Man, 5th edn. Baltimore, The Johns Hopkins University Press, 1979, p 82.

5. Eiler JL, Mortin JM. Bizarre deformitis in offspring of user of LSD. N Eng J Med 1970, 283: 395-397.

6. Beighton P. Vertebral Dysostoses. *In:* Inherited disorders of the Skeleton, 2nd edn. Ed Beighton P. Edinburgh, Churchill Livingston 1988, pp 316-319.

7. Jarcho S, Levin PM. Hereditary malformation of the vertebral bodies. Bull John Hopkins Hosp 1938, 62: 216-226.

## Benign Cystic Teratoma of the Mediastinum

**S.P. Jajoo**
**V.P. Dandge**
**A.V. Jayakar**
**W.M. Dalwai**
**S.C. Karande**
**P.B. Dharap**

Cysts and tumors of the mediastinum may originate from any of the structures contained therein or as a result of a developmental abnormality and are rare in childhood(1). Willis(2) defined a teratoma as a true tumor or neoplasm, composed of multiple tissues of kinds foreign to the part in which it arises. The components are derived from all three embryonic layers and may contain mature or immature skin, hair, teeth, bone, cartilage, CNS, respiratory or alimentary tissue.

In our case, the teratoma contained pancreatic tissue, which is an unusual finding(3). Also, in the last decade, although few cases of teratomas have been reported in Indian literature(4-7), to our knowledge, there has been no case report of a mediastinal teratoma containing pancreatic tissue in a child.

### Case Report

A 3-year-old boy, weighting 8 kg, was admitted with complaints of dry brassy cough and progressively increasing breathlessness since last one year, with exacerbation of symptoms for the last 15 days. Clinical examination revealed a conscious, well-oriented child with heart rate 140/min and respiratory rate of 64/min. There was no flushing of the face, dilated veins or edema of head and neck. On chest examination, we noticed a marked left sided precordial bulge with apex beat not well visualized. The trachea was shifted to the right side and pulsations were seen in the right parasternal region. Percussion revealed a dull note in the anterior left hemithorax, extending from the 2nd to 6th intercostal space, right upto the anterior axillary line. The left border of the heart could not be delineated, and on the right side the dull note extended upto 1 cm parasternally. Heart sounds were normal, but heard on the right side of the parasternum. Other systems were normal. The development milestones were normal.

Chest roentgenogram revealed a large mass in the left hemithorax with a mediastinal shift to the right (*Fig. 1*). Two-dimen-

*From the Department of Pediatrics and Pathology, T.N. Medical College and B.Y.L. Nair Hospital, Bombay 400 008.*

*Reprint requests: Dr. V.P. Dandge, Professor of Pediatrics, T.N. Medical College, Bombay 400 008.*

*Received for publication: November 6, 1991;*
*Accepted: January 2, 1992*

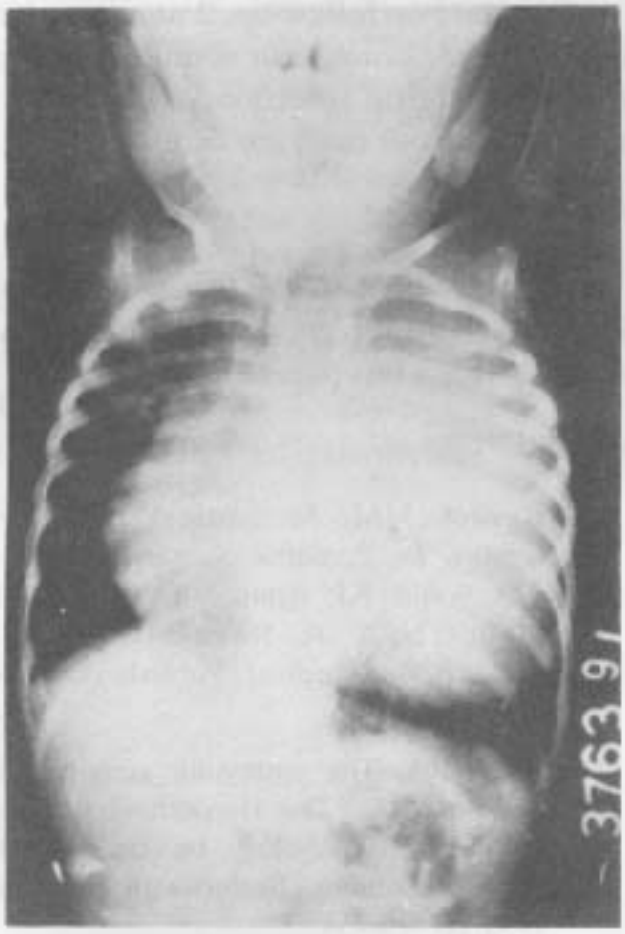

*Fig.1. Chest roentgenogram showing intrathoracic mass in the left hemithorax.*

sional echocardiogram showed a large cystic echoluscent mass in the anterior mediastinum, not continuous with the cardiac chambers. CT scan thorax confirmed the diagnosis of a cystic mass in the superior and anterior mediastinum in the left hemithorax (*Fig. 2*).

