IDIOPATHIC
INTERSTITIAL PNEUMONIAS

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MHA
MEDIHEALTH ACADEMY

ISBN: 978-625-99889-5-5

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Publisher
MediHealth Academy

Project Coordinator
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(MediHealth Academy)

Graphic-Design
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Furkan Selçuk AYDOĞDU
(MediHealth Academy)

Edition-Volume
Ankara Özgür Matbaacılık Basım Yayın Dağ. San. Tic. A.Ş.
1250 Cadde No: 25 Ostim OSB Yenimahalle / Ankara
Certificate Number: 46821

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Preface

Interstitial lung diseases; it is a group of diseases that affect the lung diffusely and show similar clinical, radiological and pathological features. Especially in recent years, as a result of the advances in diagnosis and treatment, its awareness has increased and progress has been made in treatment. Our aim in preparing this book is to guiding our colleagues working in the field of chest diseases in diagnosis and treatment, with current studies.

I would like to thank all my co-editor and writer colleagues, especially Associate Professor Berna AKINCI ÖZYÜREK, who always guided me with her experience and knowledge during the preparation of the book. We hope that this work will be useful to our colleagues

Sincerely,

Emire Pınar SEYFETTİN ÇELİK
MD, Editor
2023, Ankara

Idiopathic Pulmonary Fibrosis

Chapter 1

Berna AKINCI ÖZYÜREK, Hasret Gizem KURT

Idiopathic pulmonary fibrosis (IPF); It is a chronic interstitial lung disease with high mortality rates, characterized by advanced fibrosis, causing rapid and progressive decline in lung functions.

Epidemiology

The prevalence and incidence rates of IPF are increasing worldwide. Geographic region differences, gender and age distribution ratios of the population affect prevalence and incidence rates. The age at diagnosis of IPF patients is between 60 and 80, and it is more common in men. A systematic review revealed that IPF had a prevalence of 0.9-13/100,000 and an incidence of 3.3 to 45.1/100,000. Analysis of studies from Europe and North America revealed the incidence was 1.3-11.7 per 100,000. In the United States, the rate ranges from 7 to 16 per 100,000. In our country, the incidence of IPF between 2007 and 2009 is 5 per 100,000.

Risk Factors and Pathogenesis

It is known that IPF develops because of the interaction of genetic and environmental risk factors. Repetitive micro-damages primarily affect the alveolar epithelium. An abnormal healing process begins after epithelial damage. An imbalance between fibrotic and anti-fibrotic mediators causes myofibroblast, fibroblast, and collagen accumulation. Afterwards, tissue damage, fibrosis and honeycomb cysts occur.

There are some risk factors that cause the development of the IPF. These factors are male gender, age, smoking history over 20 pack/year, environmental and occupational factors, chronic micro-aspirations because of gastroesophageal reflux and viral factors.

Current and previous smoking history increase the risk of IPF. The clinical course of the disease is worse in patients with IPF who smoke than in patients who do not smoke.

The study in the USA identified some occupations associated with the IPF. These professions agriculture, animal husbandry, hairdressing, bird breeding, stone cutting and polishing are occupations that are exposed to metal dust and vegetable dust. Also, exposure to metals such as cadmium, aluminum, copper, arsenic, cobalt, molybdenum, tungsten, uranium, and vanadium have been closely associated with IPF. Disruption of the lung microbiome because of causes such as viral infections; There are studies showing that it has a poor prognostic factor effect.

Familial IPF is defined as IPF in at least two of the primary biological family members. Familial IPF; It is seen at a rate of 0.5-2.2%. It is autosomal dominant and is observed at earlier ages.

Acute Fibrinosis And Organized Pneumonia

Chapter 5

Kerem ENSARİOĞLU

Acute fibrinous and organizing pneumonia (AFOP), first described histologically by Beasley et al. in 2002, has a specific histological pattern of its own and in 2022 American Thoracic Society (ATS), European Respiratory Society (ERS), Japanese Respiratory Society (JPS) and Associacion Latinoamericana de Torax (ALAT) consensus is an idiopathic interstitial lung disease excluded from idiopathic pulmonary fibrosis (IPF).

