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A Case of Pulmonary Artery Stenosis

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Abstract

Stenosis of the pulmonary artery is a condition where the pulmonary artery is subject to an abnormal constriction .Peripheral pulmonary artery stenosis may occur as an isolated event or in association with Alagille syndrome, Berardinelli-Seip congenital lipodystrophy type1, Costello syndrome, Keutel syndrome, nasodigitoacoustic syndrome (Keipert syndrome), Noonan syndrome or Williams syndrome. It should not be confused with a pulmonary valve stenosis, which is in the heart, but can have similar hemodynamic effects. Both stenosis of the pulmonary artery and pulmonary valve stenosis are causes of pulmonic stenosis. The presentation of such a case may be very misleading. The case that we present here is about a 37 year old Carribean female who presented with progressive increase in SOB.

Keywords: Pulmonary Stenosis; Pulmonary Hypertension; Cardiovascular; Hypertension; Angiography; Congenital Heart Disease; Chest Pain.

Introduction

Pulmonary artery stenosis is a narrowing (stenosis) that occurs in the pulmonary artery, a large artery that sends oxygen-poor blood into the lungs to be enriched with oxygen. The narrowing may occur in the main pulmonary artery and / or in the left or right pulmonary artery branches. This narrowing makes it difficult for blood to reach the lungs to pick up oxygen. Without enough oxygen, the heart and body cannot function as they should. In an effort to overcome the narrowing, the pressure in the right ventricle (the chamber that pumps blood into the pulmonary arteries) rises to levels that can be damaging to the heart muscle. If the narrowing in the artery is less than 50 percent, patient may not experience any symptoms. However, if the narrowing of the artery is more than 50 percent, patient may experience any of the following symptoms:- Shortness of breath , Fatigue, heavy or rapid breathing , rapid heart rate ,swelling in the feet, ankles, face, eyelids, and / or abdomen.

Pulmonary artery stenosis is a congenital heart defect, meaning it is a defect that is inborn or exists at birth. Stated another way, the defect is an abnormality, not a disease. Pulmonary artery stenosis is often present in combination with other congenital heart defects. The pulmonary valve is found between the right ventricle and the pulmonary artery. It normally has 3 leaflets that function like a one-way door, allowing blood to flow forward into the pulmonary artery, but not backward into the right ventricle.

With pulmonary stenosis, problems with the pulmonary valve make it harder for the leaflets to open and permit normal blood flow from the right ventricle to the lungs in a normal fashion. In children, these problems can include:

- A valve that has leaflets that are partially fused together.
- A valve that has thick leaflets that do not open all the way.
- Narrowing of the area above or below the pulmonary valve.

There are four different types of pulmonary stenosis:

- *Valvar pulmonary stenosis:* The valve leaflets are thickened and / or narrowed.

- *Supravalvar pulmonary stenosis:* The portion of the pulmonary artery just above the pulmonary valve is narrowed.
- *Subvalvar (infundibular) pulmonary stenosis:* The muscle under the valve area is thickened, narrowing the outflow tract from the right ventricle.
- *Branch peripheral pulmonic stenosis*: The right or left pulmonary artery is narrowed, or both may be narrowed.

Pulmonary stenosis may be present in varying degrees, classified according to how much obstruction to blood flow is present.

Case History

A 37 year old female of afro carribean origin, married with two kids, had been referred to India for treatment / evaluation of gradually progressive dyspnoea, since past one year, with ejection systolic murmur.

No past history of any cardiac / systemic illness. The patient presented to the hospital with progressive dyspnoea. On presentation to the emergency the patient was dyspnoiec.

Vitals

BP: 130 / 80 mm hg

SPO2:

Pulse: 82 / Min

Respiratory Rate: 22 / Min

CVS : S1 N S2 Wide Split with ejection systolic murmur

ECG: Equivocal / Insignificant

Labs: Not Significant.

Pulmonary artery angiogram showed bilateral significant peripheral pulmonary artery stenosis.

Patient was taken for intervention in cath lab

Pulmonary stenting was done:

Procedure details: Access Rt femoral vein and Rt femoral artery catheter used Swan Ganz 6 F and pigtail 6F.

Pressures: Systemic arterial pressure: 117/70 (88) mm hg

RV Systolic pressure: 100 mm hg

Main pul artery: 100 / 21 (54) mm hg

Left lower distal PA: 21 /1 7 mm hg

RT Distal PA: 13 / 7 (10) mm hg

Pulmonary artery angoigram: Left lower PA 70% Stenosis, (Pullback Gradient 79 mm hg)

RT Pulmonary artery: 80 % Stenosis, Pullback Gradient 87 mm hg

Pulmonart Artery Stenting: Access RFA and RFV,

Systemic Pressure: 140 /80 mm hg

Pre procedure PA pressure: 98/16 mm hg

Guide – JR 6 F

In RT pulmonary artery 7 / 16mm Herculink Plus Stent was deployed @ 13 ATM Post Dilated

with Stent baloon itself.

