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A CASE OF CONGENITAL BICUSPID AORTIC VALVE WITH ANEURYSM OF ASCENDING AORTA WITH COMORBIDITIES

General Medicine

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ABSTRACT

Bicuspid aortic valve (BAV) is an inherited form of heart disease in which two of the leaflets of the aortic valve fuse during development in the womb resulting in a six-leaflet valve (bicuspid valve) instead of the normal nine-leaflet valve (tricuspid). BAV is the most common cause of heart disease present at birth and affects approximately 1.3% of adults. Here I present a case of congenital bicuspid aortic valve who also had a ascending aortic aneurysm and needed surgery due to the large diameter of the aneurysm.

KEYWORDS

INTRODUCTION:-

One of the most notable associations with BAV is the tendency for these patients to present with ascending aortic aneurysmal lesions. The extracellular matrix of the aorta in patients with BAV shows marked deviations from that of the normal tricuspid aortic valve. It is currently believed that an increase in the ratio of MMP2 (Matrix Metallopr oteinases 2) to TIMP1 (Tissue Inhibitor Metalloproteinases 1) may be responsible for the abnormal degradation of the valve matrix and therefore lead to aortic dissection and aneurysm. However, other studies have also shown MMP9 involvement with no differences in TIMP expression. The size of the proximal aorta should be evaluated carefully during the workup. The initial diameter of the aorta should be noted and annual evaluation with CT scan, or MRI to avoid ionizing radiation, should be recommended to the patient; the examination should be conducted more frequently if a change in aortic diameter is seen. From this monitoring, the type of surgery that should be offered to the patient can be determined based on the change in size of the aorta.[1]

Case Report:-

The patient was a married female aged 30 year who came with chief chief complaints of-

- Dyspnoea for the past few yrs.
- Palpitations for the past 1 year.
- Ghabrahat for the past 1 year.

The patient was alright few years back where she started experiencing dyspnoea which was more on exertion. It was grade II NYHA to start with but progreesed to Gr III NYHA in the recent times. There was no history of orthopnoea or PND. There was no history of any diurnal variation. There was no history of fever or cough.

Patient also had history of palpitations for the past 1 year, which was present intermittently. There was no h/o any giddiness, fall, syncope.

Patient also complained of ghabrahat off and on for the past 1 year. It had been present at times of stress. There was no history of chest pain.

Ther was no past history of similar illness. Patient was an old case of hypertension for previous 5 years. The treatment history showed nothing significant. The family history showed no history of diabetes mellitus, hypertension, coronary artery disease, tuberculosis. The personal history showed that the patient was vegetarian, no addictions, bowel and bladder were normal. The menstrual history was normal.

On general physical examination, the patient was conscious, cooperative, lying comfortably in the bed. The patient was afebrile. The pulse rate was 80/min, regular, other parameters normal. The respiratory rate was 18/min, thoracoabdominal. The BP was 110/70 mmHg in right arm supine. Pallor, icterus, cyanosis, clubbing, lymphadenopathy, edema were absent. JVP was not raised. On CVS examination, S1 was normal, A2 was soft, P2 loud, ejection click was present. Other systems examination was normal.

On investigation:-

Hb :- 12.3 g%, TLC :- 7000/mm3, Polys – 61%
Sr AST/ALT :- 28/12 IU/L
Bl urea :- 30 mg%, Sr Creat :- 1.1 mg%
PTI :- 1.0, RA Factor:- Negative
ESR : 10 mm I hr, ABG : normal
USG Abdomen :- normal, ECG : normal

CXR : normal RBS:- 42 mg%, Urine Exam: normal
Echocandiography :-

1)Bicuspid Aortic Valve
2) Severe pulmonary hypertension

CECT Thorax : Aneurysmal dilatation of the ascending aorta (5.5 cm diameter).

The patient was started on treatment and was sent to the cardiothoracic surgery for aneurysectomy. Surgery was done on the patient and she improved after that.

DISCUSSION:-

BAV frequently leads to significant complications in over one-third of affected individuals which often lead to significant morbidity and mortality. Notable complications of BAV include narrowing of the aortic valve opening, backward blood flow at the aortic valve, dilation of the ascending aorta, and infection of the heart valve. If aortic regurgitation and dilation of the ascending aorta are noted in someone with a bicuspid aortic valve, they should undergo yearly surveillance with transthoracic echocardiograms if the aortic root measures 4.5 centimeters or greater in diameter.[2] Bicuspid aortic valves may assume three different types of configuration.

1. "Real" bicuspid valves with two symmetric leaflets
2. A tricuspid architecture with a fusion of two leaflets
3. A tricuspid architecture with a fusion of three leaflets.

Most patients with bicuspid aortic valve whose valve becomes dysfunctional will need careful follow-up and potentially valve replacement at some point in life. Regular Echocardiography and MRI heart may be performed.[3]

Conclusion:-

Aneurysm of the aorta, when it occurs as a complication of congenital bicuspid aortic valve is a medical emergency as it can rupture and cause exsanguinations of the patient. So, it is in the best interest of the patient that we recognise this condition and give timely treatment.

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