

blastomas appear predominantly hypodense (occasionally isodense or mixed density) and often contain coarse, dense calcification. Central cystic or necrotic zones and hemorrhage are frequently seen. The location and peritumoral edema are variable. In our Case 2, apart from the diagnosis of primary intracerebral neuroblastoma, anaplastic astrocytoma glioblastoma, lymphoma, malignant meningioma and metastasis were considered preoperatively in the differential diagnosis. Although it may be difficult to distinguish a neuroblastoma from an astrocytoma by CT, the combination of intracerebral calcification, hemorrhage and location near the ventricle in a young patient should suggest primary neuroblastoma.

MRI demonstrates the macroscopic aspects more clearly and also shows strong enhancement after Gd-DTPA. However, these macroscopic features are rather non-specific and can also be seen in ependymomas and medulloblastomas(4).

Histologically primary cerebral neuroblastomas have a close resemblance to the much more common neuroblastomas of the adrenal medulla and sympathetic ganglia, as well as to cerebellar medulloblastomas. Grossly they appear as well circumscribed, rounded parenchymal masses which frequently contain areas of cystic change, necrotic foci and large intratumoral calcifications. The histopathological diagnosis based on light microscopy alone may often be difficult. Poorly differentiated oligodendrogliomas, primary cerebral sarcomas and other primary neuroectodermal tumors must be considered. The diagnosis may be confirmed by electron microscopy showing dense core vesicles(5) and immunocytochemical procedures demonstrating synaptophysin, which is the evidence of synapses or neurofilaments.

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Mitral Valve Prolapse with Wolff-Parkinson-White Syndrome

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Mitral valve prolapse syndrome (MVP), one of the most prevalent cardiac valvular abnormalities affecting 5-10% of

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the population, has not been extensively reported in Indian literature(1). Wolff-Parkinson-White Syndrome (WPM) in infants and children has also been repeatedly surveyed in the European and American literature(2,3). The combination of the conditions has been reported a couple of times in Western literature(4,5). We present a case of MVP with WPW syndrome.

Case Report

A 10-year-old girl with no previous cardiac symptoms was admitted with history of palpitations and dull aching left precordial chest pain of two days duration. No past history of breathlessness, syncope or rheumatic fever was forthcoming. On examination she was afebrile with no obvious anemia or signs of congestive cardiac failure. Her heart rate was 220/min, without any murmur. A diagnosis of supraventricular tachycardia (SVT) was made and confirmed by ECG. Hemogram and chest

X-ray were normal. She was given oral digoxin, half of the digitalising dose immediately and one-fourth after 8 hours. By the next morning her heart rate had settled. At this time, on auscultation she had a split second sound with an ejection systolic murmur. A repeat ECG showed the classical changes of WPW syndrome (short PR interval, a delta wave and wide QRS complex) as well as inverted T waves in leads II, III, aVF and V_4 to V_6 and QT prolongation (Fig.) characteristic of MVP. The digoxin was stopped. On echocardiography the diagnosis of MVP was confirmed both in B and M mode. Two subsequent ECGs taken on weekly follow up were consistent with the ECG taken on the second day. She remained symptom free and off drugs from 24 hours after admission till the subsequent follow ups.

Discussion

WPW is a pre-excitation syndrome due

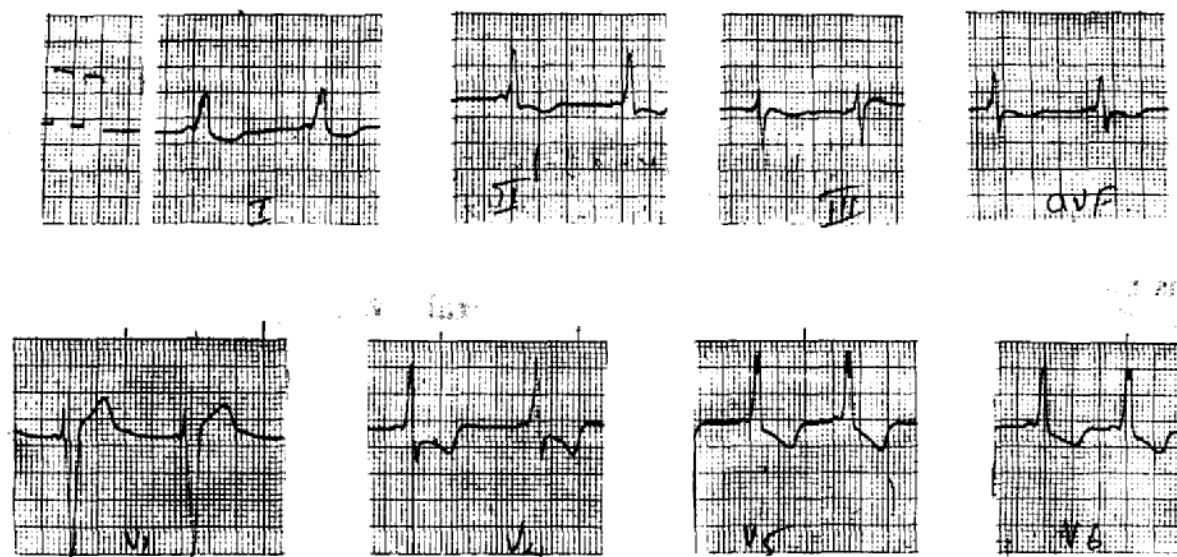


Fig. ECG showing short PR interval, QRS complex with delta wave and T wave inversion in leads II, III, aVF, V_4 , V_5 and V_6

