Bronchiectasis& Cystic fibrosis

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Bronchiectasis means abnormal dilatation of the bronchi.Chronic supportive airway infection with sputum production, progressive scarring and lung damage occur, whatever the cause
Bronchial walls become inflamed, thickened and irreversibly damaged. The mucociliary tran sport mechanism is impaired and frequent bacterial infections ensue. Clinically, the disease is characterized by cough production of large amounts of sputum and dilated and thickened bronchi, detected on CT scanning of the thorax.

Causes of bronchiectasis:

Congenital:
• Cystic fibrosis
• Ciliary dysfunction syndromes
Primary ciliary dyskinesia (immotile cilia syndrome)
Kartagener's syndrome
(sinusitis and transposition of the viscera)
• Primary hypogammaglobulinaemia

Acquired: adults:----------
• Suppurative pneumonia
• Pulmonary TB
• Allergic bronchopulmonary aspergillosis complicating asthma
• Bronchial tumours

Acquired: children----------
-Pneumonia (complicating whooping cough or measle
-Primary TB
-Inhaled foreign body

Pathology:

Bronchiectasis may result from a congenital defect affecting airway ion transport or ciliary function, such as cystic fibrosis , or may be acquired secondary to damage to the airways by a destructive infection, inhaled toxin or foreign body.
The result is chronic inflammation and infection in airways. shows the common causes, of which tuberculosis is the most common worldwide.
Localized bronchiectasis may occur due to the accumulation of pus beyond an obstructing bronchial lesion, such as enlarged tuberculous hilar lymph nodes, a bronchial tumor or an inhaled foreign body (e.g. an aspirated peanut).
The bronchiectasis cavities may be lined by granulation tissue, squamous epithelium or normal ciliated epithelium. There may also be inflammatory changes in the deeper layers of the bronchial wall. Chronic inflammatory and fibrotic changes are usually found in the surrounding lung tissue, resulting in progressive destruction of the normal lung architecture in advanced cases.

Clinical features of bronchiectasis:

chronic cough with production of copious amounts of purulent sputum, hemoptysis, and pleuritic chest pain. Dyspnea and wheezing occur in 75% of patients. Weight loss, anemia, and other systemic manifestations are common. Physical findings are nonspecific, the patient suffers from recurrent febrile episodes with malaise, and episodes of pneumonia.
When the condition is severe there is continuous production of foul-smelling, thick, brownish-yellow -colored sputum.
Hemoptysis can occur either as blood-stained sputum or as a massive haemorrhage. Breathlessness may result from airflow limitation.
Physical signs in the chest may be unilateral or bilateral.
If the bronchiectatic airways do not contain secretions and there is no associated lobar collapse, there are no abnormal physical signs.
When there are large amounts of sputum in the bronchiectatic spaces, numerous coarse crackles may be heard over the affected areas. Collapse with retained secretions blocking a proximal bronchus may lead to locally diminished breath sounds,while advanced disease may cause scarring

and overlying bronchial breathing. Acute hemoptysis is an important complication of bronchiectasis.

Clubbing of fingers:

Clubbing is infrequent in mild cases but is common in severe disease.

Obstructive pulmonary dysfunction with hypoxemia is seen in moderate or severe disease.

Investigations:

Sputum examination :

culture may reveal Pseudomonas aeruginosa, fungi such as Aspergillus and various mycobacteria, are necessary to ensure appropriate treatment of resistant organisms.

Chest X-ray:

Bronchiectasis, unless very gross, is not usually apparent .In advanced disease, thickened airway walls, cystic bronchiectasis spaces, and associated areas of pneumonic consolidation or collapse may be visible.

Chest CT scan : is much more sensitive, and shows thickened, dilated airways

Sinus X-rays: Thirty per cent have concomitant purulent rhinosinusitis.

Serum immunoglobulins: Ten per cent of adults have immune deficiency.

Assessment of ciliary function:

Screening test can be performed in patients suspected of having a ciliary dysfunction syndrome by measuring the time taken for a small pellet of saccharin placed in the anterior chamber of the nose to reach the pharynx, at which point the patient can taste it. This time should not exceed 20 minutes but is greatly prolonged in patients with ciliary dysfunction.

Ciliary beat frequency:

may also be assessed from biopsies taken from the nose. Structural abnormalities of cilia can be detected.

Management:

In patients with airflow obstruction, inhaled bronchodilators and ,corticosteroids should be used to enhance airway patency.

Physiotherapy:

Patients should be shown how to perform regular daily physiotherapy to assist the drainage of excess bronchial secretions. Efficiently executed, this is of great value both in reducing the amount of cough and sputum, and in preventing recurrent episodes of bronchopulmonary infection.

Patients should lie in a position in which the lobe to be drained is uppermost. Deep breathing followed by forced expiratory manoeuvres (the ‘active cycle of breathing’ technique) helps to move secretions in the dilated bronchi towards the trachea, from which they can be cleared by vigorous coughing.

Devices that increase airway pressure either by a constant amount(positive expiratory pressure mask) or in an oscillatory manner. The optimum duration and frequency of physiotherapy depend on the amount of sputum, but 5–10 minutes twice daily is a minimum for most patients.

