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Title: New entity of an isolated pulmonary non-Langerhans cell histiocytosis

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Body: Background: Histiocytic disorders are rare, heterogeneous diseases with abnormal tissue infiltration by histiocytes. So far, pulmonary manifestation in Non-Langerhans cell histiocytoses (NLCH) occurred only as a part of a systemic disease. Case: A 37 year old male stonesculptor suffered from progressive dyspnea and thoracic tightness. He was admitted under suspicion of pulmonary capillary hemangiomatosis (PCH). Asthma was diagnosed 6 years ago and he had a former smoking history of 5 pack years. Pulmonary function testing demonstrated obstructive lung disease (FEV1 48%) with negative bronchospasmolysis and compromised diffusion capacity (DLCO 50%, KCO 54%). Cardiopulmonary exercise testing showed a maximum work load of 150 W with VO2peak 20 ml/min/kg (56%). Surgical lung biopsies excluded PCH and revealed interstitial histiocytic infiltrates. These showed immunohistochemical positivity for CD68 and negativity for CD1a and Langerin, therefore NLCH was diagnosed. CT and metabolic imaging with F-18 FDG PET/CT scanning -as used diagnosing manifestation in Erdheim-Chester disease- demonstrated bipulmonary, FDG-avid nodular infiltrates and excluded extrapulmonary manifestations. After multidisciplinary discussion a watch and wait strategy was chosen. The patient is currently stable since 18 months with regards to clinical, functional and radiological parameters. Conclusion: Here we describe a new entity of an isolated pulmonary NLCH. Clinical, patho- & radiological features are not consistent with any other NLCH reported so far. The association with asthma might be coincidental. The etiopathogenesis is unclear and might however be occupational or smoking-related in this case. Long term prognosis seems to be dubious.