CASE REPORT

Abnormal origin of right pulmonary artery. Unusual case in an adult

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The anomalous origin of one of the branches of the pulmonary artery from the ascending aorta is a rare congenital heart disease, with 30% survival at the first year. We report the case of a 43-year-old male patient, showing how atypical this case is. The patient was maintaining in stable clinical conditions without any correction at an early age until the moment of its resolution. This case is totally unexpected according to the previously published in the literature.

Key words: **Pulmonary artery; Abnormal pulmonary origin; Pulmonary hypertension; Congenital heart disease.**

El origen anómalo de una de las ramas de la arteria pulmonar procedente de la aorta ascendente es una cardiopatía congénita poco frecuente, con sobrevida al primer año del 30%. Reportamos el caso de un paciente masculino de 43 años, mostrando lo atípico de este caso la supervivencia sin una corrección a temprana edad, encontrándose más allá de lo descrito por la literatura y conservando una condición clínica estable hasta el momento de su resolución.

***Palabras clave*: Arteria pulmonar; Origen anómalo pulmonar; Hipertensión pulmonar; Cardiopatía congénita.**

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The anomalous origin of a branch of the pulmonary artery with origin in the ascending aorta is infrequent, with a prevalence of less than 0.05%. The right pulmonary branch anomalous origin is the most common involved. This situation determines that one of the lungs assumes pressures and volumes similar to systemic ones [1,2].

Up to 40% of cases are accompanied by other cardiovascular abnormalities such as interruption of the aortic arch, aortic coarctation, septal defects, among many others. However, little is known about embryogenesis but it has been frequently associated with DiGeorge syndrome and diagnosis must be established in a timely and early manner given the poor prognosis due to the tendency to develop congestive heart failure prematurely, in addition to irreversible pulmonary vascular disease. Fortunately, 95% of patients are diagnosed within the first year of life. Nonetheless, if they are not early intervened, mortality rate can be of 70% within the first year of life [2,3].

CLINICAL CASE

We present here a case of a 43-year-old male who was admitted at our institution because of impairment of the functional class, associated with hemoptysis and multiple cases of respiratory infections for over 10 years. He had a history of cyanosis during feeding and a heart murmur at the age of 10 months. In addition, he had surgery for ductus arteriosus closure 32 years before. In NYHA CF II before admission. Echocardiogram and computed tomography showed the anomalous origin of the right pulmonary artery from the ascending aorta (**Fig. 1**). Cardiac catheterization showed off a main pulmonary artery with a left branch with systolic pressure of 50 mmHg, diastolic of 25 mmHg and mean artery pressure of 33 mmHg; right pulmonary branch with pressures equivalent to systemic of 120/60 mmHg, with mean artery pressure of 80 mmHg. Right pulmonary artery branch was emerging from the aorta. Also reported in the selective pulmonary angiography to the left branch of the pulmonary artery with a homogeneous filling of the vasculature and with adequate amount of monopedial arteries, while the right selective angiography was reported with images of the vessels with heterogeneous filling and tortuous vessels with slow filling with decrease of monopedial vessels.

Operation was performed as open chest surgery through median sternotomy on cardiopulmonary bypass. Right femoral arterial approach was used, so the aorta can be easily handled as a main part of the operation. Bicaval cannulation was used. Once on cardiopulmonary bypass, the aorta was cross-clamped and one-single dose of anterograde cold cardioplegia

