

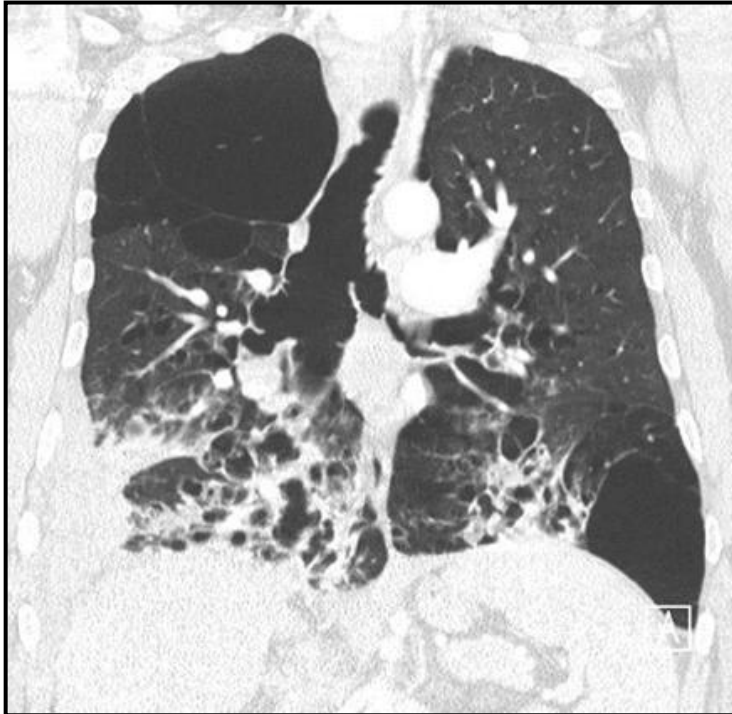
Mounier–Kuhn Syndrome: report of a case

Patientengeschichte:

Nicht-Raucher

Mit 29 Jahren erste Symptome: Dyspnoe und chronischer Husten

9.August 2010	stationäre Aufnahme im Thorax- Chirurgie LKH Graz West wegen Pleurempyem
24.August 2010	Thorax- Chirurgie AKH Wien, CT zeigt Pleuraerguss re. und Lungenemphysem
30.August 2010	LuTX-Evaluierung
29.September 2010	Sanierung des Peuraempyems
10. November 2010	LuTX



pre-operative CT-scans:

trachea 36mm (normal max. 21,8mm)

right main bronchus 17mm (normal 9-12)

