TEACHING FILES

A 10 years old girl with long standing heart disease

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Case Report: A 10 years old girl born of non-consanguineous marriage presented with puffiness of eyes, edema feet, exertional dyspnea and palpitations since 6 months. She had paroxysmal nocturnal dyspnea. There was no history of fever; however she had fever with bilateral knee joint pain 7 months back. She also had a history of a photosensitive rash over face on exposure to sun. There was no history of syncope or cyanosis. There was no history of Koch's contact. There were no urinary or neurological complaints. She had not been treated at all for the above complaints in the past. On examination, her pulse rate was 116/min; respiratory rate was 56/min. Her B.P. in right upper limb was 96/74 mm of Hg. She had pallor. There were no lymphnodes, raised JVP, edema feet or cyanosis. Joint examination was normal. On cardiovascular system, she had cardiomegaly with normal 1st and 2nd heart sound without murmur. She had hepatomegaly and other systemic examination was normal.

Investigations showed:

Hemoglobin = 12.7 gm/dl
WBC = 10,300/cumm
Platelet count = 4,35,000/cumm
ESR = 5 mm at end of 1 hour
BUN = 9 mg/dl
S. creatinine = 0.9 mg/dl
SGOT/SGPT = Normal
Urine = Normal
ASLO = Negative
ANA = positive (speckled pattern)
ds DNA = Negative
C_3 = 120 mg/dl (Normal)
S. Lactate and pyruvate = Normal

Echocardiography: - Dilated left ventricle with impaired contractility.
USG Kidneys: Right kidney = 6.1 x 2.5 cm**;** Left kidney = 8 x 3.9 cm
X-Ray chest = Normal
Mantoux test = 15 x 10 mm

Question: - What is the diagnosis?

Expert's Opinion - This child has a long standing untreated cardiac disease and echocardiography shows dilated left ventricle with impaired contractility suggestive of dilated cardiomyopathy. Though she had a history of fever with joint pains in past, there is no history of sore throat. Moreover, the joint pains were along with fever and not after fever. Also her ASLO is negative and there is no evidence of any valvular involvement. Thus, rheumatic fever seems unlikely.

She had history of a photosensitive rash, joint pains and has a positive ANA. However, she does not have any other evidence of SLE. Her urine, hemogram, C_3, are normal. Thus SLE leading to endocarditis is also unlikely.

From a metabolic point of view her serum lactate and pyruvate are normal but other metabolic disorders such as fatty acid oxidation defects, primary carnitine deficiency should also be kept in mind. Other possibility of viral myocarditis going into dilated cardiomyopathy should also be considered.

However, a significant finding on ultrasound of the kidneys should not be ignored. Her right kidney appears to be small in size. In a child with cardiac disease and small kidney, one should keep in mind, a possibility of aortoarteritis and renal artery stenosis. Though this child did not have hypertension or absent pulses, it may be due to the fact that the left ventricle was dilated and had impaired contractility leading to decreased stroke volume and decreased cardiac output and thus no hypertension. Also with her mantoux test being positive, possibility of aortoarteritis is quite high.

A colour Doppler of renal vessels as well as aorta is required. In this child, the colour Doppler showed bilateral renal artery stenosis. Infact, on treatment for her congestive cardiac failure, she developed hypertension confirming that on improvement of her cardiac output, her blood pressure also improved. She now needs to undergo angioplasty for her renal artery stenosis.

Total Answers: 44 Correct answers: 0

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