An exploratory thoracotomy was done and 350 ml of gelatinous yellowish fluid was initially aspirated from the mass, to enable its total resection. Post-operative period was uneventful. The histopathology of the mass showed features of a mature benign cystic teratoma, with tissues including gastric epithelium, respiratory epithelium, squamous epithelium like skin and hair, cartilage bits and even well formed pancreatic tissue.

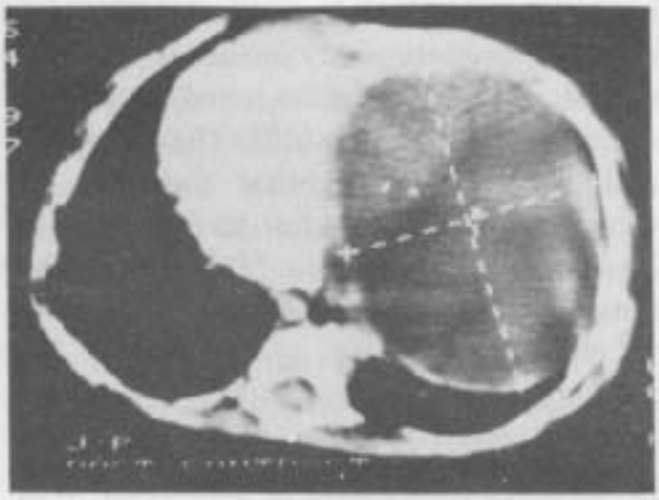

*Fig. 2. CT scan chest showing cystic mass in left anterior hemithorax.*

**Discussion**

In the mediastinum germ cell, tumors, *viz.*, dermoid cysts and teratomas are second in frequency to tumors of neurogenic origin, *viz.*, neurofibromas, ganglineuromas, neurilemomas, ganglioneuroblastomas and neuroblastomas(1). Other tumors found in the mediastinum include thymic tumors; cysts such as bronchogenic, enteric and pericardial; lymphomas; and hemangiomas(1).

Among the teratoid tumors, the benign cystic teratomas are more common than the solid ones and are often situated in the anterior mediastinum(8). Teratomas may reach enormous size. The symptoms most frequently encountered are chest pain, cough, respiratory distress, hemoptysis, dysphagia and weight loss. Remarkably enough, a large number even when large, are asymptomatic and are only incidentally found on chest roentgenograms(1). As with other mediastinal tumors, discovery is the indication for their removal, because continued tumor growth leads to serious symptoms from pressure and displacement of mediastinal structures. In addition, infection of these tumors or erosion into other structures may produce dangerous conse-

quences and greatly complicate operative removal(9). Until either infection or malignant degeneration has occurred, teratomas are quite readily removable. The criteria of malignancy are somewhat uncertain in view of the bizarre admixture of various elements. Perhaps 20 to 25% of the solid teratomas have been malignant(1).

The presence of adrenal, renal and pancreatic tissue in teratomata is uncommon(10). Berry *et al.*(10) in their review of 91 cases of teratomata in childhood from 1934 to 1969, found that the most frequent site of origin of teratomas is the sacrococcygeal area (63.7%), followed by the gonads (18.7%), mediastinum (5.5%), with less frequent sites being intracranial (3.3%), thyroid (3.3%), stomach (2.2%), palate (2.2%) and retroperitoneal (1.1%). None of their 5 mediastinal teratomas showed any pancreatic tissue. Honicky and de Papp(3) were the first to report a benign insulin-producing teratoma of the mediastinum. This 3-year-old boy had a chest roentgenogram done for dry cough. Preoperatively random blood glucose levels varied from 38 mg/dl to 63 mg/dl. Plasma insulin level was 45 $\mu$U/ml (normal 1 to 30 $\mu$U/ml) at the time hypoglycemia was documented. This asymptomatic hypoglycemia was later explained by presence of well formed pancreatic tissue in the excised teratoma.

In a child with an anterior mediastinal mass, a low blood glucose level might be a clue to the diagnosis(10). In our case, we did not document any hypoglycemia preoperatively. Random blood glucose levels were normal. Also, the presence of pancreatic tissue was revealed on histopathology 8 days after excision of the teratoma. Since plasma insulin levels even if raised earlier, would have returned to normal by this time, this expensive test was not done.