left main bronchus 20 mm

LuFu Evaluation

COPD Gold IV

ausgedehnter Pleurerguss re

St.p. bakterielle Pneumonie - **Pseudomonas**

angeborene Bronchiektasie

chron. Resp. Insuffizienz

Adipositas

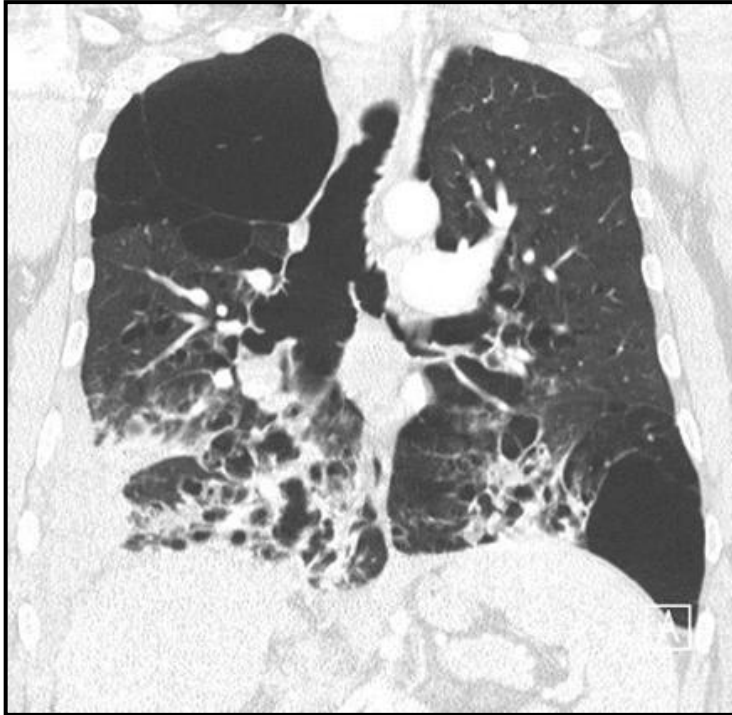
Hypertension

Stadieneinteilung nach Schwartz

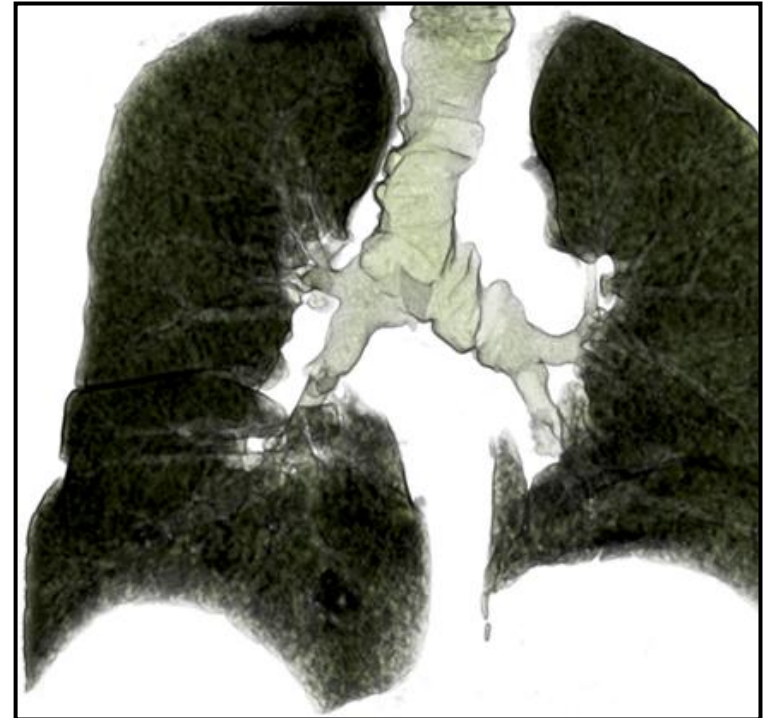
In Type 1 there is relatively subtle symmetrically diffuse enlargement of trachea and major bronchi.

Type 2 has more obvious enlargement with bizarre eccentric configurations and diverticula may also be present.

In Type 3 diverticula or sacculations extend to the distal bronchi.



Präoperatives CT



Postoperatives CT



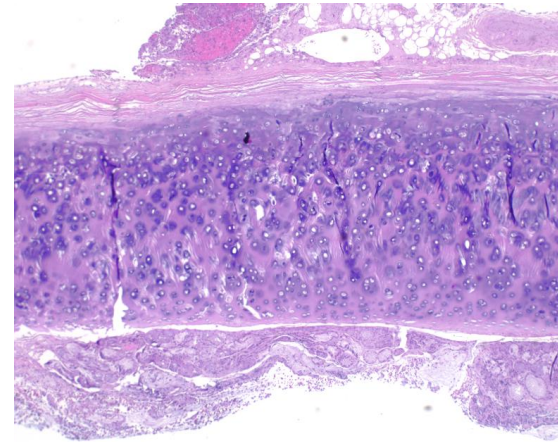
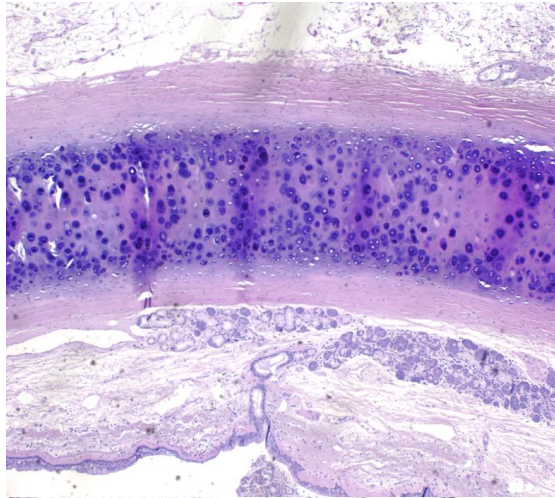
Genetische Analyse des Patienten. Blaue Balken markieren Regionen verstärkter, rote Balken Regionen verminderter DNA Kopien. Es zeigten sich keine Veränderungen in kodierenden Regionen.

