Abstract ID – 151

ISOLATED CUTANEOUS SARCOIDOSIS

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Sarcoidosis is a systemic disease that can involve almost any organ system. Infiltration with noncaseating granulomas is the hallmark of the disease. Cutaneous involvement occurs in 20 to 35 percent of patients with systemic sarcoidosis. It is more commonly observed in young and middle age individuals who present with acute-onset sarcoidosis manifested by erythema nodosum associated to bilateral hilar lymphadenopathy and polyarthralgia or polyarthritis. However, isolated cutaneous lesions, without systemic involvement, may occur in the context of chronic sarcoidosis. This report describes a case with isolated cutaneous sarcoidosis (CS). A 48 years old nonsmoking woman admitted to clinic with diffuse papules on faces and plaques on the upper extremities (Figure 1). These lesions were started 3 years ago as small red papules on the extremities than growed with time. Serum ACE level was: 8.1 (normal range: 8.0–52.0), haemoglobin level: 10.1 g/L, sedimentation rate (ESR): 45 mm/hr, serum calcium: 9 mg/dl (normal range: 8.8–10.89 mg/dl). Chest X-ray was normal. Pathology of skin punch biopsy showed noncaseating granulomas. Diagnosis of sarcoidosis is made on these findings. Treatment with 40 mg methylprednisolone was started and dose was reduced to 16 mg with time. Marked improvement occurred especially in skin lesions on the face but little improvement in plaques on extremities. Sarcoidosis occurs more commonly in the 2nd-4th decades and is more prevalent in females. An exclusive cutaneous involvement is rare and it is reported in about 4%–5% of the patients of sarcoidosis. Involvement may be mild or severe, self-limited or chronic, and limited or wide-ranging in extent. The clinical pictures of skin sarcoidosis are variegated: maculopapuler lesions of various size, changes of old scars, lupus pernio, plaque formation, subcutaneous lesions etc. CS with multisystem involvement is treated with topical steroids or systemic corticosteroids. Asymptomatic patients with isolated lesions may not need systemic corticosteroids and they are followed up. Isolated CS is a rare presentation, the diagnosis of which is likely to be missed. Skin biopsy must be done to make definitive diagnosis of sarcoidosis.

Abstract ID – 170

PREDICTION OF EXERCISE-INDUCED HYPOXAEMIA IN PATIENTS WITH IDIOPATHIC INTERSTITIAL PNEUMONIAS WITHOUT RESTING HYPOXAEMIA

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Purpose: The aim of this study was to validate a proposed cut-off of baseline-SpO2 ≤ 90% as a simple screening procedure to predict exercise-induced hypoxaemia (EIH) during a six-minute walk test (6MWT).
Method: We evaluated measurements from a lung function test, 6MWT, blood gas analysis, etc. A SpO2 of less than 90% was defined as EIH. Participants were assigned into non-EIH and EIH groups based on results from performing a 6MWT. We discussed comparing lung function, 6-minute walk distance, SpO2 of rest, etc. The factors were examined using univariate analysis and multiple linear regression analysis. The sensitivity and the specificity created a cut-off from the Area Under the Curve (AUC) and the Receiver Operating Characteristic (ROC) curve. Significances were counted at the 0.05 level.
Results: We retrospectively studied 64 idiopathic interstitial pneumonias (IIPs) patients (67 ± 10.7 y.o., 16were female and 48 male). Thirty patients exhibited EIH. In multiple linear regression analysis, the independent predictors of EIH were %RV (odds ratio (OR), 0.906; 95% confidence interval (CI), 0.844–0.971; $p < 0.001$), A-aDO2 (OR, 1.135; 95% CI, 1.033–1.246; $p < 0.001$) and %DLCO (OR, 0.951; 95% CI, 0.913–0.991; $p < 0.001$). The ROC curve showed that the %RV cut-off was 73%, with AUC 0.814, $p < 0.001$; sensitivity 65.7%, specificity 75.9%, positive predictive value 76.7%, and a negative predictive value 64.7%. A-aDO2 was 22.9 torr, with AUC 0.806, $p < 0.001$; sensitivity 72.7%, specificity 80.6%, positive predictive value 80%, negative predictive value 73.5%. %DLCO was 69%, with AUC 0.764, $p < 0.001$; sensitivity 68.8%, specificity 75%, positive predictive value 73.3%, and a negative predictive value 68.6%. If two items or more were positive, sensitivity was 85% or more.
Conclusion: This study suggests that %RV, A-aDO2 and %DLCO are objective indicators that predict EIH.
Clinical Impact: Until now, it has been difficult to predict EIH in patients without resting hypoxaemia. This study may help to better evaluate a situation and avoid EIH during patient exercise.

Abstract ID – 172

A RARE CAUSE OF INTERSTITIAL LUNG DISEASE IN IRANIAN GIRL: HERMANSKY-PUDLAK SYNDROME

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Hermansky-Pudlak syndrome (HPS) is a multisystem disorder characterized by: tyrosinase-positive oculocutaneous albinism; a bleeding diathesis resulting from a platelet storage pool deficiency; and, in some cases, pulmonary fibrosis or granulomatous colitis. HPS is common only in northwest Puerto Rico. However, it is extremely rare in other regions. A 24-year-old girl present to our clinic with cough and dyspnoea. On physical examination she had oculocutaneous albinism and nystagmus. Fine crackles in the bilateral lower lung fields were detected, but heart sounds were normal. He had neither clubbed finger nor oedema. The results of abdominal and neurological examinations were normal. Examination of gum shows no bleeding. She has no history of excessive bleeding and easy bruises. She had undergone fibrotic bronchoscopy with bronchioalveolar lavage with initial diagnosis of tuberculosis, which was negative for acid fast bacilli. She had tests for bronchiectasis. Chest x-ray showed bilateral reticulonodular infiltration. A thorax CT revealed bilateral diffuse interlobular septal thickness and CT and traction bronchiectasis. Following investigations, she was diagnosed with HPS. Therefore HPS should be actively sought, monitored and risk factors addressed in individuals with oculocutaneous albinism even without bleeding diathesis to prevent unnecessary examinations.

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THE "CRAZY PAVING PATTERN" LUNG IN A PREGNANT WOMAN

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Pulmonary Alveolar Proteinosis (PAP) is a rare disorder characterized by an abnormal accumulation of a PAS positive material within the alveolar spaces and distal bronchioles. A 29 year old lady in her 29th week of pregnancy presented with prolonged dry cough for two months which was associated with reduced effort tolerance and low grade fever. At presentation she is hypoxic and required oxygen supplement. She was initially treated empirically for tuberculosis but she continued to have worsening hypoxia. A high resolution CT thorax revealed a crazy paving pattern lung which was suggestive of PAP. She delivered a healthy baby at 36 weeks via an elective Caesarean section. Post operatively she underwent diagnostic bronchoalveolar lavage which showed acellular homogenous eosinophilic material which was positive for PAS/PASD and negative for mucin. Therapeutic whole lung lavage was performed and her symptoms and lung function tests improved. In this case report, we will discuss how we made the diagnosis of PAP and share our experience in managing a pregnant woman with this condition.