FRI0608

ASSOCIATION BETWEEN INTERSTITIAL LUNG DISEASE AND RHEUMATIC DISEASE: IMPLICATIONS IN CLINICAL PRACTICE

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Background: The relationship between interstitial lung disease (ILD) and rheumatic diseases is well known. Recently, a new clinical entity has been described that relates ILD with autoimmune findings, IPAF (Interstitial Pneumonia with Autoimmune Features), which allows identifying and condensing non-cataloged diseases with pulmonary and rheumatic involvement.

Objectives: Our aim is to establish which patients with ILD have a rheumatic disease, and which of them meet the IPAF criteria. We analyzed the role of nailfold capillaroscopy in these patients.

Methods: This is a prospective descriptive observational study. The observation period was 2 years (2016-2018), in a county university hospital. Patients diagnosed with ILD of unknown etiology, derived from Pneumology Service, were included. A medical history was obtained focused on autoimmune disease, rheumatological evaluation, rheumatological blood markers and nailfold capillaroscopy, in order to establish whether they had a concomitant rheumatic disease.

Results: Thirty patients with ILD were evaluated, with a mean age of 70,5 years (53 - 88). Of the 30 subjects, 12 (40%) had usual interstitial pneumonia (UIP), 11 (36.67%) had organizing pneumonia (OP) and 7 (23.33%) had nonspecific interstitial pneumonia (NSIP). Eleven (36.67%) of the 30 patients had an altered capillaroscopy, 6 of them (54.5%) had a rheumatic disease. In our sample, 5 cases meet diagnostic IPAF criteria, 4 OP and 1 UIP, three of them with altered capillaroscopy.

Conclusion: In our serie, one third of the patients were diagnosed with rheumatic disease associated with interstitial lung disease. Of the thirty evaluated patients, 36.67% had altered capillaroscopy, 54.5% presenting concomitant rheumatic disease. Following the IPAF classification criteria, we obtained 5 cases in our sample: four OP and one UIP, 3 of them with altered capillaroscopy. It is important to be aware of this association with a multidisciplinary approach for the adequate diagnosis and follow-up of these patients.

ILD	Capillaroscopy	Raynaud	Acropachy	Relevant issues	Rheumatology diagnosis
UIP	Altered (angiogenesis)	Yes	Yes	Aldolase, cANCA	Polymyalgia rheumatica
UIP	Altered (angiogenesis)	Yes	No	ANA 1/640 nucleolar	Systemic sclerosis
UIP	Altered (angiogenesis)	Yes	No	CK, aldolase, anti-Ro	Sjögren Syndrome
UIP	Altered (active pattern)	Yes	No	ANA 1/320 speckled	Systemic sclerosis
UIP	Normal	No	Yes		
UIP	Normal	No	Yes	ANA, anit-DNA, pANCA, anti-MPO	IgA nephropathy
UIP	Normal	Yes	No	Anti-PPC	IPAF
UIP	Normal	Yes	Yes	FR, anti-PPC	Rheumatoid arthritis
UIP	Normal	No	Yes		
UIP	Normal	No	Yes		
UIP	Normal	Yes	Yes	ANA, aldolase, FR, anti-PPC	Rheumatoid arthritis
UIP	Normal	Yes	No	Anti-Ro, anti-La	Sjögren Syndrome
OP	Altered (angiogenesis)	Yes	No		IPAF
OP	Altered (enlarged capillaries)	Yes	No	Anti-Ro	IPAF
OP	Altered (angiogenesis)	No	No	ANA 1/1280 hom	IPAF
OP	Normal	No	No	ANA 1/320 hom	IPAF
OP	Normal	No	No		
OP	Normal	No	No		
OP	Normal	No	No		

ILD	Capillaroscopy	Raynaud	Acropachy	Relevant issues	Rheumatology diagnosis
OP	Normal	No	No		
OP	Normal	No	No		
OP	Normal	No	No		
OP	Normal	No	No		
NSIP	Altered (angiogenesis)	No	Yes		
NSIP	Altered (Dermatomyositis)	No	No	Aldolase, anti-Ro, anti-MDA5	Dermatomyositis
NSIP	Altered (hemorrhages)	No	No		
NSIP	Altered (Dermatomyositis)	No	No	CK, aldolase, anti-Jo1	Antisynthetase syndrome
NSIP	Normal	No	No		
NSIP	Normal	No	No	RF, anti-PPC	Rheumatoid arthritis
NSIP	Normal	No	No	SCLE, aRo, aLa	Sjögren Syndrome

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EXPERIENCE WITH BIOLOGIC AGENTS FOR THE TREATMENT OF CARDIAC SARCOIDOSIS IN A U.S. ACADEMIC MEDICAL CENTER

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Background: Sarcoidosis is a multisystem granulomatous disease of unclear etiology characterized histologically by non-caseating granulomas. Lungs are the most common organs affected but sarcoidosis can affect almost any organ system. While clinically manifest cardiac involvement occurs in only about 5% of patients with sarcoidosis, a significant proportion have clinically silent disease. Symptomatic cardiac involvement portends a poorer prognosis with manifestations varying from heart failure and conduction abnormalities to ventricular arrhythmias including sudden death. Immunosuppression with corticosteroids and DMARDs such as methotrexate and mycophenolate mofetil has been the mainstay of treatment despite a paucity of data. There is a subset of patients that are either non-responders to these agents or in whom the side effect profile is prohibitive for their long term use. Biologic agents, mainly TNF alpha antagonists, have been used as salvage therapies in these patients. However, the evidence regarding their efficacy and safety is limited to a few case reports. In fact, there remains much apprehension regarding the use of TNF alpha antagonists in patients with systolic heart failure due to concerns that they can exacerbate heart failure.

Objectives: To study the efficacy and safety of using biologics for the treatment of cardiac sarcoidosis.

Methods: We conducted a retrospective and prospective observational study of all adult patients with cardiac sarcoidosis treated with biologics at an academic medical center in Washington D.C, USA between 2013 and 2018.

Results: We identified 9 patients (3 men and 6 women) diagnosed with cardiac sarcoidosis at our institution. The mean age at diagnosis was 49.9 (SD 8.6). 1 patient was Caucasian and the rest (n=8) were African American. Lungs were the most common extra cardiac organ involved (n=7) followed by CNS (n=4), liver (n=4) and skin (n=3). 5 of the patients presented with systolic heart failure (EF<50%), 3 with atrial and

ventricular arrhythmias and 1 was found to have incidental abnormal myocardial uptake on PET imaging. 8 of the 9 patients had abnormal myocardial uptake on PET imaging. All but 1 patient had been initially treated with oral steroids (1 refused) and 7 of the 9 patients had also been given oral DMARDs; methotrexate (n=6), azathioprine (n=2), hydroxychloroquine (n=2) and mycophenolate mofetil (n=1). Biologics used were adalimumab (n=5), infliximab (n=3) and rituximab (n=1). The most common indication for biologics was progression of disease despite optimal doses of standard therapy, followed by intolerance or contraindication to standard therapy. 75% of the patients were noted to have marked clinical improvement with the addition of a biologic. 4 out of 9 patients had decreased myocardial uptake on PET following treatment with a biologic. One patient had no change on PET and 4 have not had repeat imaging done yet. None of the patients had worsening of left ventricular systolic function with the addition of a TNF alpha antagonist. There were no reported major infections or significant adverse events that were attributable to the use of biologics.

Conclusion: Based on our small cohort, biologics (mainly TNF alpha antagonists) appear to be safe and efficacious as salvage therapy for cardiac sarcoidosis. However, there is a need for prospective studies to further validate these findings as well as to identify the subset of patients that would benefit from early initiation of these therapies.

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FRI0610

CRYOPYRIN-ASSOCIATED PERIODIC SYNDROME: A SERIE OF CASES IN A ADULT'S PATIENTS COHORT WITH AUTOINFLAMMATORY SYNDROME IN A THIRD LEVEL HOSPITAL

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Background: The Cryopyrin-associated Periodic Syndrome (CAPS) or cryopyrinopathies are part of the spectrum of autoinflammatory diseases. The term cryopyrinopathy encompasses three clinical entities such as: CINCA/NOMID (chronic infantile neurologic, cutaneous and articular/neonatal onset multisystem inflammatory disease), Muckle-Wells disease and FCAS (familial cold-associated periodic syndrome). There are three different diseases that would be part of the same clinical spectrum, ranging from CINCA/NOMID (more serious) to FCAS (the less severe). It is more frequent in pediatric age although they can also be observed in adulthood. They are characterized by mutations in the NLRP3 gene and have an autosomal dominant inheritance pattern

Objectives: To describe the clinical characteristics and genetic variants of a cohort of patients in adulthood diagnosed with cryopyrinopathy and with follow-up in a 3rd level hospital

Methods: Retrospective descriptive study in adults patients with diagnosis of cryopyrinopathy since 2013 (year of introduction of genetic tests in the hospital laboratory) until now. The data was obtained from the review of medical records. All patients with mutations in NLRP3 gene and clinically compatible with this diagnosis were reviewed.

Results: Of a total of 44 patients in adulthood diagnosed with periodic fever syndromes (FMF, TRAPS, cryopyrinopathies, HIDS) and compatible genetic mutations, 7 patients (15.9%) were diagnosed with cryopyrinopathies, presenting 6 of them (13, 6%) mutations in NLRP3 gene. 1 patient was diagnosed with Muckle Wells based on clinical criteria without genetic test. 6 patients (13.6%) were women. 4 patients (9.1%) presented mutations in heterozygosis in exon 3 of NLRP3 gene (p.V198M). 1 patient presented heterozygous mutation in exon 3 of NLRP3 gene (p.R260W) and another patient presented a mutation in heterozygosis in exon 3 of the NLRP3 gene (p.S726G). The mean age at diagnosis was 38 years (IR 13-71 years). The totality of patients diagnosed with cryopyrinopathy showed elevation of acute phase reactants. 6 patients (13.6%) presented fever, joint symptoms (arthralgias and/or arthritis) and myalgias. 4 patients (9.09%) showed cutaneous involvement in the form of urticarial rash. 4 patients (9.09%) showed neurosensory deafness since childhood. 3 patients (6.8%) presented ocular involvement in form of conjunctivitis and/or uveitis. In 3 cases (6.8%), anti-IL-1 (anakinra) or anti-TNF were used.

Conclusion: Cryopyrinopathies are autoinflammatory diseases that occur mainly in the pediatric age, but there are also cases in adulthood. We must consider its diagnosis in those cases with periodic fever, arthralgia or arthritis, sensorineural deafness, APR elevation and urticarial rash. The genetic diagnosis will help us to confirm the diagnosis and avoid a delay in it.

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FRI0611

ACUTE AND LONG-TERM EFFICACY OF IL-1 INHIBITOR ANAKINRA IN IDIOPATHIC AND SECONDARY PERICARDITIS IN PATIENTS REFRACTORY OR INTOLERANT TO CONVENTIONAL THERAPY

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Background: In contrast to its use in idiopathic recurrent pericarditis (IRP), efficacy and optimal treatment duration of Anakinra are unclear in pericarditis secondary to known autoimmune or inflammatory conditions.

Objectives: a) Assess the efficacy of Anakinra in secondary and idiopathic pericarditis, refractory or intolerant to conventional therapy (CT; colchicine and corticosteroid [CS]).

b) Study the effect of extended Anakinra continuation on post-treatment relapse risk.

Methods: Retrospective chart review of 12 adults hospitalised between January 2016 and October 2018 treated with Anakinra 100mg SQ daily for pericarditis refractory or intolerant to CT (CT+A group), and 22 consecutive hospitalised patients treated with CT only (CT group).

Results: Diagnosed autoimmune or inflammatory condition was present in 83.3% (10/12) of CT+A and 40.9% (9/22) of CT group (p=0.07, Table 1). The most common etiologies in CT+A group were RA and Undifferentiated Connective Tissue/Overlap Disease (Figure 1).

Despite CT-intolerance or resistance (Figure 2), all CT+A (12/12) patients had symptom relief at discharge vs. 72.7% (16/22) in CT group (p=0.04). There was a strong trend in CT+A compared to CT group towards shorter time to symptom improvement (1.75±1.29 vs. 3.55±3.06 days; p=0.06) and symptom relief (3.75±1.87 vs. 5.63±3.28 days; p=0.08). CS were tapered successfully in 71.4% (5/7) vs. 66.7% (6/9) of patients in the CT+A and CT groups (p=0.56). Anakinra was associated with a longer mean length of stay (13.0±16.2 vs. 8.68±5.07 days; p=0.25).

No recurrence during treatment was noted in CT+A group (0/12) vs. 40.9% (9/22) in CT group (p=0.009). There was a trend towards reduced post-treatment relapse risk in CT+A group [16.7% (2/12) vs. 41.7% (5/12), p=0.18]. In relapsed patients, there was no significant difference in time to relapse (1.75±0.34 vs. 1.72±0.71 months, p=0.93). Within CT+A group, extended Anakinra use in 6 patients (mean: 5.58±6.5 months) did not reduce post-treatment relapse risk [16.7% (1/6) vs. 16.7% (1/6), p>0.9]. Adverse effects were infrequent; elevated transaminases (n=1) and local injection-site reaction (n=1). Neither required permanent Anakinra discontinuation.

Table 1. Baseline Characteristics

	Anakinra plus conventional therapy % (n=12)	Conventional therapy only % (n=22)	p- value
Age (years +/- SD)	60.3±17.3	56.8±17.8	0.590
Female	66.7 (8)	50 (11)	0.349
Diagnosed autoimmune or inflammatory condition	83.3 (10)	40.9 (9)	0.074
Recurrent pericarditis (>1 episode)	33.3 (4)	27.3 (6)	0.710
Time since initial episode (months +/-SD)	49.0±79.9	9.21±11.7	0.441
Prior NSAID use	25.0 (3)	18.2 (4)	0.638
Prior Colchicine use	25.0 (3)	9.09 (2)	0.210
Prior CS use	50.0 (6)	18.2 (4)	0.051
Pericardial effusion on ECHO	75.0 (9)	81.8 (18)	0.638
Treatment during hospitalisation			
NSAID	50.0 (6)	54.5 (12)	0.799
Colchicine	58.3 (7)	63.6 (14)	0.761
CS	58.3 (7)	40.9 (9)	0.331
Risk factors for pericarditis			
PCI, cardiac surgery or chest trauma	16.7 (2)	45.5 (10)	0.093
Chest radiation	8.33 (1)	4.55 (1)	0.653