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Review Article

Idiopathic non-specific interstitial pneumonia: a brief review

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ABSTRACT

Non-specific interstitial pneumonia (NSIP) is one of the better variants of interstitial lung disease in terms of outcomes. Timely diagnosis with early treatment has excellent 5-year survival figures. Typical clinical symptoms and radiologic findings makes diagnosis easy. Ruling out various associated connective tissue disease (CTD) is of paramount importance as they are amenable to immunosuppressant therapies. Majority of cases still remain idiopathic which forms the major chunk of NSIP pool. This article highlights the natural history, epidemiology and key differentiating features of NSIP and current treatment options.

Keywords: Idiopathic NSIP, Connective tissue disease, Ground glassing

INTRODUCTION

Interstitial lung diseases (ILD) are a diverse group of lung disorders with non-specific interstitial pneumonia (NSIP) a distinct entity with mostly unknown cause. They are often associated with connective tissue diseases (CTD) and less frequently with drug toxicity, infection and smoking.

NSIP was initially not considered a separate disease entity. Traditional classification of interstitial pneumonia was given first by Liebow in 1969.[1] He proposed five classical pathological histological types of diffuse idiopathic interstitial pneumonia, namely, universal interstitial pneumonia (UIP), desquamation interstitial pneumonia (DIP), occlusive bronchitis obliterates with interstitial pneumonia (IIP), lymphocytic interstitial pneumonia (LIP), bronchiolitis obliterates with interstitial pneumonia (BIP), and giant cell interstitial pneumonia (GIP). Later on NSIP drew attention in 1994 after Katzenstein and Fiorelli described histopathologic changes in it.[2] These findings were the initial landmark description of uniform inflammation and fibrosis distinct from UIP. Still there were doubts regarding labelling of NSIP as separate disease entity due to association with various CTD and other secondary pathologies.[3]

Some consensus was made in the American Thoracic Society (ATS) international classification, thus labelling NSIP as "provisional type" interstitial pneumonia. Subsequently it was labelled as a separate disease entity in 2008.[4] It was also stressed upon that NSIP is different from other interstitial pneumonia and mostly seen in non-smokers, middle aged women with a good long term prognosis. Recent ATS/ERS guidelines further classified idiopathic NSIP as distinct entity among all interstitial pneumonias.[4,5]

EPIDEMIOLOGY

Exact data on prevalence of NSIP is still unknown. Retrospective cohorts however estimate it to be around 1 to 9 per 100000 population.[6,7]

NSIP is more commonly seen in females' sex and non-smokers in their fifth decade of life. Idiopathic NSIP has relatively good prognosis with 5-year survival >80% and that too with predominant cellular NSIP.[8] Data says that around 15% of patients diagnosed with NSIP are found to have an underlying CTD and around 40% of patients with CTD are diagnosed with ILD later on in their course of illness.[9]

Indian data on incidence and prevalence of idiopathic NSIP is still sparse with few studies on limited patients having concordant findings as of western literature.[10,11] Almost all the studies from north India described male preponderance of ILD in test subjects. Most common aetiology being still CTD and idiopathic. Large multicentre studies are needed to depict the correct demographic pattern of subtypes in our vast country.

CLINICAL FEATURES AND DIAGNOSIS

Detailed history is very important to rule out secondary causes of ILD. Idiopathic NSIP patients are usually middle age females without any smoking history. A good history can rule out specific causes like smoking related ILD namely respiratory bronchiolitis (RB) and Diffuse interstitial pneumonia (DIP), exposure to certain drugs like methotrexate, nitrofurantoin, bleomycin, amiodarone and rituximab.[12] Similar findings are seen in patients with hypersensitivity pneumonitis (HP), thus occupational exposure to birds, hay or industrial particulates must be ruled out. Before making a diagnosis of NSIP always rule out features of CTD like Raynaud's phenomenon, arthralgia, skin rash, dry eyes or xerostomia.[13]