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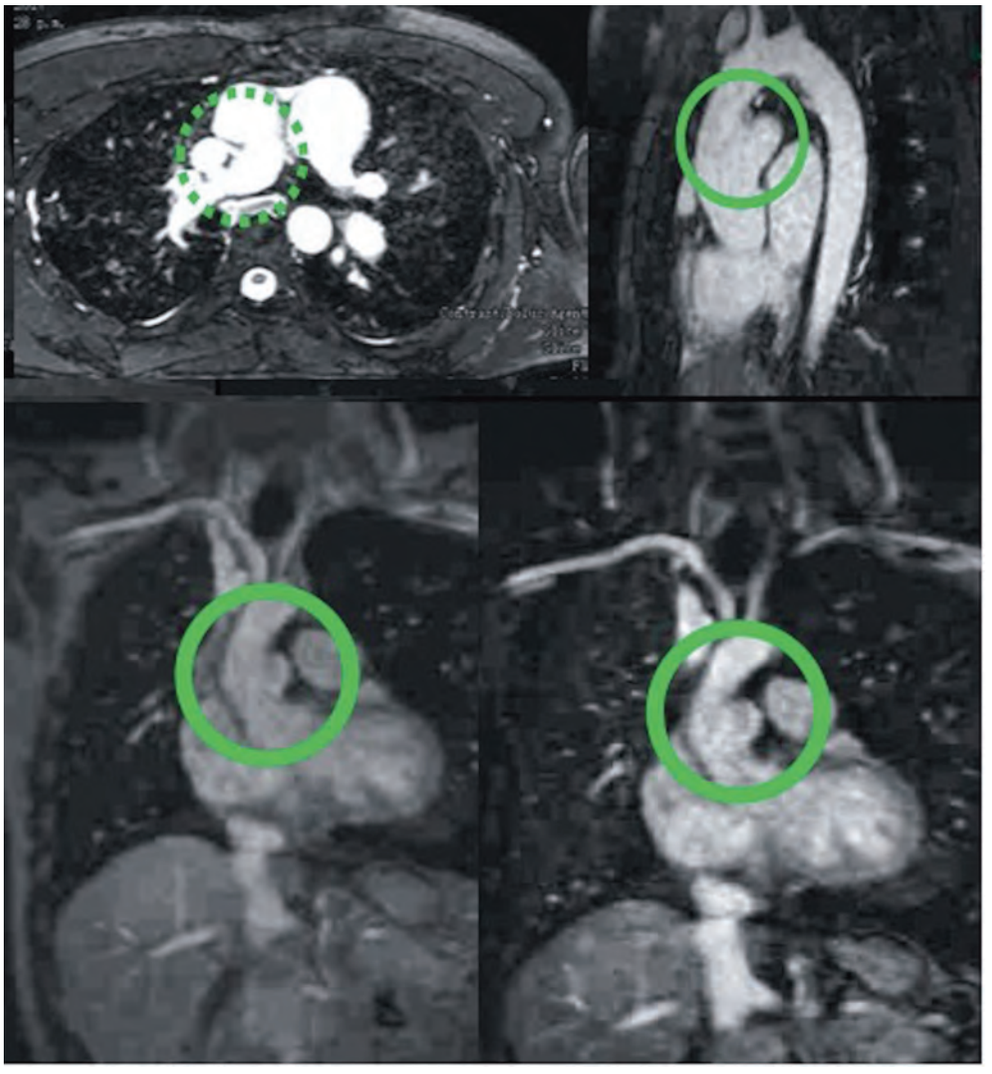

Figure 1. Cardiac CT shows anomalous pulmonary artery anomaly inside the green circle.

through the ascending aorta was administered. After heavy and full dissection around the ascending aorta to identify the anomalous branch of the pulmonary artery was carried out. Next step was sectioning this anomalous branch from de aorta, while a bovine pericardium patch was placed to cover the hole in the aorta. Reinforcement of the pulmonary ending was made by using a Teflon band. A 16mm Woven Dacron © tube was interposed between the reinforced end and the pulmonary trunk by using continuous 4-0 USP polypropylene suture (**Fig. 2**). The rest of the operation was made as usual. The postoperative course in Intensive Care Unit (ICU) was uneventful. It is worth highlighting the systemic pressure in ICU stay was on average 104/60 mmHg with mean systemic pressure of 77 mmHg versus pulmonary pressures of 47/17 mmHg and mean pulmonary pressures of 31 mmHg.

Control was performed with a selective angiography at the level of the main trunk of the pulmonary artery, observing the left branch of adequate size with a homogeneous pulmonary circulation and the right branch angled in origin. In the same way, the postoperative echocardiogram reported pulmonary artery pressure at 44 mmHg (**Fig. 3**). At in-hospital discharge, 5-phosphodiesterase inhibitors were given as supplementary therapy. According to the modified McLaughlin scale, the prognosis for this case is low risk. At a 2-year follow-up, our patient remains asymptomatic.

