Acute Intermittent Porphyria

I read with interest the recent case report of acute intermittent porhyria (AIP)(1). The authors have rightly stressed the rarity of this disorder in the pediatric age group. It was in the year 1967 that we reported a case of AIP in a child(2). At that time we had reviewed the world literature (39 references) and came across references to only 13 cases in the pediatric age group. According to the present report, in all 37 cases had been recorded till 1974. It is, therefore, reasonable to infer that on an average only three cases of AIP in the pediatric age group per year appear in the world literature.

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Pulmonary Blastoma

Pulmonary blastoma is an extremely uncommon intrathoracic neoplasm in children. It was first described by Barrett and Barnard in 1945. Since then over 120 cases have been reported with about one-fifth occuring in children(l-5). Because of rarity of pulmonary tumor in children, they are not commonly considered in the differential diagnosis and are treated as infection. We report clinical features, radiological findings and treatment of a child with pulmonary blastoma.

A 9-year-old boy presented with history of chest pain, low grade fever, weight loss,, decreased appetite, progressive pallor and dyspnea for two and half months. The patient had dysphagia to solids and engorgement of neck veins for one month. He received antitubercular therapy for 2 months from a general practitioner with a presumptive diagnosis of tuberculosis. There was no response to this therapy. On examination, he had tachycardia (pulse rate 160/min), tachypnea (respiratory rate 60/min) and mild pallor. Neck veins were engorged with absence of any wave form and bilateral conjunctival congestion. On examination of chest, trachea was shifted to the right. The left hemithorax was more prominent, percursion note was dull, air entry was reduced and vocal resonance decreased. Right side of the chest was normal. There was no hepatosplenomegaly.

X-ray film of chest revealed an opaque left hemithorax and CT scan of chest showed a large soft tissue mass with areas of necrosis and calcification occupying almost the entire left hemithorax. Mediastinal structures were displaced to the right and posteriorly. An ultrasound guided bi-

opsy was performed. It revealed an undifferentiated round cell tumor with areas of necrosis. The material was negative for neuron specific enolase and faintly positive for desmin and myoglobin. Based on the findings on histopathology and CT chest, a diagnosis of pulmonary blastoma was made. The patient was treated with combination chemotherapy including vincristine, cyclophosphamide and actinomycin-D. Despite chemotherapy and supportive care, he succumbed to his illness on day 10 of hospitalization.

Pulmonary blastoma is a mixed tumor containing both mesenchymal and epithelial malignant elements. Histological features are similar in appearance to a fetal lung of 3 months gestation(5). Clinical features and findings on X-ray film of chest may be nonspecific. Computerized axial tomographic scan is useful to investigate the origin, extent and consistency of neoplasm. Intraneoplastic calcifications suggest the possibility of neuroblastoma or pulmonary blastoma. The neuroblastoma can be differentiated by measurement of urinary catecholamines. An open or thoracoscopic biopsy or needle biopsy and fine needle aspiration cytology with histochemistry may help to differentiate these tumors(l,2,5).

There is no single large experience, in the management of pulmonary blastoma. Combination chemotherapy with agents active against both sarcomatous (Vincristine, cyclophospamide, d-actinomycin) and carcinomatous elements (doxorubicin and cisplatinum) and radiotherapy have been attempted with some evidence of objective response. The prognosis remains poor with overall response rate ranging between 20-40%(4-7).

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