Cardiology in the Young

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Massive bilateral pulmonary embolism in a child

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Brief Report

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Abstract

We describe a rare case of acute pulmonary artery thromboembolism in a 17-year-old male patient who presented to our emergency department following a syncopal episode. A chest radiograph showed a convex pulmonic cone and an increased cardiothoracic index, and two-dimensional echocardiogram suggested near-occlusion of both pulmonary arterial branches. Multi-slice pulmonary angio-tomography revealed massive thrombosis of the pulmonary artery. He was treated with systemic anticoagulation and subsequently required surgical thrombectomy, with favourable early outcome. Although the cause of the thromboembolism remains unproven, we discuss possible etiologies.

Pulmonary thromboembolism is a potentially lethal clinical expression of blood clots embedded in the pulmonary tree, and it is usually a secondary entity that can originate from multiple factors that involve endothelial damage, venous stasis, and thrombophilia (Virchow's triad). In most cases, the source is the deep venous system of the lower extremities and less frequently the pelvic veins. The entity is extremely rare in children and adolescents. Although systemic anticoagulation and control of the primary problem are the mainstays of treatment, occasional patients may require surgical thrombectomy as a life-saving measure. Most paediatric teams will not have encountered this situation, which prompts us to submit this case report.

Case report

A 17-year-old 59-kg male presented to our emergency department with 4 brief syncopal episodes (a few seconds duration), over the past 4 months, associated with running. He complained of a dry cough and mild ankle and lower leg edema for 2 weeks prior to admission. There was a history of pneumonia and a gluteal abscess 3 years earlier, with negative blood cultures. Both problems resolved, apparently without residua. He was currently taking no medications but was an occasional smoker of cannabis. He had a history of anabolic steroid (oxandrolone, exact dose unknown) use during his previous 2 years of baseball training.

Physical exam showed a hemoglobin SO_2 of 98% by pulse oximetry. Pulse was 130/minute. Blood pressure was 100/70 mmHg with respiratory rate 24/minute. Temperature was 37 degrees Celsius. Mild pedal and calf edema were present bilaterally. There was a prominent right parasternal heave. A para-systolic systolic murmur was present, grade II/VI, and S2 was pathologically split.

His chest X-ray showed mild cardiomegaly, with a prominent convex pulmonary cone. Pulmonary vasculature was prominent. Electrocardiogram showed sinus tachycardia with tall p-waves, a right axis, and augmented right-sided voltage (Figs. 1 and 2).

Laboratory results were as follows: hemoglobin 13.5 g/dL; hematocrit 41.3%; leucocytes $4.7 \times 10^3/mm^3$; prothrombin time 17.9 seconds; partial thromboplastin time 25.7 seconds. Antiphospholipid panel was normal. D-dimers were elevated at 12,615 ng/ml. Sputum culture showed no growth. Viral screen for human immunodeficiency virus, hepatits B, hepatitis C, and antibodies for COVID-19 were all negative. Mycobacterial DNA was not detected (GeneXpert assay).

A transthoracic 2D echocardiogram was performed, revealing a dilated inferior vena cava with less than 50% inspiratory collapse, with spontaneous contrast inside the vessel (Fig. 3a–d). The interventricular septum was flattened, with paradoxical movement. A hyper-refrigerant area was observed in the left branch of the pulmonary artery, suggestive of a 29 × 22 mm thrombus, which occluded the entire lumen. There was significant dilation of right atrium and ventricle. Tricuspid annular plane systolic excursion (TAPSE) was 16, with severe tricuspid regurgitation. Pulmonary artery systolic pressure was estimated to be 94 mmHg.

A lower limb Doppler ultrasound suggested chronic deep vein thrombosis in both lower extremities. There was compromised flow in both femoral and popliteal veins. There was chronic superficial thrombophlebitis and valvar insufficiency, involving both great saphenous veins, including both femoral saphenous junctions.