COMMENT

The anomalous origin of the pulmonary artery is infrequent, first described in 1868 by Fraentznel [4]. Since then, there have been several reports of isolated cases and different clinical series, chiefly in relation to the right pulmonary branch [4]. This entity is characterized by a lung receiving perfusion directly from the aorta with systemic pressure. As isolated finding, it can raise up to 20% of cases, but it is also described in association with other intracardiac lesions with left-to-right cardiac shunt, where the hemodynamic situation is further exacerbated [2,3].

Because the vasculature of both lungs becomes vulnera-

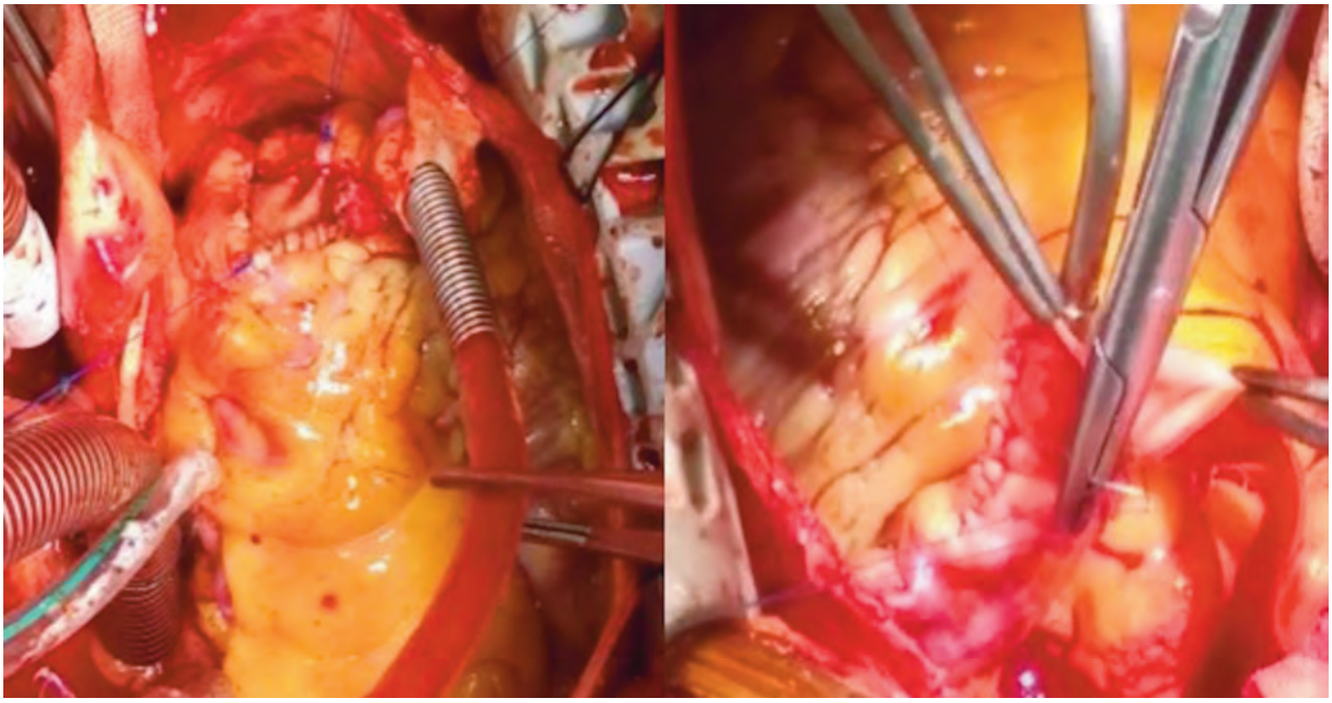

Figure 2. Exclusion and reinsertion of pulmonary branches. Repair of the ascending aorta.

ble, the natural history of this pathology shows an irreversible progression towards severe obstructive pulmonary vascular disease. With no prompt surgical correction, survival at one year of life can be worse than 30 % [2]. The clinical picture is characterized by the early onset of respiratory distress due to an increase in pulmonary flow, congestive heart failure and cyanosis when pulmonary pressure and pulmonary vascular resistance are too high [3], frequently associated with recurrent respiratory infections [2,3].