Our child on follow-up, 2 months later, is doing well. Long term results following complete surgical resection in benign teratomas, as in our case, are excellent(10).

**Acknowledgement**

The authors thank the Dean, Dr. (Mrs) K.D. Nihalani, for granting them permission to publish this paper.

**REFERENCES**

1. Ravitch MM. Mediastinal cysts and tumors. *In:* Pediatric Surgery, 4th edn. Eds Welch KJ, Randolph JG, Ravitch MM, O'Neill JA, Rowe MI. Chicago, Year Book Medical Publishers 1986, pp 602-618.
2. Willis RA. The embryonic tumors and teratomas. *In:* The Borderland of Embryology and Pathology, 1st edn. Ed Willis RA. London, Butterworth and Co, 1958, pp 410-454.
3. Honicky RE, de Papp EW. Mediastinal teratoma with endocrine function. Am J Dis Child 1973, 126: 650-653.
4. Mathur NB, Raicada RM, Bhatia BD, Dubey AP, Chaturvedi P, Dhabate S. Asphyxiating nasopharyngeal teratoma at birth. Indian Pediatr 1985, 22: 325-326.
5. Narasimharao KL, Mohanthy HK, Mitra SK. Posterior mediastinal teratoma. Indian Pediatr 1985, 23: 469-470.
6. Murthy TVM, Bhargava RK, Sood KC, Rakas FS, Kamble G. Cervical teratoma in newborn. Indian Pediatr 1989, 26: 600-603.
7. Sharma V, Newton G. Intracranial teratoma in early infancy. Indian Pediatr 1991, 28: 70-72.
8. Mahour GH, Landing BH, Woolley MM. Teratomas in children. Clinicopathologic studies in 133 patients. Z Kinderchir 1978, 23: 365-380.

9. Thompson DP, Moore TC. Acute thoracic distress in childhood due to spontaneous rupture of a large mediastinal teratoma. J Pediatr Surg 1969, 4: 416-423.

10. Berry CL, Keeling J, Hilton C. Teratomata in infancy and childhood: A review of 91 cases. J Pathol 1969, 98: 241-252.

# Wilson's Disease: Initial Worsening of Neurologic Syndrome with Penicillamine Therapy

**A. Kher**
**B.A. Bharucha**
**N.B. Kumta**

Wilson's disease was described in 1912(1). It results from expression of a locus on chromosome 13(2). Interpretation of diagnostic tests often pose problems especially in heterozygote detection. D-penicillamine, the gold standard of therapy was introduced by Walshe in 1956(1). This was proved quite dangerous at times as exemplified by this report.

## Case Report

A 10-year-old girl presented with neurologic deterioration in the form of unsteady gait, worsening of handwriting and speech over two years. She had no prior history of jaundice or neurological disorder. On nervous system examination, she had dysarthria, slurred speech, inco-ordination, unsteady gait and brisk tendon jerks. There were no involuntary movements, focal deficits or cranial nerve palsies. There was no hepatosplenomegaly. A Kayser Fleischer ring was detected unilaterally and confirmed on slit lamp. Serum copper (Cu), ceruloplasmin, and urine copper values were 50 $\mu$g/dl (n 70-160 $\mu$g/dl), 0.032 OD (n 0.25-0.49 OD) and 401.85 $\mu$g/dl (n 40 u/day) respectively. Liver functions and tests for renal tubular involvement were normal. The patient was put on 250 mg six hourly of D-penicillamine (40 mg/kg/day) with a diet restricted in copper.

The child had an acute neurologic worsening in the form of loss of sensorium, speech and all higher functions and frequent vomiting within 15 days of instituting the above therapy. Biochemical parameters, cerebrospinal fluid examination and CT scan of the brain failed to reveal any abnormality. She slowly recovered with mannitol, dexamethasone and oral glycerol therapy given to treat cerebral edema. D-penicillamine was omitted during this time and restarted after 15 days in a low dose of 125 mg 12 hourly (10 mg/kg/day) with a daily dose of 100 mg pyridoxine. A similar episode occurred again within 12 days of restarting D-penicillamine and the child improved with similar treatment. D-penicillamine was reintroduced in the dose of 25 mg only (1 mg/kg/day). It was gradually increased to 250 mg 12 hourly (20 mg/kg/day). Urine copper was closely monitored and this dose was tolerated well.

## Discussion

D-penicillamine acts by promoting

*From the Department of Pediatrics, Seth G.S. Medical College and K.E.M. Hospital, Parel, Bombay 400 012.*

*Received for publication: August 16, 1991; Accepted: February 5, 1992*