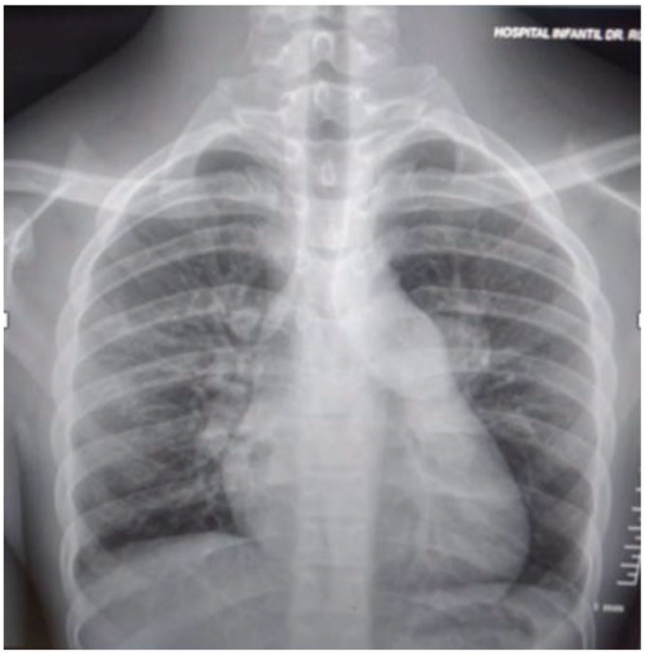

Figure 1. Chest X ray (see text).

Multi-slice tomographic angiography of the thorax was performed. Acute central and peripheral pulmonary thromboembolism was observed in the right, left, segmental, and subsegmental pulmonary arteries (Fig. 4). There was dilatation of the pulmonary trunk. Also noted was bilateral interstitial pneumonia, with evidence of left endobronchial dissemination, suggesting infected bronchiectasis. Bilateral basal chronic pulmonary inflammatory changes and hepatomegaly were also noted.

Clinical course

Enoxaparin was started (1 mg/kg subcutaneously) but after 45 days of therapy the echocardiogram was unchanged. Because the patient remained symptomatic, we decided to offer bilateral pulmonary thrombectomy. The operation was performed through median sternotomy with normothermic full flow cardiopulmonary bypass, employing aorto-bicaval cannulation. The intracardial pulmonary arteries were quite dilated, as was the right heart. The main PA and both primary branches were incised out to the hilum, and all of the organised thrombus was dissected and extracted out to and including the lobar branches (Fig. 5). The patient was separated easily from cardiopulmonary bypass despite initially systemic pulmonary pressure, which fell to 70% systemic with inhaled nitric oxide 40 ppm.

Postoperatively, the patient required 24 hours of ventilation, 2 days of inotropes, and 2 days of nitric oxide, with subsequent conversion to sildenafil. A post-operative echocardiogram on day 2 showed notable improvement in right atrial and ventricular dilation, reduced pulmonary arterial pressure, and no evidence of thrombi. He was discharged free of symptoms on day 5, with continued anticoagulant treatment (oral rivaroxaban) under outpatient follow-up (cardiology, vascular surgery, and hematology). An echocardiogram 3 months post-operatively showed resolution of all pre-operative abnormalities. He continues to receive oral anticoagulants awaiting resolution of the deep venous thrombosis.

Discussion

Pulmonary thromboembolism requiring surgery is an extremely rare disease in children, especially in otherwise healthy patients such as ours. The true incidence is unknown since the entity is probably under-diagnosed. By exclusion, we speculate that in our case the use of anabolic steroids (synthetic testosterone analogues) could have been the most important contributory factor.

Anabolic steroids are infamous sports performance enhancing medications. High-dose or multiple anabolic steroid use has been associated with serious side effects, including cardiovascular adverse events such as dyslipidemia, hypertension, cardiac hypertrophy, and increased vascular procoagulants.[1,2] Experimental data would suggest that testosterone increases human platelet thromboxane A_2 receptor density and aggregation

