The Case of the Unlucky Heart

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A 58 year-old woman presents to the emergency department with severe dyspnea at rest, progressing over the last two days. She is quickly admitted to general medicine. She does not have a history of smoking, COPD, fever, cough, chest pain, or weight loss. She does have a positive history of a congenital heart defect, consisting of: a ventricular septal defect; right outflow tract obstruction, an overriding aorta, which receives blood from both ventricles, and ventricular hypertrophy. This patient also has a history of ventricular tachycardia and atrial fibrillation. Due to her arrhythmia she takes amiodarone and has a dual chamber pacemaker. Although amiodarone is a very effective anti-arrhythmic, it has many potential side-effects.

Questions

Q1: What is this congenital heart defect? How should it be treated?
Q2: What is the most likely cause of the pansystolic murmur at the LLSB?
Q3: What is your differential diagnosis?
Q4: What are the side effects of amiodarone? Could they be a factor?

Answers

A1: This is a tetralogy of fallot. The treatment is surgical correction consisting of closure of the ventricular septal defect and enlargement of the pulmonary artery outflow tract. Most patients then go on to be asymptomatic adults. [1,2]

On examination the patient: appears fatigued; has an elevated jugular venous pressure (JVP); abdominal ascites, a pulsatile liver; a pansystolic murmur over the lower left sternal border; coarse crackles at the lung bases; and edematous swollen ankles.

A2: The pansystolic murmur at the LLSB is likely that of tricuspid regurgitation. This is related to pressure overload in the right ventricle, related to the right ventricular hypertrophy caused by an outflow obstruction. These patients often also have an ejection murmur at the upper left sternal border from the right outflow tract obstruction. [1]

A3: This appears to be congestive heart failure; though the specific cause could be: acute or chronic right sided failure as a consequence of her congenital defect leading to left sided failure; silent ventricular infarction; interstitial lung disease and/or pulmonary secondary to amiodarone toxicity. Also included in the differential, would be: failed tetralogy of fallot repair; pulomonary vascular disease from long standing right heart failure; anemia, pleurisy, pneumonia, or pulmonary embolism. [1,3]

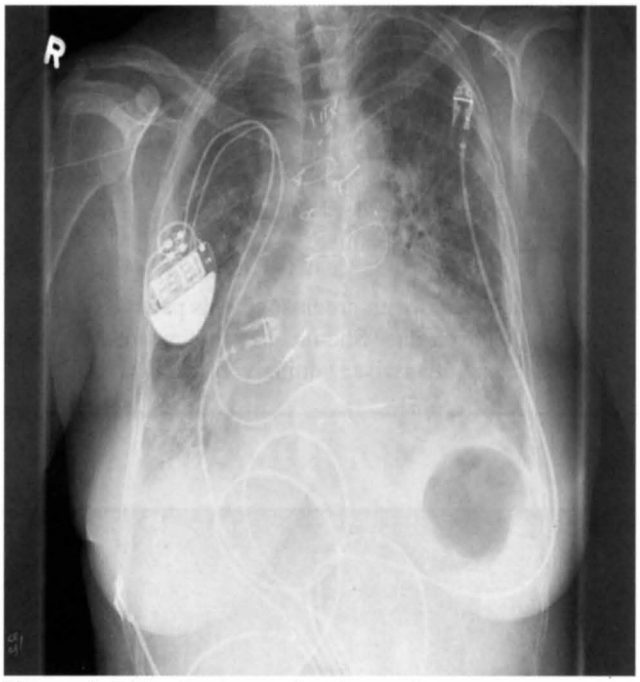

Figure 1. This chest x-ray has been taken at the emergency department. Note the florid pulmonary edema, cardiomegaly, wires from previous sternotomy, dual chamber pacemaker.

