A pictorial review of complications following lung transplantation for cystic fibrosis.

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Learning objectives

Within this pictorial review we illustrate the common pathologies that affect the transplanted lung following lung transplantation for cystic fibrosis as well as including some rarer complications. Categories include; Infection, malignancy, anastomotic complications as well as both acute and chronic rejection.

It must be remembered that although the individual has undergone lung transplantation this is not a cure and cystic fibrosis has continuing effects on the rest of the body.
Background

Lung transplantation has been available for several decades. The first successful lung transplant for cystic fibrosis took place in 1985. Lung transplantation is considered the 'ultimate therapy' with between 150 and 200 cystic fibrosis patients receiving transplants annually in the UK. A significant increase in five year survival has been documented following lung transplantation, however transplantation and immunosuppressant therapy is not without risks.

This pictorial review illustrates both common and rarer complications experienced following lung transplant for cystic fibrosis at our large regional cystic fibrosis centre in Leeds, UK.
Infection:

Infection remains one of the most important causes of morbidity and mortality in lung transplant recipients. The increased susceptibility is multifactorial with commonest cause being bacterial infections. Cystic fibrosis transplanted patients have a higher risk of Pseudomonas Aeruginosa infection. Viral infections such as Cytomegalovirus and Respiratory Syncytial Virus are common as are fungal infections including Aspergillus and Candida.

Case 1:

This patient presented with cough. HRCT revealed a thin walled cavity within the right upper lobe with a small soft tissue component seen within (Fig 1&2). The patient was treated with anti-fungal medication and initially the cavity decreased in size. A year later she became increasingly unwell with fever and cough. Imaging demonstrated an increase in size of both the right upper lobe cavity and the soft tissue component within (Fig 3). Two days later the patient became acutely unwell with massive haemoptysis. Fig 4 & 5 demonstrate high density material within the cavity and central airways in keeping with acute haemorrhage.

Malignancy:

The overall risk of cancer within the non transplanted cystic fibrosis patient is similar to that of the general population. Post transplant cystic fibrosis patients have an increased risk of malignancy, with the risk of small bowel, colon and biliary tract malignancy being particularly elevated. Post transplant lympho proliferative disease and lymphoma are also relatively frequently seen within this population.

Case 2:

This 51 year old female patient presented with abdominal pain, weight loss and anaemia. Staging CT of the abdomen and pelvis identified an 8.7cm left upper quadrant mass, highly suspicious for lymphoma (Fig 6&7). B cell lymphoma was confirmed on endoscopic biopsy.

Case 3:
This 35 year old female presented with abdominal pain and weight loss. CT (Fig 8) showed a large pelvic mass centered on the cervix with evidence of extensive pelvic and retroperitoneal nodal disease. PET- CT (Fig 9) confirmed the CT findings and the patient underwent surgery with adjuvant chemo-radiotherapy. Despite aggressive treatment, the subsequent staging CT demonstrated disease progression with multiple sub centimeter pulmonary metastases seen within the transplanted lungs (Fig 10).

Case 4:

This 36 year old patient presented with abdominal pain and constipation. CT identified multiple pathologically enlarged retroperitoneal and mesenteric lymph nodes (Fig 11&12). Subsequent CT guided biopsy of a retroperitoneal node confirmed a diagnosis of diffuse B cell lymphoma.

Anastomotic Complications:

Lung transplantation involves three anastomotic sites: The airways, pulmonary arteries and pulmonary veins / left atrium. Multiple case reports of leaks, fistula, stenoses at anastomotic sites and thrombus formation within the pulmonary veins are described in the literature.

Case 5:

This 40 year old patient presented within a year of lung transplant with increased shortness of breath. She initially underwent bronchoscopy and washings which were normal. Subsequent CXR (Fig 13) showed a right upper lobe cavity and CT (Fig 14 & 15) confirmed the presence of an abscess within the right upper lobe. Following a prolonged course of antibiotics the cavity decreased in size (Fig 16). Several months later the patient described coughing when lying flat. CT (Fig 17) demonstrated increase in size of the right upper lobe abscess and appearances suspicious for fistulous connection to the mediastinum. A fistula between the oesophagus and right upper lobe was subsequently confirmed with contrast swallow(Fig 18).

Allograft rejection:

The peak incidence of acute rejection is said to be around 100 days post transplant. Typical clinical manifestations include fever, dyspnoea, cough and malaise with low oxygen saturations. CXR may show alveolar, interstitial or nodular infiltrates as well as pleural effusions. Diagnosis is confirmed with trans bronchial biopsy.
Chronic rejection limits the long term survival of lung transplant recipients. Histologically it resembles "bronchiolitis obliterans", a fibroproliferative process affecting small airways. Unlike acute rejection the transbronchial yield for histological confirmation is low. For this reason chronic rejection is defined based on lung function tests and demonstration of air flow limitation. HRCT shows bronchial wall thickening, peribronchial opacities, tree in bud pattern and air trapping (expiratory sequence required).