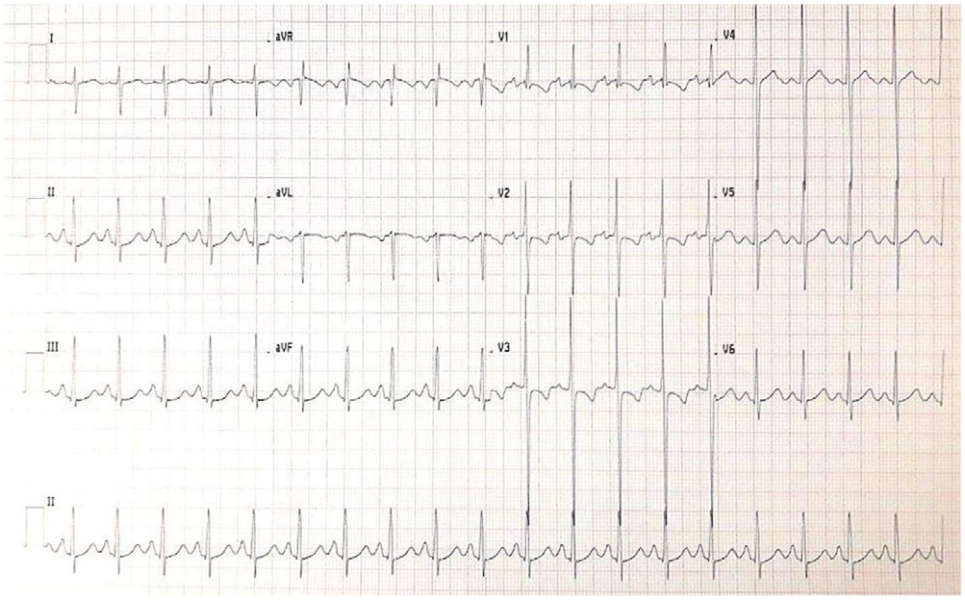

Figure 2. Electrocardiogram (see text).

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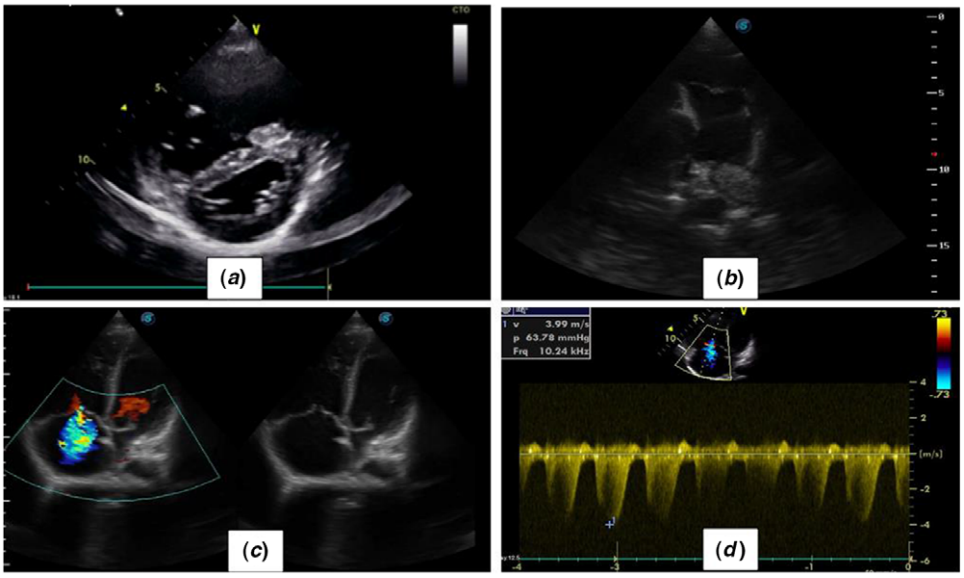

Figure 3. Transthoracic echocardiogram (see text).

