CASE REPORT

Ebstein's anomaly with recurrent massive pericardial effusion: a rare presentation

Nirdesh Jain,¹ Dharam Prakash Saran,² Sharad Chandra Yadav,² Sudhanshu Dwivedi³¹Department of Cardiology, CSMMU (earlier KGMU), Lucknow, Uttar Pradesh, India²Department of Cardiology, King George's Medical University, Lucknow, Uttar Pradesh, India³Department of Cardiology, KGMU, Lucknow, Uttar Pradesh, India**Correspondence to**Dr Nirdesh Jain,
nirdeshjain8@gmail.com

Accepted 11 July 2016

SUMMARY

Ebstein's anomaly is known for its varied presentations and exceptionally long survival in some cases. Here the authors describe a case of previously undiagnosed Ebstein's anomaly in a patient who presented with massive pericardial effusion. Over the next 6 months, the patient was hospitalised twice for pericardiocentesis. To the best of our knowledge, a case of Ebstein's anomaly with recurrent massive pericardial effusion has not been described previously.

CASE PRESENTATION

A 42-year-old woman presented with symptoms of progressive dyspnoea for 2 years and generalised body swelling for 2 months. She had no history of cyanosis. Her blood pressure was 100/60 mm Hg, pulse rate 120/min, and irregular and pulsus paradoxus was present. Pedal oedema was present. Cyanosis (peripheral oxygen saturation 95%) and clubbing were absent. Jugular venous pressure was raised with absent and descent. Auscultation revealed muffled heart sounds.

INVESTIGATIONS

ECG of the patient revealed low voltage complex with atrial fibrillation (figure 1A). Chest X-ray revealed cardiomegaly with globular heart, small aortic root, a small main pulmonary artery shadow and decreased pulmonary vasculature (figure 1B). Echocardiography revealed massive pericardial effusion to an extent that the cardiac chambers could not be delineated precisely (figure 2A and video 1). Emergent pericardiocentesis was performed by the subcostal route and 1.5 L of pericardial fluid was drained. Echocardiography after pericardiocentesis showed apical displacement of the septal and posterior leaflets of the tricuspid valve (12 mm/m²) with severe tricuspid regurgitation and right atrial enlargement with small right ventricle (RV; figure 2B and video 2). The left atrium and left ventricle were within normal limits and left ventricular ejection fraction was 64%. Systolic function of the RV was markedly reduced with the RV fractional shortening being 20%. Pericardial fluid was straw coloured, total cell count was 60/mL with 40% neutrophils and 60% lymphocytes, protein was 2.2 g/dL (serum

BACKGROUND

Ebstein's anomaly is an uncommon congenital malformation of the tricuspid valve which occurs in 1/20 000 live births and accounts for 1% of all cases of congenital heart disease.¹ It was first described by Wilhelm Ebstein in 1866.² The hallmark of Ebstein's anomaly is apical displacement of the septal leaflet of the tricuspid valve from the insertion of the anterior mitral leaflet by at least 8 mm/m² body surface area.³ Ebstein's anomaly has a broad clinical spectrum depending on age of presentation, haemodynamics, anatomic severity and degree of right-to-left shunt.⁴ No two patients of Ebstein's anomaly have similar presentation. Radiologically, Ebstein's anomaly closely mimics massive pericardial effusion. Coexistence of Ebstein's anomaly with a haemodynamically significant pericardial effusion is highly uncommon and only one such case has been reported to the best of our knowledge.⁵ Here we report a rare case of Ebstein's anomaly who presented with recurrent massive pericardial effusion.

Figure 1 ECG of the patient revealing low voltage complex with atrial fibrillation (A). Chest X-ray posteroanterior view showing cardiomegaly with globular heart, small aortic root and main pulmonary artery shadow and decreased pulmonary vasculature (B).

CrossMark

To cite: Jain N, Saran DP, Yadav SC, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-216176

Figure 2 Echocardiogram apical view showing large pericardial effusion lateral to left ventricle (before pericardiocentesis) (A). Echocardiogram apical view showing apical displacement of tricuspid leaflet with atrialised ventricle (after pericardiocentesis) (B). LA, left atrial; LV, left ventricle; PE, pericardial effusion; RA, right atrium; RV, right ventricle.

protein was 4.3 g/dL, albumin was 1.3 g/dL (serum albumin was 2 g/dL), sugar was 120 mg/dL and LDH was 120 IU/L (serum lactate dehydrogenase was 1295 IU/L). All features were suggestive of transudative effusion. Malignant cytology of pericardial effusion was negative. PCR for mycobacteria was also negative. Systemic causes of pericardial effusion were ruled out by finding a negative antineutrophil antibody titre, normal thyroid and renal function. Contrast-enhanced CT of the thorax and ultrasonography of the abdomen failed to find out any malignant focus.