Case 6:

This patient presented 10 weeks following lung transplant with fever, lethargy, cough, dyspnoea and low oxygen saturations. CXR on admission showed bilateral symmetrical perihilar / lower zone opacification in keeping with acute rejection (Fig 19).

Case 7:

Several years following lung transplantation this patient had a steadily declining in lung function.

HRCT demonstrated bronchial wall thickening, air trapping and sub pleural fibrotic changes. (Fig 20 & 21)

Extra pulmonary manifestations:

Multiple extra pulmonary manifestations of cystic fibrosis exist and persist following lung transplantation. More commonly encountered abnormalities include a meconium ileus equivalent small bowel obstruction, biliary cirrhosis, pancreatic insufficiency, bone demineralization, sinusitis and infertility.

Case 8:

This 36 year old male patient presented following lung transplantation with abdominal pain & distention. Clinically he was felt to be obstructed and abdominal radiograph confirmed the presence of dilated loops of small bowel. CT demonstrated dilated loops of small bowel with a small bowel faeces sign indicating stagnation of bowel contents (Fig 22 & 23). The same examination demonstrated fatty replacement of the pancreas, correlating with the known pancreatic insufficiency. (Fig 24)

Other factors that have been reported to affect the patient following lung transplantation for cystic fibrosis include:
Venous thromboembolism, pulmonary embolism, phrenic nerve / diaphragmatic dysfunction, drug induced pulmonary toxicity and graft versus host disease.
Images for this section:

**Fig. 1:** Thin walled cavity within the right upper lobe.

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Fig. 2: Small soft tissue component within the right upper lobe cavity consistent with a mycetoma.

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Fig. 3: Increasing right upper lobe cavity with an increase in size of the soft tissue component.

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Fig. 4: Increase high density material seen within the right upper lobe cavity. Radiological appearances are consistent with acute haemorrhage.

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Fig. 5: High density material, in keeping with blood is seen within the airways. Correlating with the clinical presentation of massive haemoptysis.

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**Fig. 6:** 8.7cm mass within the left upper quadrant extending from the duodenal-jejunal flexure to the proximal jejunum.

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**Fig. 7:** Coronal imaging best demonstrates the circumferential jejunal thickening.
Fig. 8: CT showed enlarged retroperitoneal and pelvic side wall lymphadenopathy as well as a large cervical mass.
**Fig. 9:** PET confirmed a locally advanced cervical malignancy with pelvic side wall and retroperitoneal lymph nodes.

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Fig. 10: Despite surgery and aggressive chemo and radiotherapy the patient developed multiple pulmonary metastases within the transplanted lungs.

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Fig. 11: Multiple pathological mesenteric lymph nodes.

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Fig. 12: Multiple enlarged retroperitoneal and mesenteric lymph nodes confirmed as diffuse B cell lymphoma following CT guided biopsy.

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**Fig. 13:** CXR showing right upper lobe cavity.

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**Fig. 14:** Ct showing right upper lobe cavity.
Fig. 15: CT Right upper lobe cavity with fluid and gas within.

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Fig. 16: CT showing initial resolution of the right upper lobe cavity with antibiotic treatment.

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**Fig. 17:** CT showing recurrence and increase size of the right upper lobe cavity with an air fluid level within.

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Fig. 18: Contrast swallow confirmed fistulous tract between the proximal oesophagus and the right upper lobe cavity.

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Fig. 19: CXR shows Increase opacification within the lower zones bilaterally.
**Fig. 20:** Bronchiectasis and evidence of fibrosis in keeping with bronchiolitis obliterates/transplant rejection.

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Fig. 21: Evidence of bronchiectasis and fibrosis in keeping with suspected transplant rejection.

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Fig. 22: Coronal images showing multiple loops of dilated small bowel with small bowel faeces sign. No transition point was identified and features were compatible with a meconium ileum equivalent.

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Fig. 23: Small bowel faeces sign within the terminal ileum. Dilated loops of small bowel on this coronal CT.

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Fig. 24: Fatty replacement of the pancreas on CT.

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Conclusion

Post lung transplant complications in the cystic fibrosis population can be divided into several categories. Within our pictorial review we have illustrated common as well as rarer complications seen at our regional cystic fibrosis unit in Leeds, UK.
References

www.cysticfibrosis.org.uk

Personal Information

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