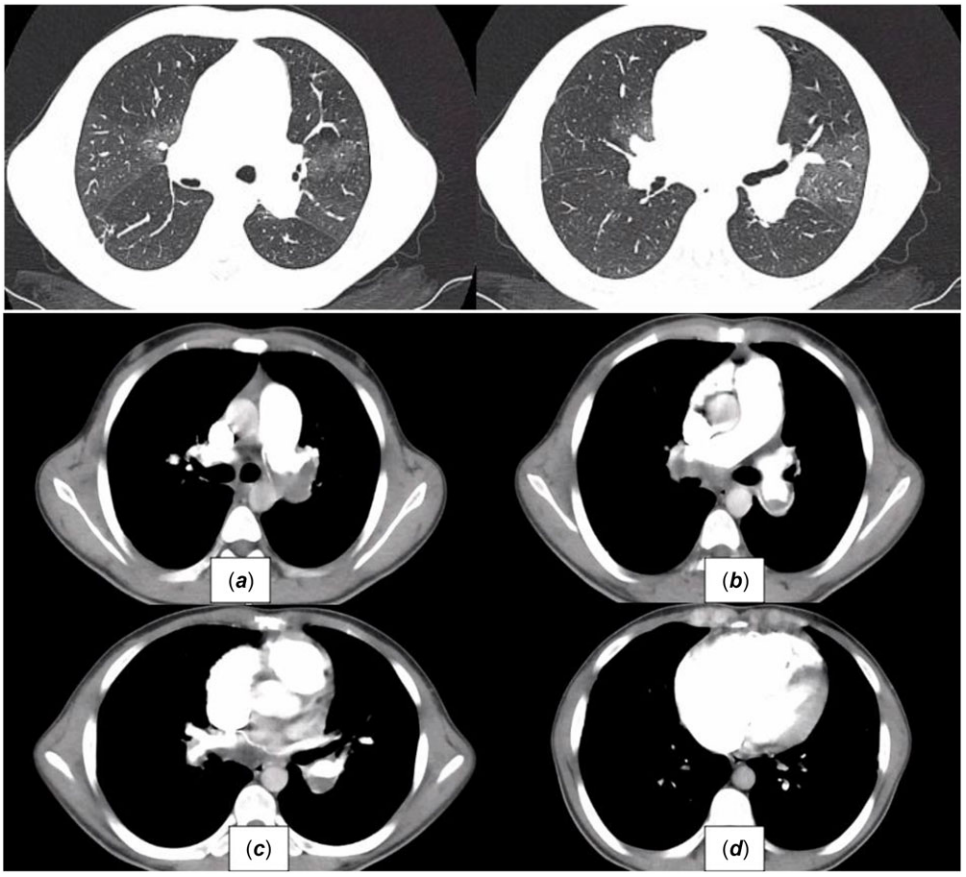

Figure 4. CT images (see text).

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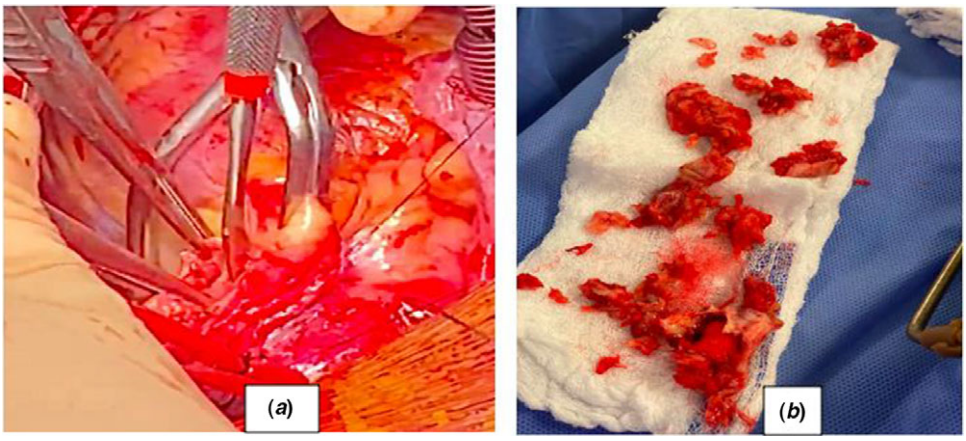

Figure 5. ***a***, ***b***. Surgical thrombus removal from pulmonary circulation via opened main and branch pulmonary arteries (a) and extracted clots (b). (See text).

responses.[3,4] There is only limited information available associating anabolic steroid use with clinical pulmonary embolism.[2]

Surgical pulmonary endarterectomy may be required in cases not responsive to anticoagulation, especially in the presence of haemodynamic compromise.[5] Balloon pulmonary angioplasty may be an alternate treatment for patients that are unsuitable for operation.[5] Although it is complex and possibly high risk, a catheter-based approach may allow dilation of occlusions to the level of subsegmental vessels, which are usually inaccessible to surgery.

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Informed consent. The authors indicate that they have the informed consent of the patient, family, and institution for the publication of this article.

Competing interests. The author declares that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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