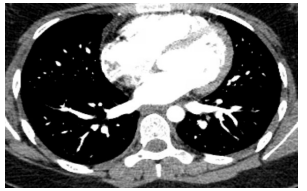

Figure 12: CTA PE 3 months post-thrombectomy demonstrating normal right ventricle size.

Discussion & Conclusion

Our patient experienced a sub-massive PE, provoked by recent initiation of combined OCP, and was successfully treated with mechanical thrombectomy.

Mechanical thrombectomy was performed on our patient because of the extent of the PE, the degree of right heart strain, and concerns for right heart failure from persistent pulmonary hypertension. Guidelines for PE management in children are limited and are based on information from adult populations. Previously the management of PE involved initiation of anticoagulation, with the use of systemic thrombolytics reserved for severe cases of massive PE. Mechanical thrombectomy has been growing in popularity as an intervention for sub-massive PE because of its potential benefits regarding length of hospital stay and reduction in ICU admissions. Saleh et. al reported a statistically significant thirty-day (4.8% vs 9.1%), one year (12.9% vs 25.6%), and two-year (16.1% vs 25.6%) mortality benefit in adult patients who had undergone mechanical thrombectomy vs systemic anticoagulation alone [3]. While the results of this study are statistically significant, the small sample size limits the generalizability. Larger studies investigating this topic such as the PEERLESS-II study are currently underway and will provide further insight.

Combined OCPs containing Estrogen are known to increase the risk of thrombus development in young and adolescent women, by approximately 3-4-fold [4]. In the study conducted by He et. al., approximately 70% of thromboembolism development in adolescent girls was associated with the use of estrogen containing OCPs [4]. The increased risk of thromboembolism development is attributed to the acquired resistance to activated Protein C as well as increased thrombin production from elevated estrogen levels [4]. Patients with thrombus development within 6 months of initiating estrogen containing OCPs were found to have the greatest prevalence of traits (Factor V Leiden, Protein C/S, Lupus anti-coagulant, etc.) predisposing to the development of activated protein C resistance [4]. Our patient had no known personal or family history of thromboembolism development, however hypercoagulable evaluation revealed a weakly positive Anti-cardiolipin antibody. Given the positive Anti-Cardiolipin and development of PE shortly after initiation of Estrogen containing OCP, it was recommended that the patient forgo future use. She was ultimately considered low risk for future thrombotic events and did not need to continue long-term anticoagulation.