to accessory pathways (bundles of Kent) in the conduction of system of the heart. Its incidence in the general population is unknown. Among hospitalized children it ranges from 0.04 to 0.31%. Even in children with congenital heart disease the incidence is only 0.27 to 0.86%(2). The criteria for diagnosis are short PR interval for age, with the initial slurring of the QRS complex (the 'delta wave') on ECG. Rosenbaum classified this pattern into Type A: the major QRS deflection in lead V_1 is positive, predicting a left lateral accessory connection, and type B: negative major QRS deflection in V_1 predicting a right sided accessory connection(6). WPW syndrome may be present without any heart disease or in association with a spectrum of congenital heart disease (Ebstein anomaly, coarctation of the aorta, tricuspid atresia, atrial and ventricular septal defects, patent ductus, dextrocardia, corrected transpositions, tetralogy of Fallot, pulmonary stenosis)(2,3,7), as well as with familial cardiomyopathy(2,7). In rheumatic heart disease it has been noted during a recurrence of rheumatic carditis(2). It has also been reported in association with some neurological disorders like mental retardation and Friedreich's ataxia, and other congenital anomalies like branchial cyst, *etc.*(2). Most commonly it is noted with Ebstein anomaly when 90% of the cases have Rosenbaum type B pattern. The present case also had the ECG features of type B anomaly.

The WPW syndrome is a benign condition with no subjective manifestations or hemodynamic alternations as long as there are no ectopic tachyarrhythmias(5). Our child was symptom free until she was 10 years and developed SVT. Recurrent SVTs do occur; in the absence of primary cardiac disease, the prognosis is good(3). Paradoxically, digitalis is effective in the treatment

of SVT associated with WPW syndrome. Recently propranolol and lidocaine are mentioned as the drugs of choice; if SVT is refractory, DC shock may be needed. Simple Valsalva maneuver may also be of help if used early(5).

MVP is an auscultatory syndrome of either a mid systolic click, a late systolic murmur or both, with a prevalence of 1.4 to 8% in the normal population(8). The ECG changes include inverted or biphasic T waves and nonspecific ST segment changes in lead II, III, aVF and occasionally anteriolateral leads as well(4). M-mode echo criteria for MVP are mid systolic buckling and pansystolic hammocking. The ECG and echo findings were very characteristic in our patient. SVT is the most common tachyarrhythmia in MVP. What triggered SVT in the present case is difficult to say. If there is family history of MVP, or T wave, and QT abnormalities associated with arrhythmias in the ECG, then the individual is at risk for infective endocarditis and may need prophylaxis(8).

WPW syndrome and MVP need to be further studied for their interrelationship, specially in Indian children, as there is paucity of reporting. Regular ECG and ECHO studies in children presenting with arrhythmias are advocated for this purpose.

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Pulmonary Alveolar Microlithiasis in Siblings

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Pulmonary alveolar microlithiasis is a rare disease of unknown etiology, in which

calcium phosphate crystals are deposited throughout the lungs. We report 2 cases of pulmonary alveolar microlithiasis occurring in siblings.

Case Reports

Case 1: A 11-year-old male child presented with symptoms of increasing weight gain for the past 6 months. A routine chest X-ray (*Fig. 1*) showed bilateral diffuse miliary like mottling of both lung fields, almost obliterating the cardiac silhouette. The radiological picture was suggestive of a diffuse interstitial lung disease and possibilities of miliary tuberculosis, pulmonary alveolar proteinosis, fibrosing alveolitis, pulmonary hemosiderosis and pulmonary alveolar microlithiasis were considered. Careful examination of the cardiovascular, respiratory and other systems did not reveal any significant findings. There was no family history of any respiratory problems except that the child's grandfather had died of 'asthma' at 60 years of age.

On investigations hemogram and routine investigations were normal; serum Na^+ was 138 mEq/L, K^+ 4.9 mEq/L, calcium 7 mg/dl and Cl^- 100 mEq/L. Gastric lavage for acid fast bacilli was negative. Arterial blood gas showed evidence of hypoxia (pH 7.37, PO_2 64.7 mm Hg, PCO_2 37.7 mm Hg; TCO_2 22.9 mmol/L; O_2 sat 91.6%; HCO_3^- 21.7 mmol/L). Pulmonary function tests showed a restrictive airways

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