Antibiotic therapy:

For most patients with bronchiectasis, the appropriate antibiotics are the same as those used in COPD.

but larger doses and longer courses are required, and resolution of symptoms is often incomplete. When secondary infection occurs with staphylococci and Gram-negative bacilli, in particular Pseudomonas species, antibiotic therapy becomes more challenging and should be guided by the

microbiological sensitivities. For Pseudomonas, oral ciprofloxacin (500–750 mg twice daily) or ceftazidime by intravenous injection or inhalation (1–2 g 3 times daily) may be required.
Hemoptysis in bronchiectasis often responds to treatment of the underlying
infection, although, in severe cases, percutaneous embolisation of the bronchial circulation by an interventional radiologist may be necessary.

Surgery:

Unfortunately, it is rare for bronchiectasis to be sufficiently localized for a resection to be performed. Lung or heart-lung transplantation is sometimes required .

Complications:

The incidence of complications has fallen with antibiotic therapy. Pneumonia, pneumothorax , empyema and metastatic cerebral abscess can occur. Severe, life threatening hemoptysis can also occur, particularly in patients with cystic fibrosis.

Prognosis:

The disease is progressive when associated with ciliary dysfunction and cystic fibrosis, and eventually causes respiratory failure. In other patients, the prognosis can be relatively good if physiotherapy is performed regularly and antibiotics are used aggressively.

Prevention:

As bronchiectasis commonly starts in childhood following measles, whooping cough or a primary tuberculous infection, adequate prophylaxis for and treatment of these conditions are essential.
The early recognition and treatment of bronchial obstruction are also important.

Cystic fibrosis:

Most common fatal genetic disease, with autosomal recessive inheritance. mutations affecting a gene on the long arm of chromosome 7 which codes for a chloride channel known as cystic fibrosis transmembrane conductance regulator (CFTR),
In cystic fibrosis (CF) there is an alteration in the viscosity and thicken of mucus produced at epithelial surfaces .
The classical form of the syndrome includes bronchopulmonary infection, bronchiectasis and with high sweat sodium and chloride concentration.
The gene defect also causes disorders in the gut epithelium, pancreas, liver and reproductive tract.
the clinical picture (bowel obstruction, failure to thrive, steatorrhea and/or chest symptoms in a young child) supported by sweat electrolyte testing and genotyping. Patients with unusual phenotypes were commonly missed, however, and late diagnosis led to poorer outcomes.
Neonatal screening for CF using immunoreactive trypsin and genetic testing of newborn blood samples is now routine in the UK.

Clinical pictures :

Although the lungs of babies born with CF are structurally normal at birth, frequent respiratory infections soon develop and are often the presenting feature.
Sinusitis is almost present and nasal polyps are common. Breathlessness and haemoptysis occur in the later stages as airflow limitation and bronchiectasis.
At this stage, the lungs are most commonly infected with **Staphylococcus aureus**; however, many patients become colonized with **Pseudomonas aeruginosa** by the time they reach adulthood.
Recurrent exacerbations of bronchiectasis.
initially in the upper lobes but subsequently throughout **both lungs,** cause progressive lung damage and Spontaneous pneumothorax may occur resulting ultimately in death from **respiratory failure**.

Gastrointestinal effects:

About 85% of patients have symptomatic steatorrhea due to pancreatic dysfunction, meconium ileus, intestinal obstruction ,gall stones and liver cirrhosis .

Other presentation of C.F:

Males are almost always infertile due to failure of development of the vas deferens and epididymis

Females are able to conceive, but often develop secondary amenorrhoea as the disease progresses.

Arthropathy and diabetes mellitus also occur.

Diagnosis:

The diagnosis of CF in older children and adults is based on the clinical history and:

-A family history of the disease

-A high sweat sodium concentration over 60 mmol/L regular sweat analysis is essential, but the test is still difficult to interpret in adults).

■ Blood DNA analysis of gene defect

■ Radiology showing features seen in bronchiectasis

■ Absent vas deferens and epididymis

Treatment:

All patients with CF who produce sputum should perform chest **physiotherapy** daily, and more frequently during exacerbations.

Oxygen should be given as necessary

Antibiotics: treatment for respiratory infections is as described under bronchiectasis

Inhaled antibiotics and corticosteroids can also reduce inflammation and improve lung function**.**

Novel therapies for cystic fibrosis:

Ivacaftor (a CFTR 'potentiator'), is now an established oral treatment for few percent of CF patients.

Somatic gene therapy for CF is under development.

Other treatment :

Treated with oral pancreatic enzymes .and insulin therapy for DM

Good nutrition: vitamin supplements. Malabsorption occurs in 85% of patients.

Lung transplantation can produce dramatic improvements but is limited by donor organ availability

The main diseases treated by Lung transplantation:

-Pulmonary fibrosis

-Primary pulmonary hypertension

-Cystic fibrosis

-Bronchiectasis

-Emphysema - particularly alpha one antitrypsin deficiency .

-Eisenmenger's syndrome

Screening:

Genetic screening is available for the most common mutations and this identifies 85-95% of carriers. Screening for the carrier state should be offered to persons or couples with a family history of CF, together with counselling.