Histologie Bronchusspange

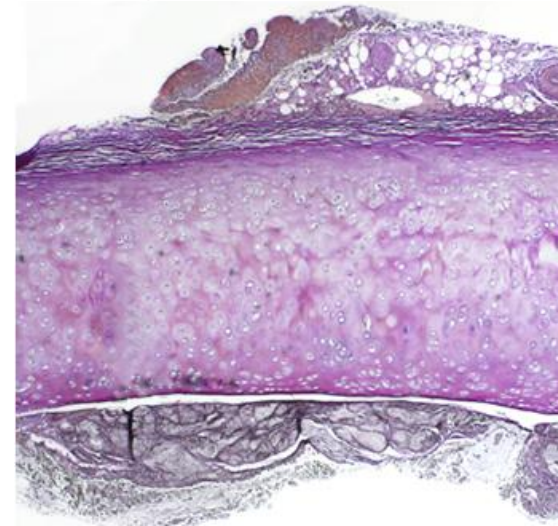
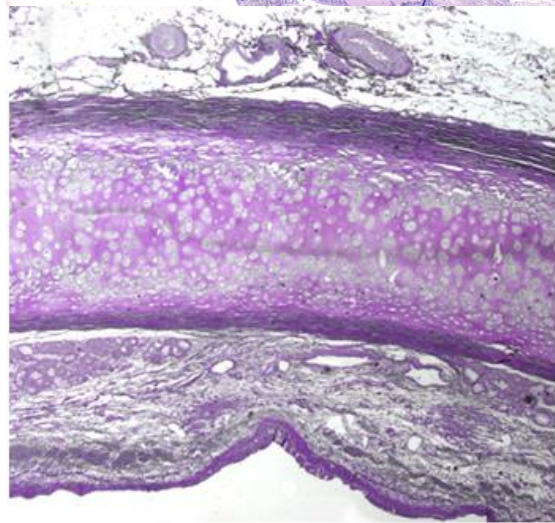
control

MKS

HE



EVG



Immunohistochemie: Matrix Metalloproteinasen

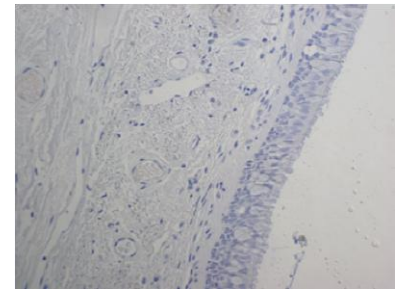
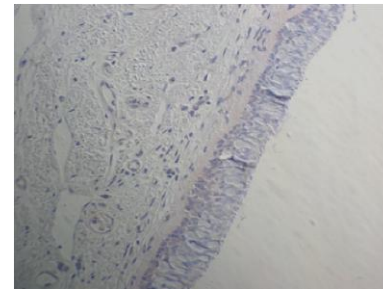
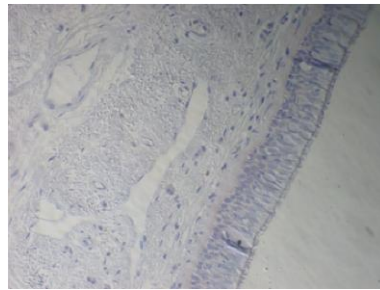
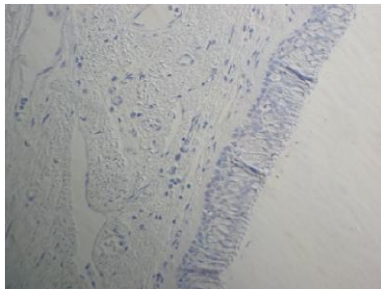
MMP1