A4: Amiodarone works by prolonging the action potential duration and refractoriness of cardiac fibers. This results in a decreased rate of sinus node firing. It also suppresses automaticity, interrupts reentrant circuits, and prolongs PR, QRS, and QT intervals in addition to possesing vasodilatory and inotropic properties. Amiodarone is used for a wide range of arrhythmias. [1,3,4]

Its most serious side effect is pulmonary toxicity, probably from a hypersensitivity reaction. Other serious side effects relate to its cardiac toxicity; by causing bradycardia, aggravating ventiricular arrhythmia, and potentially precipitating congestive heart failure from the negative inotropic effects. A wide range of thyroid, neurological, and gastrointestinal disturbances can also occur.[1]

In this case, there is some dose related pulmonary toxicity causing pneumonitis, as well as congestive heart failure.

Consequently, she is taken off the amiodarone and responds to short term, high-dose prednisone to manage the pulmonary toxicity. Her consequent ECG tracing is shown in Fig. 2.

Questions

Q5: What type of rhythm does this ECG demonstrate?
Q6: What are the treatment options now, without amiodarone?

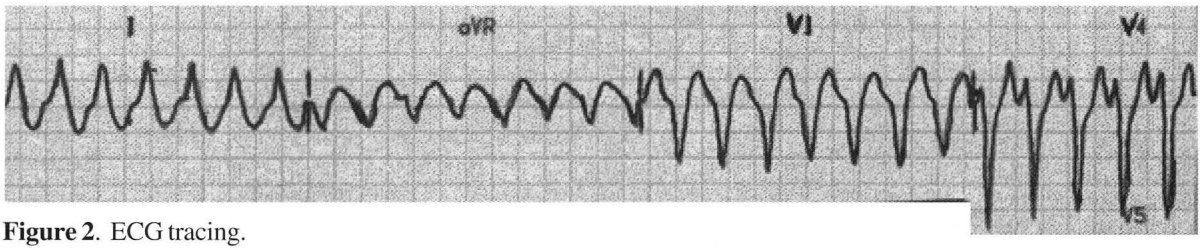

Figure 2. ECG tracing.

Answers

A5: This ECG clearly demonstrates ventricular tachycardia, (VT) resulting from her discontinuation of amiodarone. A wide QRS tachycardia is VT until proven otherwise. Ventricular arrhythmias are associated with a number of congenital heart defects, particularly tetralogy of Fallot. [5]

A6: Treatment of ventricular arrhythmias in this population include antiarrhythmic medications, catheter ablation, surgery, and implantable cardiac defibrillators. As seen in Figure 3, this patient, who needed a more chronic treatment, had an external defibrillator implanted. [1,2,4]

After being treated with prednisone, this patient's pneumonitis resolved. The new pacemaker/defibrillator controlled her ventricular tachycardia. She has had one episode of the defibrillator responding to an arrhythmia since its placement 4 months ago, and otherwise is doing very well.

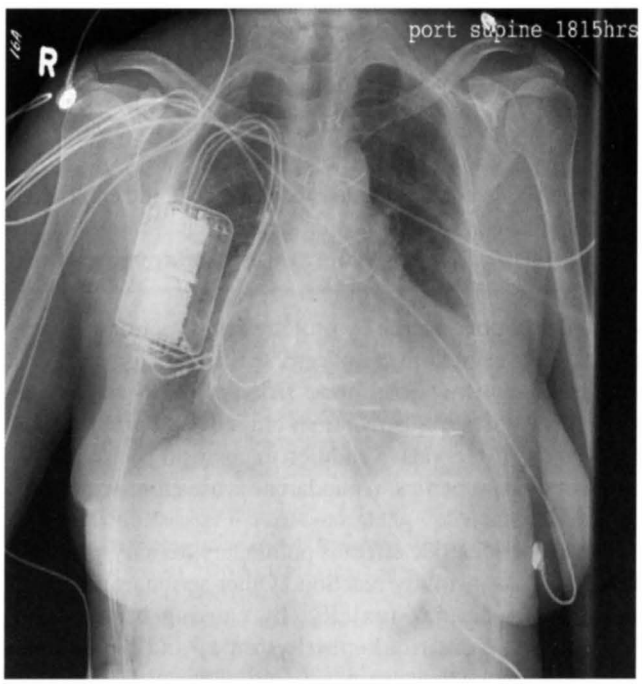

Figure 3. This x-ray, taken after a week in the hospital shows an implantable defibrillator/pacemaker. The pulmonary edema has mostly cleared up.

References

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