Case Report: Angio Behçet in a child [version 1; peer review: awaiting peer review]

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\textbf{Abstract}

Behçet's disease (BD) is rare in children. Herein, we report a case of 12-year-old male child with a history of left superficial femoral vein thrombosis and bilateral pulmonary embolism, admitted for multiple thrombosis located in the inferior vena cava, right atrium and ventricle associated with multiple pulmonary artery aneurisms and fissurations concomitant to oral aphthosis, which was suggestive of BD. Despite corticoids and immunosuppressive therapy, the child had a massive hemoptysis resulting in his death. The association between intracardiac thrombosis, pulmonary aneurism and venous thrombosis should raise suspicion of angio-Behçet, which could be lethal.

\textbf{Keywords}

behcet, pulmonary artery aneurism, thrombosis

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Introduction
Behçet’s disease (BD) is the only primary vasculitis that affects both arteries and veins of any size. Cardiac complications are rare but they represented one of the prognostically devastating manifestations of BD. They predominate in the right heart and are often associated with pulmonary artery aneurysm. Physicians are frequently faced with a therapeutic dilemma due to the absence of consensus for treatment of such complications. To the best of our knowledge, there is no evidence about the prevalence and the prognosis of pulmonary artery involvement in children with BD. Herein, we reported a case of a child presenting with Angio-Behçet, revealed by intra right ventricle thrombus and pulmonary artery aneurysm. It should be kept in mind that some life-threatening features in the pediatric population may occur earlier than mucocutaneous findings.

Case report
An African 12-year-old male child was admitted for cough, hemoptysis and progressive dyspnea. He had a history of left superficial femoral vein thrombosis and a massive bilateral pulmonary embolism three months before, which had been treated with low molecular weight heparin (15 UI/kg/h intravenously) and oral vitamin K antagonists (VKAs) (1 mg per day). He mentioned an episode of oral ulcers with no genital lesions. He denied any history of joint pains, skin rash or visual complaints but he has suffered from recurrent headaches and anxiety. Family history was non-contributory.

Physical examination revealed thoracic collateral venous circulation and hepatomegaly with jugular veins distension. Initial laboratory results showed hemoglobin=11.2 gm/dL, platelets=232,000/mm³, and white blood cells=10×10⁹/mm³, International Normalized Ratio (INR)=2.3 and D-Dimer=1,500 ng/L. Chest X-ray showed images of bilateral perihilar condensations and electrocardiogram showed a right bundle branch block. Pulmonary computed tomography angiography showed inferior vena cava, right atrial and right ventricular thromboses (Figure 1) associated with multiple pulmonary artery aneurisms (PAAs) with fissurations (Figure 2). There were also bilateral infiltrates and peripheral subpleural opacities suggestive of pulmonary infarcts (Figure 1). Thrombophilia and connective tissue screening were negative.

During hospitalization, the child developed oral aphthosis suggestive of the diagnosis of angio-Behçet. VKAs were maintained with IV methylprednisolone at a dose of 15 mg/kg/day over three days. After that, a cyclophosphamide bolus at 700 mg/m² was introduced. Despite these treatments, the child died from a massive hemoptysis leading to respiratory compromise.

Discussion
BD is characterized by multisystemic involvement of unknown etiology, for which viral, bacterial, genetic, environmental and immunological factors have been implicated. The prevalence of BD in children is unknown but is probably very low, as a range of 3.3–26% of cases have been reported. 

Figure 1. CT scan (axial reconstruction) of right atrial and right ventricular thrombosis. CT, computed tomography.
In 2015, a recent publication established a classification criteria from the largest prospective cohort ever reported for BD in children. Three of six items are required to classify a patient as having pediatric BD. In fact, our patient showed three criteria for BD, oral aphthosis, neuropsychiatric signs as headaches, anxiety and cardiovascular signs that confirmed the diagnosis.

Cardiovascular involvement has been reported in 7–29% of patients with BD. Intracardiac thrombosis is extremely rare with an obvious predilection to the right heart location. Arterial damage occurs in 1–7% of patients with BD but it may be at the forefront of the clinical manifestation, as shown by our case, leading to a worse prognosis. PAAs occur more in men and appear in younger patients. The inflammatory process in PAAs affects the vasa vasorum of the artery, causing weakness in the vessel wall. These lesions frequently erode bronchi and cause massive hemoptysis. The treatment of BD is not well established, but colchicine, thalidomide, corticosteroids and immunosuppressants, can be used depending on the severity of systemic manifestations.

Due to rarity of intracardiac thrombosis as a complication of BD, there is no evidence-based consensus regarding best treatment. Mogulkoc et al., suggested that medical management with anticoagulants and immunosuppressant may be associated with better outcomes than surgical intervention. The use of anticoagulation could be dangerous in patients who presented both thrombosis and hemoptysis. The EULAR recommendations for treatment of BD concluded that there is no evidence of the benefits for the use of anticoagulants for arterial lesions of BD. Huong et al., reported the case of a patient with intracardiac thrombosis and bilateral pulmonary aneurysms who had complete resolution of thrombus by immunosuppressant therapy without anticoagulants. The use of immunosuppressants has been shown to significantly improve survival and prognosis in patients with thrombotic and arterial lesions. Regarding the choice of immunosuppressant, there are no strong recommendations to support superiority of one regimen over another. Cyclophosphamide has been used to induce remission. Anti-TNF agents are more frequently used in patients with multi-systemic involvement and show promising results but they are more expensive and not frequently available in some underdeveloped countries.

Despite treatment of our patient with an immunosuppressant, the prognosis was poor.

**Conclusions**

The association between intracardiac thrombosis, pulmonary aneurism and venous thrombosis are very characteristic of angio-Behçet in children. The prognosis is poor despite treatment. Anticoagulation therapy may be more beneficial in patients that only show intracardiac thrombosis but it should be avoided when the patient is experiencing pulmonary arteries aneurysms.

**Data availability**

All data underlying the results are available as part of the article and no additional source data are required.

**Consent**

Written informed consent for publication of their clinical details and clinical images was obtained from the parent of the patient.

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**Figure 2.** CT scan (axial reconstruction) of pulmonary artery aneurisms with fissurations. CT, computed tomography.
References


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