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Congenital Pulmonary Airway Malformation (CPAM): A case series; Radiographic importance in diagnosis

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Congenital Pulmonary Airway Malformation (CPAM) is a Crare (1-4/100,000 births) congenital anomaly, with male predominance (95% cases), presenting as a multi-cystic mass of segmental lung tissue with abnormal bronchial proliferation; considered as a part of the spectrum of Broncho-pulmonary foregut malformations. It is believed to be a result of arrested development of the fetal bronchial tree during sixth and seventh week of fetal development. Thyroid transcription factor 1 defect is also considered a possibility. Malignancy and frequent airway infections are major concerns in these patients. Surgical excision is recommended to make a definite diagnosis, to exclude hidden malignancies, and is also the treatment of choice.

Herein, I report a three cases series of different types of Congenital Pulmonary Airway Malformation (CPAM); all presenting with infection. Two of them (2 month and 2 year old males respectively) who were diagnosed as type IV presented with a clinical diagnosis of large pleural effusion. Secondary post chest tube drainage, they were diagnosed (using radiographic and CT scan correlation) with localized large multi-cystic radiolucency's in the right lung field with significant midline shift on chest radiography. The third case is of a 3 month old female; being a type II, who presented with empyema thoracis. Contrast CT aided in the diagnosis of multiple small cystic changes in the lungs. All these cases were managed in pediatric ICU and pediatric surgery services. The recent trend of antenatal detection using MRI and ultrasound as well as management, to some extent has tremendously improved the life expectancy of such cases. However, availability of timely diagnosis, necessary radio-imagining modalities and appropriate management still are quite a challenge in the developing countries.

Speaker Biography

Pradesh Ghimire has completed his MBBS from Manipal College of Medical Sciences affiliated to Kathmandu University, Nepal. He is a fourth year radiology resident in Bicol Medical Center, Naga city, Philippines.

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