Patients with ILD or NSIP present with chronic cough and exertional breathlessness. There may be constitutional symptoms of arthralgia or weight loss indicating some underlying CTD. On examination there may be fine inspiratory basal crepitations more commonly called as Velcro crepitations. Pulmonary function tests reveal a restrictive type defect with reduced FVC and DLCO.[14]

Making a diagnosis of idiopathic NSIP requires a multimodality approach as it involves ruling out CTD, ocular, renovascular, occupational and drug exposure related factors. Serological tests to rule out CTD includes testing for rheumatoid factor (RAF), anti CCP, antinuclear antibody (ANA) and anti-ds-DNA antibody levels.[8,15] Despite positive serologic results for autoantibodies, patients who do not fit into any CTD can be labelled as idiopathic NSIP. Diagnosing an underlying CTD is therefore important as treatment options vary with each diagnosis, and the prognosis of CTD-ILD is much better than IPF/UIP.

Radiological diagnosis is difficult on basis of chest X-ray alone. Usually, X-ray is performed at beginning, as signs and symptoms may mimic more prevalent conditions such as pneumonia or pulmonary oedema and ARDS. Chest X-ray usually shows increased interstitial markings and predominant lower lobe infiltrates.[16]

However, HRCT shows pretty straightforward changes of classical bilateral Subpleural reticular opacities with traction bronchiectasis and fibrosis.[17,18] Ground glassing and reduced lung volumes may be seen in both X ray and CT scan. Subpleural sparing is not classically seen in all cases. Honeycombing is a feature of UIP and is absent in NSIP patients.[19] HRCT may also reveal any other coexistent pathology thus it is imperative to perform CT in all suspected cases. Thus, it is not possible to diagnose NSIP on the basis of CT alone and needs correlation with history and serological tests to rule out CTD's. Lung biopsy plays a role when cases of UIP may be missed without any changes of honeycombing on HRCT chest.

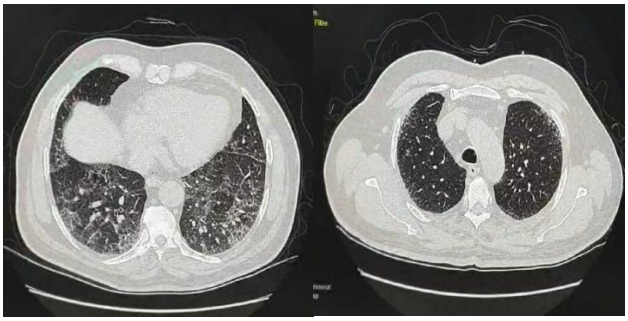

Figure 1: High resolution computed tomography (HRCT) chest in lung window showing subpleural ground glass reticulations and traction bronchiectasis at level of both lower lobes (left) and along subpleural lungs in upper lober (right).

Table 1: Differences in radiographic and pathologic findings in NSIP and UIP.

NSIP	UIP
Subpleural, basal predominance	Subpleural, basal
Ground-glass frequent	Minimal ground-glass appearance
Diffuse or peripheral	Not extensive
Honeycombing rare (~5%)	Honeycombing seen
Reticular abnormality	Reticular abnormality
Traction bronchiectasis	Traction bronchiectasis

Bronchoscopy is an essential tool to rule out other differential diagnosis in patients with strong history of smoking. Some infectious diseases like pneumocystis carinii pneumonia and ABPA are ruled out by specific BAL findings.[14] Otherwise it is of not much use in diagnosing idiopathic NSIP cases. Lymphocytic pleocytosis is seen in cases of IPF versus neutrophilic predominance in NSIP.[21]

For making a definitive diagnosis of NSIP lung biopsy is essential. Predominantly shows diffuse homogenous interstitial inflammation and fibrosis with relatively preserved alveolar architecture.[22] NSIP histopathology pattern can be classified into cellular or fibrotic NSIP based on imflammation or fibrosis. In cellular pattern, infiltration with chronic inflammatory cells is seen with hardly any alveolar wall fibrosis. Fibrotic NSIP is associated with alveolar wall thickening and fibrosis with or without any inflammatory cells infiltration.[13,23,24] Fibrotic NSIP is more common and amounts to almost

80% of the total cases. The extent of organizing pneumonia is less than 10% and associated with smoking or CTD.[18,25]

Table 2: Differences in pathologic findings in NSIP and UIP.