DIFFERENTIAL DIAGNOSIS

Massive pericardial effusion.

TREATMENT

Considering a transudative effusion with normal left ventricle and depressed RV systolic function along with failure to find a secondary cause, pericardial effusion was attributed to right-sided heart failure, hence optimal heart failure therapy was started.

Video 1 Echocardiogram apical view showing large pericardial effusion lateral to left ventricle (before pericardiocentesis).

Video 2 Echocardiogram apical view showing apical displacement of the tricuspid leaflet with atrialised ventricle (after pericardiocentesis). LA, left atrial; LV, left ventricle; RA, right atrium; RV, right ventricle.

OUTCOME AND FOLLOW-UP

While awaiting definite surgery she had recurrent pericardial effusion which required pericardiocentesis twice over the next 6 months along with optimisation of the diuretic dose.

DISCUSSION

Clinical presentation of Ebstein's anomaly is variable but it usually presents with cyanosis, right-sided heart failure and arrhythmia. While cyanosis and right-sided heart failure are more common in paediatric population presentation with arrhythmia it is more common in adults.⁶ In the era of cardiovascular intervention the survival of Ebstein's anomaly has increased but still <50% of patients survive beyond 10 years.⁷ The index case presented first time at 42 years with right-sided heart failure and massive pericardial effusion making it an unusual presentation. ECG is often abnormal in Ebstein's anomaly and shows tall and broad P wave, complete or incomplete right bundle branch block, bizarre terminal QRS complexes and atrial fibrillation.⁶ In our patient the only obvious ECG abnormality was low voltage complex (in precordial and limb leads) and atrial fibrillation. Chest radiography of Ebstein's anomaly usually shows a globe-shaped heart with a narrow waist that closely resembles massive pericardial effusion. In our case both conditions coexisted. Echocardiography is the diagnostic test of choice in such patients and it readily establishes

the diagnosis. In the present case an inconspicuous RV with huge right atrium and low pressure tricuspid regurgitation made the diagnosis of Ebstein's anomaly obvious. Such large pericardial effusion due to right-side heart failure is unusual.

Here the authors tried to rule out all differential diagnoses which might have caused the transudative pericardial effusion. Our patient responded well to diuretics therapy which also favoured right-sided heart failure as the possible aetiology of pericardial effusion. Definite management of Ebstein's anomaly is surgical repair while those patients with heart failure who are not candidates for surgical repair should be managed with optimum heart failure therapy.

Acknowledgements The authors would like to acknowledge professor V S Narain, head of the department of cardiology for helping us in the clinical case discussion and chest X-ray reading.

Contributors NJ gave the concept of the manuscript. The literature was searched by DPS and SCY. All authors contributed in drafting and final review as well as revision of the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- Correa-Villaseñor A, Ferencz C, Neill CA, *et al.* Ebstein's malformation of the tricuspid valve: genetic and environmental factors. The Baltimore-Washington Infant Study Group. *Teratology* 1994;50:137–47.
- Ebstein W. Ueber einen sehr seltenen Fall von Insufficienz der Valvula tricuspidalis, bedingt durch eine angeborene hochgradige Missbildung derselben. *Arch Anat Physiol* 1866;7:238–54.
- Edwards WD. Embryology and pathologic features of Ebstein's anomaly. *Prog Pediatr Cardiol* 1993;2:5–15.
- Giuliani ER, Fuster V, Brandenburg RO, *et al.* Ebstein's anomaly: the clinical features and natural history of Ebstein's anomaly of the tricuspid valve. *Mayo Clin Proc* 1979;54:163–73.
- Llamas-Esperón G, Alemán-Villalobos R, Zamora-Muciño A, *et al.* Ebstein's anomaly related with significant pericardial effusion. Report and analysis of a case. *Gac Med Mex* 1998;134:479–86.
- Celermajer DS, Bull C, Till JA, *et al.* Ebstein's anomaly: presentation and outcome from fetus to adult. *J Am Coll Cardiol* 1994;23:170–6.
- McElhinney DB, Salvin JW, Colan SD, *et al.* Improving outcomes in fetuses and neonates with congenital displacement (Ebstein's malformation) or dysplasia of the tricuspid valve. *Am J Cardiol* 2005;96:582–6.

Learning points

- ▶ Ebstein's anomaly with massive pericardial effusion is difficult to delineate by transthoracic echocardiography.
- ▶ Multiple hospitalisations for massive pericardial effusion in the index case of Ebstein's have made the notion that right-sided heart failure can present as massive pericardial effusion.
- ▶ Ebstein's anomaly should always be kept in mind as differential diagnosis of a right-sided heart failure irrespective of the age of the patient.

Copyright 2016 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow