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Brief Report

Lung Abscess in Adults with Tetralogy of Fallot and Pulmonary Atresia

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Abstract We describe 5 adults with tetralogy of Fallot and pulmonary atresia who developed lung abscesses, including some infected with atypical microbial pathogens, with important morbidity. We hypothesize that patients with such anatomy are at risk for chronic pulmonary infection due to hypo-perfusion of the pulmonary parenchyma. This previously unreported clinical association should be considered in the differential diagnosis of patients with tetralogy of Fallot and pulmonary atresia who alter their respiratory state.

Keywords: Adult congenital heart disease; pulmonary atresia and ventricular septal defect; pulmonary disease

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Tetralogy of Fallot with pulmonary atresia is a congenital cardiac lesion marked by hypoplastic pulmonary arteries fed by flow from a source other than the heart, often through aortic-to-pulmonary collateral arteries. Corrective treatment for patients with this combination, if possible, involves the restoration of continuity from the right ventricle to the pulmonary arteries, and aggressive transcatheter rehabilitation of the pulmonary arteries, including any incorporated collateral vessels.[1] Advances in treatment have resulted in improved survival, with many patients now reaching adulthood. This growing cohort of patients mandates increased vigilance for associated complications. In this report, we emphasis the significance of pulmonary abcesses.

Case Reports

We have now encountered 5 adults with tetralogy of Fallot and pulmonary atresia who developed pulmonary abscesses. Unusual pulmonary collateral circulation was noted in 4 of the patients, with 1 patient having experienced prior pulmonary infarction subsequent to a transcatheter interventional procedure. The patients had varied clinical presentations, including hypoxia, cough, haemoptysis, and pleuritic chest pain. We have summarized the clinical features in the Table 1.

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Discussion

We hypothesize that patients with tetralogy of Fallot and pulmonary atresia are at risk for chronic pulmonary infection due to maldevelopment of the pulmonary circulation, and hypoperfusion of the pulmonary parenchyma. The pulmonary vascular abnormalities in patients with tetralogy of Fallot and pulmonary atresia may create an opportunity for chronic colonization and infection by facultative or anaerobic organisms. Hypoplasia or absence of the intrapericardial pulmonary arteries, often with persistence of systemic-to-pulmonary collateral arteries or supply through a patent arterial duct results in dramatically altered pulmonary vascular distribution. In those with collateral arteries, the absence of protective proximal stenosis can trigger focal pulmonary vascular disease, with the propensity for stasis, thrombosis, and haemorrhage. Pulmonary infarction in this setting can result in both aseptic pulmonary cavitation and development of lung abscesses[2–4] This may be similar to the

Table 1. The clinical findings in our patients with pulmonary abcesses in the setting of tetralogy of Fallot and pulmonary atresia.

Age/Sex	Prior Surgery	Pulmonary Collateral Circulation	Signs and Symptoms	Microbiology	Outcome
30/Male	Left Blalock-Taussig shunt, right ventricle-to-left pulmonary arterial conduit, closure of ventricular septal defect, right upper lobe unifocalization	Systemic-to-pulmonary collateral arteries to left lung and right upper lobe	Fatigue, cyanosis, weight loss, sweats, chills; cavitary lesion right lower lobe*	*Mycobacterium intracellulare*	Triple antibiotics; slow improvement
36/Female	No prior surgery	Multiple systemic-to-pulmonary collateral arteries	Recurrent pneumonia; hypoxia; left cavitary lesion †	*Not isolated*	Cavitary lesion developed; death
22/Female	Left Blalock-Taussig shunt to unifocalized collaterals	Multiple systemic-to-pulmonary collateral arteries	Haemoptysis, dyspnoea, wheeze; multiple cavitary lesions left lung ‡	*Aspergillus; Enterobacter cloacae*	Progressive symptom; death
40/Male	Left, right Blalock-Taussig shunts, central shunt	Multiple systemic-to-pulmonary collateral arteries to left lung, and small collateral arteries to right lung	Fever, cough, hypoxia, skin papules; cavitary lung lesions	*Mycobacterium abscessus*	Disseminated infection; death
19/Female	Right ventricular outflow tract patch, ventricular septal defect patch, history of pulmonary infarct	None	Pleuritic chest pain, multiple bilateral peripheral cavitary lesions	*Staph non-aureus*	Antibiotics, eventual resolution

* Figure 1a.
† Figure 1b.
‡ Figure 1c.

Figure 1.
*The computed axial tomographic scan for our first patient (**a**) shows a cavitary lesion in the right lower lobe with adjacent calcification. A similar scan in our second patient (**b**) shows a large left cavitary lesion with anterior parenchymal consolidation, while a scan from our third patient (**c**) reveals a cavitary lesion of 2 to 3 centimeters in diameter in the left lower lobe.*

propensity to infection described secondary to micro-infarction in sickle cell anemia.[5]

Development of abscesses in the lungs is associated with high rates of morbidity and mortality despite aggressive pharmacologic therapy.[6,7] Pathogens which are nosocomially acquired, associated with underlying immunocompromise or with pulmonary anatomic defects, are particularly concerning.[8,9] The impact of chronic cyanosis on immunity has not been well explored, but may play a role in the development of infection in this population. In addition, patients with tetralogy of Fallot and pulmonary atresia are frequently associated with microdeletion of chromosome 22 and DiGeorge syndrome. The clinical aspects of this syndrome can include absence of the thymus, and defects in T cell immunity.[10] No patients in our series, however, had been genetically screened to establish this combination of features. Chronic malnutrition was problematic, particularly for our second and third patients, although in the third patient the long-lasting infection appeared a contributor rather than a consequence.

From our small cohort, 3 of the patients have now died. Although the development of pulmonary abscesses was only part of their complex clinical picture, the chronicity of infection, and its associated morbidity, highlight the impact of this complication. All 5 patients were managed without aggressive surgical intervention. The pulmonary abscesses in these patients were part of multisystem morbidity, and consideration of surgical therapy mandated weighing risks and benefits of thoracic invasion in the setting of chronic cyanosis, pulmonary vascular disease, abnormalities of lung function, and ventricular dysfunction.

The evolution of pulmonary findings on the computed tomographic scans of the chest is well illustrated by these patients. Scoliosis, cardiomegaly, and pulmonary vascular abnormalities may complicate the interpretation of plain films. Prompt evaluation using computed tomographic imaging, therefore, is recommended whenever the diagnosis of lung abscess is considered (Fig. 1). Bronchoscopy, or needle aspiration guided by computed tomography, may be helpful in identification of pathogens and assist in therapeutic decisions.

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