NSIP	UIP
Temporal uniformity with contiguous involvement	Temporal heterogeneity with patchy involvement
Cellular or fibrotic	Fibrosis predominant
No architectural distortion	Architectural distortion
Honeycombing rare	Honeycombing prominent

The term "interstitial pneumonia with autoimmune features (IPAF)" was also proposed for patients with clinical and serological features of any CTD which do not fit into ACR diagnostic criteria.[26] Kinder and colleagues reported that out 75 cases with an 'idiopathic' aetiology of NSIP at their institution, 28 patients met the criteria for undifferentiated connective tissue disease (UCTD). They labelled them as UCTD by the presence of autoimmunity, but without all diagnostic features of a defined rheumatologic disease on the basis of symptoms associated with a CTD and a positive autoantibody marker. Though it is not a validated term for clinical use, we might use it for labelling patients who might benefit from immunosuppressant therapies.

Thus, the diagnosis of idiopathic NSIP can be surely made upon clinical features with insidious onset, typical radiological findings in HRCT chest, lung biopsies and ruling out all other causes including smoking and CTD.

MANAGEMENT

Lacking randomized studies in the treatment of idiopathic NSIP, there is no clear consensus regarding treatment guidelines. As this entity is recently described, we found mixed opinions about whether to treat the condition with immunosuppressants or follow natural course of remission.[20,27] Once progression of disease is noticed, immunosuppression should be started without delay. As a rule, corticosteroids are the first choice of therapy in idiopathic NSIP.[28]

Milder form of disease can be closely followed up with serial HRCT and spirometry readings.[29,30]

Natural remission is not described in idiopathic NSIP; however, it is evident from retrospective data that good number of patients show benefit from steroids and other immunosupressants.[31] Similar results were seen by Watanabe and colleagues, after 1 year of corticosteroid treatment. All 10 patients showed improvement in lung function and oxygen requirement with single mortality after median follow up of 4.5 years.[32]

There is no clear consensus regarding dose and average duration of corticosteroids, but a starting dose of 0.5 to 1.0 mg/kg/day Prednisolone can be used. It is usually tapered over weeks to a minimum required dose as per disease activity. For severe disease, high-dose pulse methylprednisolone therapy (1-1.5 g/day for 3-5 days) may be given followed by a maintenance oral dose as mentioned prior.[32] Park et al also reported that the mean duration of steroid treatment in NSIP was 17.4±12.1 months but with 36% recurrence rate.[33] Kim et al reported recurrence in 20% patients on steroids, which was associated with an initial low dose (0.5 mg/kg) short treatment course of corticosteroid.[34]

Treatment strategy with corticosteroids and other immunosuppressive agents has its limitations. It is most effective in inflammatory phenotype, such as cellular NSIP or organizing pneumonia, but not so in patients with fibrotic NSIP pattern.[35]

Thus, patients not responding to initial corticosteroids or with severe disease to begin with have been deemed fit for other steroid sparing immunosuppressants. Azathioprine, cyclosporine, mycophenolate mofetil, cyclophosphamide, are the commonly used drug for this purpose. They not only have an excellent safety profile but also complement steroid therpay.[36,37]Among this cyclophosphamide has been most frequently studied drug with promising results.[38,39] However these are small retrospective studies and patient profile has to be kept in mind while prescribing these immunosuppressants for long term basis.

Recent literature supports use of rituximab for idiopathic NSIP patients. Keir et al reported beneficial results with rituximab in 50 patients with severe interstitial lung disease who did not respond to any conventional immunosuppressive agent.[40] There are case reports mentioning good results of rituximab in both idiopathic NSIP and CTD associated ILD.[41] It is obvious that with a better safety profile and long duration of action rituximab has a long shot in the treatment of NSIP. Difficult to treat and steroid resistant cases can be managed with it.

Opinion have been raised regarding use of anti-fibrotic drugs in these fibrotic subtypes of IPF.[42] Anti fibrotic like nintedanib has shown promising results in other subtypes of ILD and can be tried in fibrotic variant of idiopathic NSIP as well to halt the progression of disease. Further studies are required in this field.

PROGNOSIS

Overall prognosis of idiopathic NSIP is favourable as compared to its counterparts and similar to CTD-ILD.[10,33,43] Survival depends on co morbidities and baseline lung function along with age. 5-year mortality of 20% has been described in this illness.[44] Out of total 67 cases of idiopathic NSIP, 5 and 10-year survival rates of 82% and 73%, respectively were reported in the 2008 ATS report.[4]

There are widely available tools for assessment of severity and prognosis of ILD like The ILD-GAP scoring systems and gender–age–physiology (GAP) (Table 3). These are simple and easily applicable tools that aid in determination of prognosis using readily available information from patient.[45] Histopathologic features at presentation also provide prognostic information about NSIP. Travis and colleagues examined histologic features from 101 patients with idiopathic ILD and compared survival rates. They found out that cellular NSIP patients on lung biopsy at diagnosis had better 5-year survival rates that fibrosing type.[4,46] Thus, performing histopathologic examination at diagnosis is utmost important for detailing the outcomes and counselling.

Table 3: The ILD-GAP model for prediction of prognosis and mortality in idiopathic NSIP.

Abbreviation	Predictor		Points
ILD	**ILD subtype**		
	IPF		0
	Unclassifiable ILD		0
	CT-ILD/idiopathic NSIP		-2
	Chronic HP		-2
G	**Gender**		
	Female		0
	Male		1
A	**Age (years)**		
	60		0
	61-65		1
	>65		2
P	**Physiology**		
	FVC, % predicted		
	> 75%		0
	50-75%		1
	<50%		2
	DLCO, % predicted		
	>55%		0
	36-55%		1
	35%		2
	Cannot perform		3
	Total possible points		8
ILD-GAP index	**1-year**	**2-year**	**3-year**
0-1	3.1	6.6	10.2
2-3	8.8	18.0	26.9
4-5	18.2	35.0	49.2
>5	33.5	58.4	74.8

MONITORING AND FOLLOW UP

Clinical examination with repeated imaging is the gold standard of care and monitoring in idiopathic NSIP. Dyspnoea can be quantified using several clinical parameters like modified medical research council scale (mMRC) and baseline dyspnoea index (BDI).[47] An effective tool for assessment over time is timely monitoring of pulmonary function tests. Changes in spirometry values are useful indicators of survival.[48] Decline in DLCO by over 12 months has been found to correlate with high (15%) mortality. Measurements are however having objective variation with improper techniques and training. Restrictive lung diseases have a more reliable parameter i.e. forced vital capacity (FVC). It is easily reproducible and has an independent association with mortality.[46,49] Patients who show decline in FVC >10% or DLCO >15% should ideally be referred for escalation of treatment and/or lung transplantation.

CONCLUSION

Idiopathic NSIP (iNSIP) is a relatively favourable and treatable variant of ILD. Histologic confirmation helps in diagnosis and prognostication. Similarities with CTD-ILD makes necessary for thorough immunological workup. Patients who are ideal candidates must be started on immunosuppressants soon. Long term survival with treatment is much better than UIP, thus all patients must be offered the choice of treatment. Monitoring and follow up can be done by simple spirometry on an outpatient basis. Thus, it is a lesser evil when seen on a broad perspective.

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