MMP2

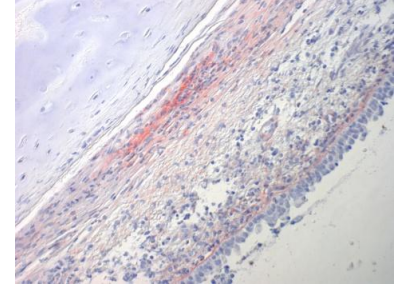
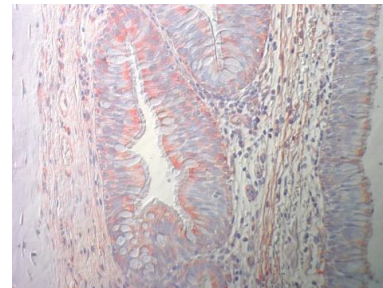
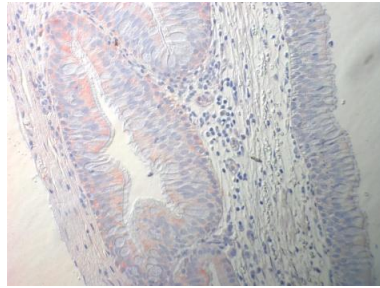
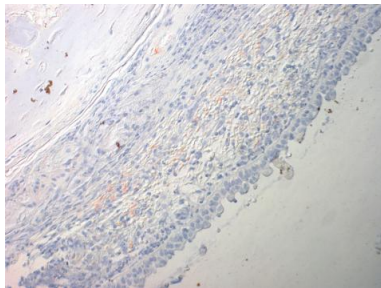
MMP3

MMP9

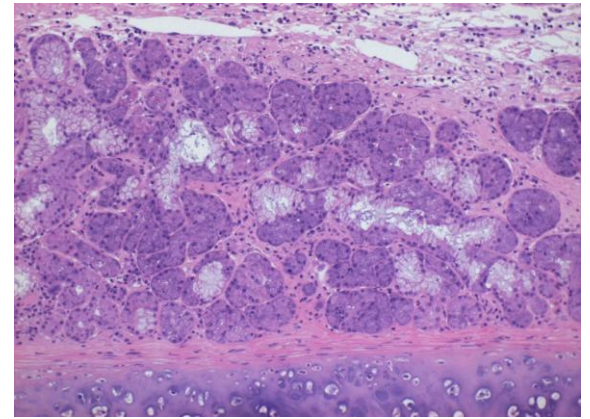
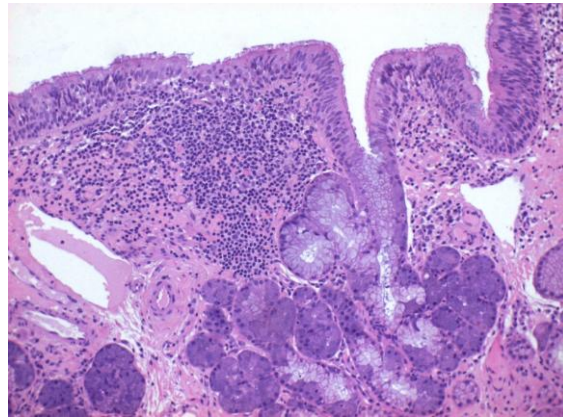
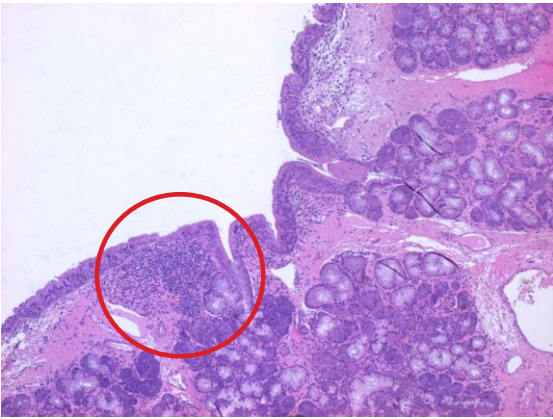
control



