

Quiz Section

Answer to Quiz No 1 (PMR Vol 1 No 01 March 2009)

Answer to Quiz No. 3(ECG)

1. *What is the diagnosis?*
Pleuropulmonary blastoma (PPB)
2. *What more could have been done after surgery at 6 months of age?*
Post-operative chemotherapy should be added in the treatment protocol.
3. *How will you treat it?*
Palliative chemotherapy on the line of soft tissue sarcoma.

Discussion

Manivel et al first described the term, PPB, which includes tumours that has previously been described as pulmonary blastoma, pulmonary sarcoma, embryonal sarcoma, pulmonary rhabdomyosarcoma, embryonal rhabdomyosarcoma, and malignant mesenchymoma. In paediatric patients, the lesion is a true dysembryogenic neoplasm of the thoracopulmonary mesenchyma, without malignant epithelial cells. This tumour is characterised histologically by primitive blastema and a malignant mesenchymal stroma that often shows multidirectional differentiation (rhabdomyosarcomatous, chondrosarcomatous or liposarcomatous pattern). In 25% of cases, PPB patients or their siblings have either dysplastic or neoplastic condition.

Based on the gross and microscopic features, PPB can be divided into three types. The exclusively cystic or type I PPB is the least complex and presents at an earlier age. Type II PPB has both cystic and solid lesions, while type III PPB is a purely solid tumour consisting of friable, gelatinous to mucoid, lobulated tissue often accompanied by haemorrhage and necrosis.

In the case of type I PPB, the diagnostic pitfall is failure to sample or recognise the immature mesenchymal cells, often with a rhabdomyoblastic immunophenotype, beneath the epithelial surface of the cysts. The cambium layer of primitive cells beneath the surface epithelial layer is not a continuous band of tumour cells in all cases. For this reason, extensive tissue sampling from any cystic or multicystic structure submitted as an intrathoracic or pulmonary cyst from a child is necessary in the course of the pathological examination.

Complete surgical resection remains the primary goal of treatment of children with PPB. Tagge et al convincingly admonished against the indiscriminate non-surgical management of thin-walled cystic structures that are found radiologically in the lungs of neonates and children. The PPB Reference Centre suggests that postoperative chemotherapy should be considered strongly for children with type I PPB to minimise the possibility of recurrence, as seen with the more aggressive type II or type III lesions. Recent data from the registry suggest that adjuvant chemotherapy may decrease the risk of recurrence and improve the outcome for these children. Of the 20 registry patients who had surgery alone for type I lesions, eight developed recurrent disease, and five died despite all efforts at salvage. In contrast, among 17 children who underwent surgery and received adjuvant chemotherapy as well, only one died, and the rest are disease-free at an average follow-up of 4.9 years. A combination of vincristine, actinomycin-D and cyclophosphamide has been advocated as adjuvant chemotherapy by the registry. Adjuvant therapy has also been used consistently for patients with type II and type III tumours, while radiotherapy