The treatment of choice is early surgical correction to avoid pulmonary hypertension and the subsequent development of obstructive pulmonary vascular disease [1-3]. Direct implantation of the anomalous pulmonary button to the pulmonary artery trunk was described by Kirkpatrick et al. in 1967 [5]. However, over the years, several surgical strategies have been proposed, such as using vascular grafts, direct reimplantation and homograft patches. However, as most of the reported cases take place in childhood, post-surgical restenosis at the site of anastomosis is frequent in this group of

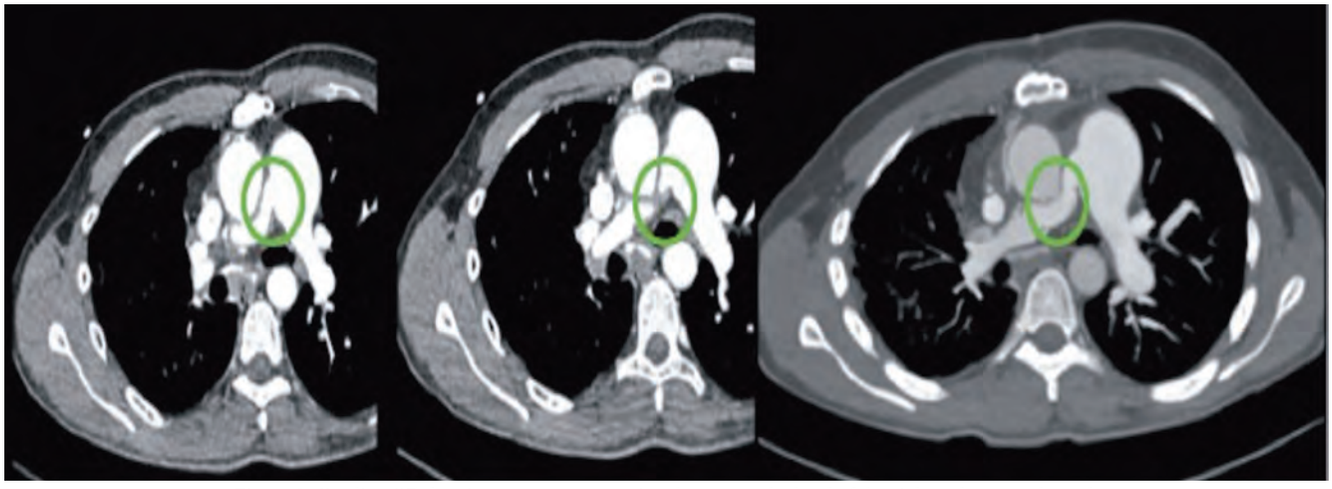

Figure 3. Cardiac CT showing some postoperative changes at the level of the right pulmonary branch.

patients [6-8].While surgical experience in pediatric patients is quite acceptable, on the other hand, the postoperative evolution in more advanced ages is unknown. There are few cases of survival in adult patient. Thus, the experience is fairly limited. Nevertheless, a good evolution after correction has been observed in most of the isolated cases, especially in adolescent patients [6,7].

In 2012, Garg et al. [9] published a series of 17 patients, where the oldest one was 25 -years old; in 14 of them, direct implantation was performed, and 3 required any graft interposition. There was a surgical death by persistent hypoxia in a child with associated Tetralogy of Fallot. Early repair resulted in acceptable hemodynamic and anatomical parameters. Operative mortality varies according to different series with ranges from 0% to 21%, and the medium-term reintervention rate for interventional catheterization with balloon catheter and stent due to pulmonary anastomosis stenosis is 2.5% to 36% [10].

Finally, it is important to highlight that in our case it was an adult patient, in stable clinical conditions in relation to the time of evolution. However, we must stress the high importance of having some degree of clinical suspicion, given the fact that it can go unnoticed. Some especial conditions as congenital heart diseases make this suspicion even more difficult as tetralogy of Fallot and pulmonary atresia, in which the circulation of a lung can depend on a major aorto-pulmonary collateral originating from the ascending aorta.

In conclusion, special caution should be taken with the decision of surgical correction, focused on the presence or absence of pulmonary hypertension and irreversible vascular damage that may coexist. In addition, a lung biopsy should be performed as a complement to establish an adequate prognosis.

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