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Title: Pathological findings in histiocytosis X with pulmonary hypertension

Dr. Javier 19367 Villuela jvilluela@gmail.com MD [1], Dr. Jose 19368 Cifrian jose492@separ.es MD [1], Dr. Beatriz 19370 Abascal babascal@humv.es MD [1], Dr. Alejandro 19371 Daly adaly@humv.es MD [1], Dr. Carlos 19374 Amado camado@humv.es MD [1], Dr. Sonia 19379 Fernandez-Rozas sfernandez@humv.es MD [1], Dr. David 19385 Iturbe diturfe@gmail.com MD [1], Dr. Felipe 19386 Zurbano nmlzgf@humv.es MD [1], Dr. Roberto 21385 Mons rmons@humv.es MD [3] and Dr. Javier 19369 Gomez-Román apagrj@humv.es MD [2]. [1] Pneumology, Hospital Marques Valdecilla, Santander, Spain ; [2] Patholgy Department, Hospital Marques Valdecilla, Santander, Spain and [3] Thoracic Surgery, Hospital Marques Valdecilla, Santander, Spain .

Body: Objective. To evaluate the hemodynamic characteristics, pulmonary function and pathological findings in patients with Langerhans cell histiocytosis (LCH) and Pulmonary Hypertension (PH) not explained otherwise. Methods. A retrospective study was conducted in patients with Langerhans cell histiocytosis. Echocardiogram, lung biopsies, lung function tests and hemodynamic registries were reviewed. Results. Twenty patients were studied, with a mean age of 41 ± 10 years. 8 patients (40%) had severe PH.The median delay between the diagnosis of LCH and PH was 2.1 years. Six minute walk distance was 409 ± 101m. Systolic PAP: 62 ± 9.9 mmHg. FVC was 62±15% of predicted, FEV1 45±21 % and DLco 41±13. All patients were on long-term oxygen therapy. After a median follow-up of 9.1 years,1 patient is clinically stable, 1 patient had died of cardiac arrest while waiting for lung transplant, and 6 patients had undergone lung transplantation with 83% survival at 1 and 3 years. Histopathological lesions studied in explanted lungs suggested a veno-occlusive origin for this kind of complication, with capillary haemangiomatosis-like changes and fibrosis of septa, something that has been rarely described in LCH. Otherwise, it is interesting to remark the presence of three cases of malignant neoplasias in the global serie. This association has been described as isolated cases in literature review. Two patients suffered from malignant lymphoma and one female patient had an uterine cervix carcinoma. Conclusions: Pulmonary Hypertension could have a veno-occlusive origin in LCH. A relationship between malignant neoplasias and LCH may be possible.