In 2002, Beasley et al., in their evaluation at their clinic, mentioned patients who they thought did not fit the classification in the ATS and ERS joint guideline, which was prepared a year ago and published in 2002. In this guideline, where the nomenclature of the idiopathic forms of diffuse alveolar injury (DAD) and bronchiolitis obliterans (BOOP) has been changed, it is recommended to use the definition of organizing pneumonia histologically, and eosinophilic pneumonias (EP) are divided into acute and chronic. In this scan performed by Beasley on patients who were seen with an acute or subacute clinic and did not meet the definitions of DAD, BOOP and EP in the guideline, it was observed that 17 patients had clinical features that had a common histological character and were compatible with each other. In this study, in which six patients did not have any triggering etiology, the definition of AFOP was made for the first time and pathology findings independent of DAD, BOOP, and EP were specified. Although it was reported to have similar mortality with DAD, it was emphasized that it characteristically followed two different clinical patterns, the acute presentation was more mortal and was defined as fulminant, respiratory failure was observed less frequently in the milder subacute variant, and mortality was not observed.

Following Beasley's classification, in the 2013 ATS/ERS interstitial lung diseases update, AFOP was classified as a rare histological pattern. In the same report, it was emphasized that it can be seen in the presence of DAD and OP, although it can be idiopathic, there may be rheumatological lung disease, drug reaction or hypersensitivity pneumonia in the background, and because it is similar to eosinophilic pneumonia, it can be ruled out in the absence of peripheral eosinophils. Following this report, it was seen that more than a hundred patients were diagnosed in case reports and series until 2016 for AFOP, which is generally considered as being a subtype of COP, and its importance in differential diagnosis in the evaluation of COP. AFOP, which was reclassified in 2022 according to the current ATS/ERS/JPS and ALAT guidelines, is now considered a rare non-IPF interstitial lung disease that requires pathological sampling because it is in its own category and is diagnosed as exclusion.

Etiology

AFOP occurs in a wide age range and in a non-sexist spectrum of patients. Although idiopathic cases have been reported, case series in which the underlying etiology is known and AFOP has been attributed to it has been published. Basically,

Acute Interstitial Pneumonia

Chapter 7

Büşra BALKAY BABAEV

Acute interstitial pneumonia (AIP) is a rare and fulminant disease with diffuse lung damage, first described by Hamman and Rich in 1935. AIP is classified as idiopathic Interstitial Pneumonia (IIP) and is the most acutely onset and rapidly progressive among IIPs. The course of AIP is similar to that of acute respiratory distress syndrome (ARDS) and represents a subgroup of idiopathic ARDS cases.

Epidemiology

AIP usually affects individuals who are previously healthy and have no prior history of lung disease. It occurs with equal frequency in men and women. It has nothing to do with smoking. Most of the patients are over 40 years old, with an average age range of 50-55.

Histopathology

AIP has the histopathological appearance of diffuse alveolar damage (DAD). DAD is a reaction pattern that occurs in response to a number of known causes of lung injury, but is idiopathic in the case of AIP (Table 1). Acute exudative, organized proliferative, and healed (or fibrotic) stages are seen in the development of DAD, respectively.

Table 1. Causes of diffuse alveolar damage

Acute hypersensitivity pneumonia	*Legionella*
Idiopathic acute interstitial pneumonia (Hamman Rich syndrome)	Viral infections, including influenza and severe acute respiratory syndrome (include SARS)
Medications	*Mycoplasma*
Chemotherapeutic agents	Ingestants
Ethchlorvinol	Paraquat
Aspirin	Kerosene
Radiation therapy	Rapeseed oil toxic oil syndrome
Oxygen toxicity	Toxic inhalants
Heroin	Chlorine gas
Cocaine	Nitrogen dioxide
Connective tissue disease	Phosgene
Polymyositis	Smoking
Lupus pneumonia	