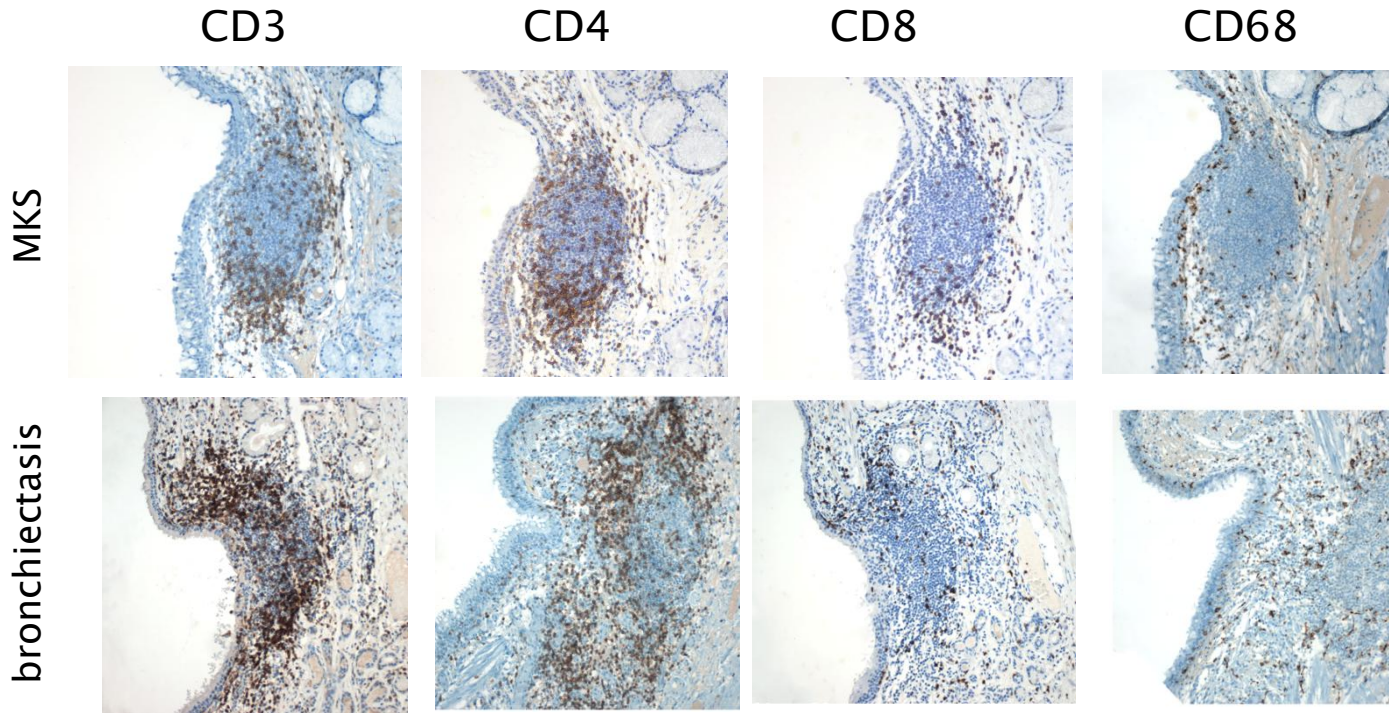
MKS



Histologie Lymphozytäre Infiltrate



Immunohistochemie des Lymphozytären Infiltrats



Pathophysiologie

- Chronische Entzündung mit Verlust der Elastischen Fasern und Zerstörung des Gewebes mit Hochregulierung der Matrix Metalloproteinasen
 - Erbkrankheit, mögliche Single-Gen Mutation
 - Autoimmunerkrankung
 - Eine Form von Bronchiektasie. 60% der Patienten mit Bronchiektasie zeigen Besiedlungen von Pseudomonas oder Haemophilus in den Atemwegen.
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