Prospective MRI study of cystic fibrosis (CF)-associated changes in the pancreas with regard to their precancerous potency

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Aims and objectives

Besides pulmonary manifestation, up to 90% of patients with cystic fibrosis (CF) suffer from pancreatic disorders, including exocrine (80%) or endocrine insufficiency (30-50%). The mutation of the cystic fibrosis transmembrane conductance regulator gene (CFTR) leads to abnormally thickened, viscous secretions which provoke the obstruction of pancreatic ducts. They also release lipases and proteases leading to autolysis of the pancreas, followed by fibrosis and atrophy. Thereafter the pancreatic tissue is replaced by fat.

Previous studies described partly or totally fatty replacement in CT- or MRI- studies, as well as cystic lesions and ductal calcifications. The calcifications are often difficult to detect in MR imaging [1].

It has been shown that the loss of CFTR expression through mutation can also further the development of a pancreatic ductal adenocarcinoma (PDAC) [2].

An important precursor lesion to PDAC is the intraductal papillary mucinous tumor (IPMN). Its pathology originates from the mucinous epithelium of the pancreatic duct and is characterized by papillary masses and a ductal dilatation. A connection between the cystic lesions and the pancreatic ducts is important for differential diagnosis. In some cases solid mural nodules can be detected. IPMN have a premalignant potential. In particular IPMN concerning the main duct, as well as large IPMN (> 2 cm) originating in the side branches require surgery. Another precursor lesion is the intraepithelial Neoplasm (PanIN), which can only be differentiated histologically. Occurrence of PanIN3 seems to define a high risk for malignant transformation [3].

As MR imaging is highly sensitive in depicting IPMN and MR cholangiopancreatography (MRCP) is helpful in showing a possible connection to the pancreatic duct, MRI in combination with MRCP is the preferred imaging method to analyze possible pancreatic alterations in patients with CF.

Methods and materials

So far, 17 patients (11 male, 6 female, average age 32 years) with confirmed CF were examined as part of this ongoing prospective study by MRI and MRCP for changes of the pancreas (Magnetom Skyra 3 Tesla, Siemens Healthcare AG, Germany).
Prior or during the examination, patients received the following:

- 40 mg N-Butylscopolaminiumbromid intravenously to reduce intestinal mobility (<50 kg: 20 mg)
- 300 ml Lumirem® (+300 ml H2O) orally for the MRCP
- an intravenous injection of Gadovist® (1 ml/ 10 kg bodyweight)

The sequence protocol encompassed:

- T2w HASTE coronal and axial
- T2w HASTE fat-saturated coronal
- T2w TIRM coronal
- T2w SPACE 3D coronal with MIP
- diffusion-weighed EPI-sequences with ADC
- dynamic T1w sequences
- T1w FLASH fat-saturated coronal and axial with contrast medium

Results

The preliminary evaluation of the executed MRI screenings revealed a partial or complete fatty replacement of individual sections of the pancreas (caput, corpus or cauda) in 13 patients (76%). In four cases (24%) no pancreatic tissue could be separated after a complete parenchymal atrophy and fatty replacement (Fig.1). Five patients (29%) showed cyst-like changes, resembling retention cysts, in line with the underlying disease (Fig.2). In one of these cases a total cystic transformation of the pancreas was found (Fig.3). Further four patients (24%) presented one or more cystic formations with contact to the pancreatic duct in the MRCP. Morphologically, these are compatible with intraductal papillary mucinous neoplasms (IPMN), of which three seem to concern the minor duct only (Fig.4), as well as a hybrid form affecting the minor and major duct (Fig.5). No signs of malignancy like diffusion impairment or papillary projection could be found in any of the patients.

These results suggest that in the group of patients examined so far, the prevalence of IPMN lies considerably above the overall population.

However, it has to be mentioned that a 29-year old male patient with the assumed hybrid IPMN shown in MR imaging, was further examined by ERCP, where again an IPMN was confirmed. His blood tests showed an increase in the Carboanhydride Antigen CA 19.9 to 988 IU/ml (laboratory reference<32.0 U/ml), so he underwent surgery. After pancreatectomy, histology showed no IPMN and no malignancy, but instead a PanIN2-
lesion. Therefore, a thorough and further study of IPMN-like lesions in these patients is crucial.

Images for this section:

**Fig. 1:** T2w HASTE demonstrating complete fatty replacement of the pancreas (cauda) in a 37-year-old woman with cystic fibrosis.
Fig. 2: Coronal T2w HASTE showing large cyst-like lesions (max. 4 cm), presumably retention cysts, in a 50-year-old man with cystic fibrosis.
**Fig. 3:** Axial T2 HASTE demonstrating complete cystic pancreatic transformation in a 28-year-old man with cystic fibrosis.
**Fig. 4:** Coronal T2w Rare (MRCP) demonstrating cystic formations with contact to the pancreatic duct, resembling intraductal papillary mucinous neoplasms (IPMN), in this case concerning the minor duct.

**Fig. 5:** Coronal T2w Rare (MRCP) depicting the assumed hybrid IPMN in a 29-year-old male patient with cystic fibrosis.
Conclusion

In addition to the MRI appearances of the pancreas described in literature so far, this ongoing study suggests that an increased risk for IPMN in patients with cystic fibrosis cannot be precluded through critical evaluation. It is under discussion whether these cyst-like lesions with connection to the pancreatic duct are IPMN or retention cysts. As most cystic fibrosis patients present with unusual cystic abnormalities, it is difficult to clearly identify the type of lesion and it is therefore suggested to screen these patients every 12 months. If a progress of growth (> 2 cm), a contrast enhancement or a diffusion restriction presents, these patients are recommended to undergo surgery.

This study is limited by the number of patients analysed so far. As the study proceeds, further patients will be admitted, as well as laboratory figures like the Carboanhydrate Antigen (CA 19-9) correlated with MRI data.

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References

