

A Tracheal Mucoepidermoid Tumor with CRTC1-MAML2 Fusion Gene in a 12-Year-Old Boy

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Clinical Images



Figure 1a: Primary malignant tracheal neoplasms in children are rare and most mucoepidermoid tumor (MET) usually occurs in the lobar or segmental bronchi and manifest as a transluminal nodule adapting to the branching features of the airways.

Mucous pluggin, obstructive pneumonia and atelectasis are frequently associated. 19 months ago, a 12-year old boy was referred to our hospital who was diagnosed with asthma because he had dyspnea and was coughing without any improvement in prior 6 months. On CT, a well marginated lobulated tumor (Figure 1a) of 13x11x11mm in diameter was revealed in his trachea.

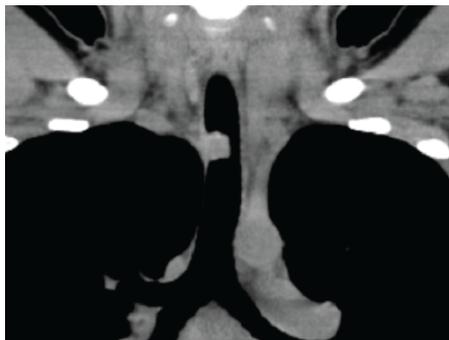


Figure 1b: On coronal view (Figure 1b), it attached with the lateral wall of trachea. A small calcification was found.

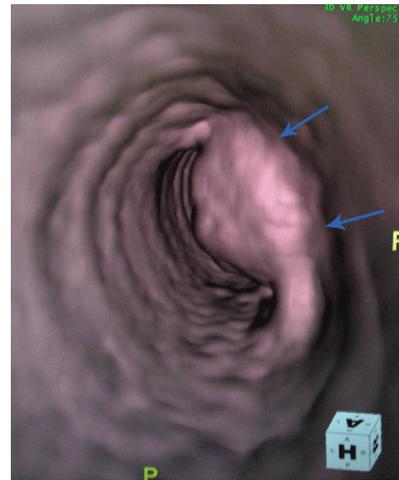


Figure 1c: The three-dimensional reconstructed virtual endoscopy showed a sessile broad based tumor (Figure 1c).

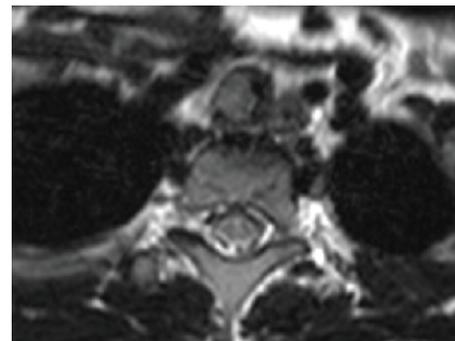


Figure 2a: MRI examination after 2 months previous CT showed an iso intensity mass on T1WI, and a slightly high intensity mass on T2WI (Figure 2a).

He underwent an emergency endoscopic subtotal resection and the histological diagnosis was made. A successful resection of the residual tumor with two tracheal rings followed by end-to-end anastomosis was done. No apparent sub mucosal invasion was observed.



Figure 2b: Biologically NAML2 split was identified on FISH, and the fusion of CRTC1 and MAML2 suggesting good prognosis was also observed in the analysis of gene(RT-PCR)

The diffusion weighted image showed almost iso signal intensity (Figure 2b) Macroscopically a 12x12x5mm tumor mass was observed.

Intermediate and goblet a typical cells. Glandular nest was also formed. The differentiation to the squamous cell was not observed, therefore, the diagnosis of low grade (grade1-2) mucoepidermoid carcinoma was made. No apparent submucosal nor vascular invasion were observed histologically. The epithelioid cells were also observed with admixed collagenous fibrous tissues. No apparent necrosis nor mitosis were recognized. The final pathologic examination confirmed the diagnosis of primary low-grade MET originating broadly from the trachea, without the involvement of the surgical margins. At present, 17 months after surgery, the child is in good general health and free of recurrence on CT. The diffusion-weighted imaging (DWI) is a form of MR imaging based upon measuring the random Brownian motion of water molecules within a voxel of tissue. The densely cellular tissues exhibit lower diffusion coefficients, and thus the signal is elevated. In our case, the mass showed almost iso signal compared with muscle which was assumed to be consistent with relatively low cellular tumor such as low grade MET.