Letter to the Editor

Chronic Myeloid Leukemia Associated with Ebstein's Anomaly

To the Editor,

Ebstein's anomaly is the most common congenital abnormality of the tricuspid valve and accounts for about 0.5% of cases of the congenital heart diseases. It has occasionally been associated with tumours of the heart and other neoplasms. Here we report a case of chronic myeloid leukemia (CML) occuring in a patient with Ebstein's anomaly.

A 14 year old boy presented with abdominal distension of four months duration. He did not gave any history of any other systemic complaints including exertional breathlessness, palpitation or dizzy spells. There was no family history of any significant illness. Physical examination showed cyanosis and prominent v wave in the JVP. There was a pansystolic murmer in the tricuspid area. Liver was palpable 3 cm and spleen was palpable 6 cm below the costal margin. Hemogram showed hemoglobin of 12.5 g/dL, white cell count of 122 x 109/L and platelet count of 344 x 109/L. Leukocyte alkaline phosphatase (LAP) score was 4. Peripheral smear and bone marrow findings were consistent with features of chronic myeloid leukemia in chronic phase. Karyotype showed Ph' positivity in all the 20 metaphases analysed. Echocardiogram (ECG) showed features of right-axis deviation and right bundle branch block with type B WPW syndrome. ECG showed enlarged right atrium and small atrial septal defect with bi-directional flow. Septal leaflet of tricuspid valve was displaced down and the antirer leaf let of the tricuspid valve was large-typical of Ebstein's anomaly. There was severe tricuspid regurgitation and moderate

mitral regurgitation. He was treated with hydroxyurea and is continuing in hematologic remission.

Neoplasms have been reported to occur rarely in cases of Ebstein's anomaly. Russel, et al, reported one case of rhabdomyoma of the heart with Ebstein's anomaly^[1]. Extracardiac tumours like plasmacytoma and mesothelioma of the pericardium has also been reported^[2,3]. Leukemia has never been reported previously. A common etiology is not possible as CML is an acquired stem cell disorder. Whether Ebstein's anomaly carries an increased risk of malignancy cannot be ascertained as there are only few case reports.

REFERENCES

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