38

CCAM TYPE 1 ASSOCIATED WITH MUCINOUS ADENOCARCINOMA: FIRST SURVIVOR OF NEONATAL SURGERY

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Aim of the Study: To report survival in a newborn with CCAM type 1 associated with mucinous adenocarcinoma and *KRAS* mutation in codon 12. Only five paediatric cases of this sporadic association are reported. *KRAS* accounts for 25 % of non-small cell lung cancer, and approximately 97% of *KRAS* mutations involve codons 12 or 13. There is a relationship between *KRAS* mutations and primary resistance to chemotherapy.

Method: A case report following a retrospective review of 120 patients who underwent resectional surgery for CPAM in our department (1994 - 2015).

Case presentation: A male fetus was diagnosed at 18 weeks of gestation with macrocystic CCAM. He developed significant mediastinal shift and polyhydramnios documented on fetal MR scan. His mother was HIV +ve and he was born by caesarean section at 39 weeks, requiring ventilation immediately after. A chest CT (Figure 1) showed a large lesion occupying the entire left hemithorax. Expedited surgery (left upper lobectomy) was performed on day 4 with uneventful recovery and discharge after 2 weeks.

Histology confirmed CCAM type 1 but also highlighted unsuspected multiple foci of mucinous adenocarcinoma. A somatic point mutation was detected in codon 12 of the *KRAS* gene. Extensive investigations for residual disease included further CT imaging, and negative excision biopsies of two suspicious intrathoracic areas. In view of the chemotherapy resistance associated with *KRAS* mutation, adjuvant therapy was not considered beneficial. At 7 months of life he is well and thriving with no sign of residual or recurrent disease.

Conclusion:

1. Our experience suggests an incidence for the association of CCAM and adenocarcinoma of about 1%.
2. We report the first antenatally-detected survivor for this condition.