SIX7: PLEUROPULMONAR BLASTOMA IN INFANT: A DIAGNOSE TO CONSIDER

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Background The congenital pulmonary airway maformations are frequently diagnosed in the prenatal period, but their postnatal approach is controversial. The risk of infection and the malignant transformation are arguments in favor of early resection. Bilateral involvement, mediastinal deviation, complex morphology and the presence of symptoms or DICER1 gene mutation should raise the suspicion of pleuropulmonar blastoma (PPB). This diagnose is less probable if prenatal diagnose of cystic pulmonary lesion, evidence of systemic vascular anomaly or hyperinsuffled pulmonary areas exist.

Materials and methods 7-weeks-old infant without previous obstetrical event, was admitted because of respiratory distress which had presented as tachypnea and subcostal drawing. The absent respiratory sounds in the 2/3 of left superior hemithorax was observed with auscultation. The Computed Tomography of the thorax revealed a multilocular cystic lesion, which occupied the whole left superior lobe, except the lingula.

Results Thoracoscopic left upper lobectomy was performed, after disinflation of the cystic lesion and the specimen was removed through one of the ports. The baby was discharged on the 5th post-operative day asymptomatic with total pulmonary expansion. The histological exam revealed type I PPB. Currently is being followed in collaboration with the Pediatric Oncology Center and has been submitted to chemotherapy.

Conclusions The described clinical case proves the necessity of considering the diagnosis of PPB when we have a symptomatic infant specially if presenting a complex cystic pulmonary lesion.

Key words pleuropulmonar blastoma, infant