In lpa 6 / 16 mm herculink plus stent was deployed @15 atm post dilated with stent baloon itself .

Post procedure

Systemic pressure: 142 / 80 mm hg

Pul Artery: 77/15 mmhg

Good end result, no procedural complications.

Post procedure RV pressure almost half of systemic.

Post procedure hospital stay was uneventful, the patient was symptomatically better and was discharged in a stable condition.

Discussion

This case tells us that as emergency physician our differences have to be very broad for patients presenting with sob and dyspnea on exertion. The patient was well managed and the different modalities that can aid us reaching a diagnosis in such atypical / typical presentation are as under.

During examination in ED, doctor may hear abnormal heart sounds (a murmur) when listening to the heart. If abnormal sounds are identified, we can order for the below mentioned diagnostics from the ED itself.

- An electrocardiogram (ECG or EKG): A test that records the electrical changes that occur during a heartbeat; reveals abnormal heart rhythms (arrhythmias) and detects heart muscle stress.
- Chest X-ray: A test to show the size and shape of the heart and lungs and pulmonary arteries
- Echocardiogram: A test that uses sound waves to create a moving picture of the heart's internal structures.

- Doppler ultrasound: A test that uses sound waves to measure blood flow; usually combined with echocardiogram to evaluate both the internal structure of the heart and blood flow across the heart's valves and vessels.
- Cardiac magnetic resonance imaging (MRI): A test that uses three-dimensional imaging to reveal how blood flows through the heart and vessels and how the heart is working.
- CT scan: An X-ray procedure that combines many x-ray images with the aid of a computer to generate cross-sectional views of the heart. Cardiac CT uses the advanced CT technology with intravenous (IV) contrast (dye) to visualize cardiac anatomy, coronary circulation, and great vessels.
- Cardiac catheterization: A procedure that involves inserting a thin tube (a catheter) into a vein or artery and passing it into the heart to sample the level of oxygen, measure pressure changes, and make X-ray movies of the heart and its internal structures.
- Pulmonary angiography: A dye-enhanced X-ray of the pulmonary arteries and veins of the heart
- Perfusion scan: A test in which the patient is injected with a small amount of a radioactive material. A special machine shows how well blood is flowing through each of the two lungs.
- Additional tests may be ordered as necessary.

ED Management

Stabilizing A-B-C and then need to think of definitive management as under:

Balloon dilation

This treatment method consists of moving a balloon dilation catheter into the narrowed area of the artery. The balloon is carefully inflated – first under low pressure and then under higher pressure – until the narrowed area is widened. The balloon is then deflated and removed. Although the narrowing is improved in a majority of patients following balloon dilation, overtime the artery can again become narrow in as many as 15% to 20% of cases, requiring further ballooning. Different types of balloons are currently being developed that will likely lead to better and longer-lasting results.

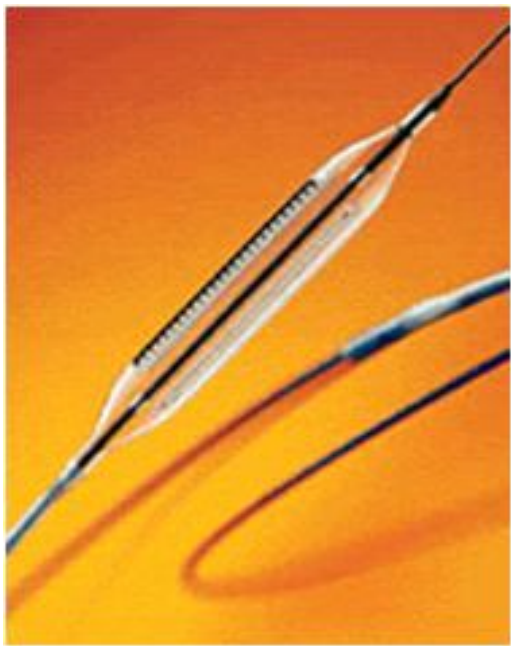
Balloon dilation and stent placement

In an effort to improve on the results of balloon dilation, a search for a more effective treatment was begun and led to the development of the stainless steel balloon-expandable stent. Stent placement is accomplished by positioning the stent across the narrowed segment of the artery. The stent is mounted on aballoon angioplasty catheter and covered with a sheath as it is moved into position. The sheath then is withdrawn off the stent-balloon angioplasty assembly and the balloon is inflated to its recommended pressure, expanding the stent and anchoring it in place.

The Cutting Balloon

This procedure is similar to standard balloon dilation but the balloon has been specially designed with small blades running up and down its length. When the balloon is inflated, the blades are activated and they cut through the narrowed area, making the vessel easier to dilate and resulting in a larger opening. Cutting balloons are available in different sizes.

Fig. 1: The Cutting Balloon Image with permission, from Boston Scientific Corporation



Surgery

Various methods of surgical repair of pulmonary artery stenosis are used, the choice of which depends on the characteristics of the stenosis and the surrounding vessels and other structures.

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