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CASE REPORT

Idiopathic Pulmonary Lymphoid Diffuse Hyperplasia

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ABSTRACT: ***Idiopathic Pulmonary Lymphoid Diffuse Hyperplasia. P.A. Canessa, L. Pratticò, F. Fiasella, A. Cavazza, B. Bacigalupo, F. Fedeli.***

We describe a 70 year old woman affected by diffuse pulmonary lymphoid hyperplasia (DPLH). DPLH is a rare clinical-pathological entity generally associated with connective tissue diseases, but we diagnosed our case as an idiopathic DPLH. To the best of our knowledge, this is the first case of idiopathic DPLH reported in medical literature.
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Case report

In February 2003, a 70 year-old non-smoker housewife without any para-occupational condition was examined. She had a one-month's history of dyspnoea, cough and fever; antibiotics had been administrated for three weeks without improvement and she was admitted to the hospital for further evaluation.

She denied any drug use, a history of toxic inhalation or drug abuse. Physical examination was negative. The laboratory tests produced results that were within the normal range, and an ECG was normal.

Arterial blood gas values were: pH 7.461, PaO_2 95.9 mmHg, $PaCO_2$ 33.6 mmHg. A chest X-ray showed bilateral lower-zone coarse reticulonodular infiltrates with consolidated alveolar opacities.

The pulmonary function tests gave the following results: TLC 75% of predicted value, RV 76%, VC 73%, FVC 76%, FEV_1 76%, DLCO 53%, DLCO/VA 56%. The bronchodilatation test was negative.

A chest-CT showed a number of small nodular opacities in the centrilobular and in the subpleural areas of the superior and inferior lobes with interlobular septal thickening and with alveolar consolidation in inferior lobes (figure 1A).

The immunity tests were negative (Rheumatoid factor, ANA, anti SSA/Ro and anti SSB/La antibodies, anti Sm, anti RNP, SCL70, Jo1, ASMA, anticardiolipin antibodies).

No infectious pathogens were detected in blood (antibody dosage for toxoplasma, mycoplasma, mononucleosis, chlamydia, rubeola, herpes simplex, borrelia; CMV-DNA and CMV-antigen, HIV, Aspergillus), in urine (pneumococcus and legionella antigens) and in BAL (P Jirovecii, Koch Bacillus).

Bronchoscopy was normal; the transbronchial biopsies performed in the left lower lobe showed a dense lymphoid infiltration of the chorion. The BAL was: macrophages 29%, neutrophils 28%, lymphocytes 43% with CD4/CD8 ratio 0.4.

No specific diagnosis was reached, and a videothoracoscopic biopsy of the lingula was performed, showing a dense lymphoplasmacellular infiltration localised mostly along the lymphatic routes (fig. 2A). This infiltration had produced follicles and occasional lymphoepitelial complexes with small accumulations of fibrin, endoalveolar macrophages and foci of subacute organising pneumonia (figure 2B). A Grocott and a Ziehl-Neelsen stains were negative. Immunohistochemically, the lymphoid infiltrate was mixed, B (CD20 +) and T (CD3 +), and the plasmacells were polyclonal with light chain immunoglobulins. A bone-marrow biopsy was normal.

The patient was treated with 25 mg/die of prednisone for six months, and she had a clinical improvement. In November 2003 chest CT was improvement (fig 1B), DLCO values were normal (90%) and prednisone was reduced at 12.5 mg/die. After 18 months autoimmunity tests remained negative. After 36 months the patient's follow up showed clinical and radiological stability with 12.5 mg/die of prednisone. She did not have any CVD clinical and serological evidence. This was the minimal effective dosage to control the disease; below this dosage, the patient suffered from coughing and dyspnoea.

Discussion

Lymphoid tissue is uncommon in normal adult human lungs. In non-smoking adults, the majority of

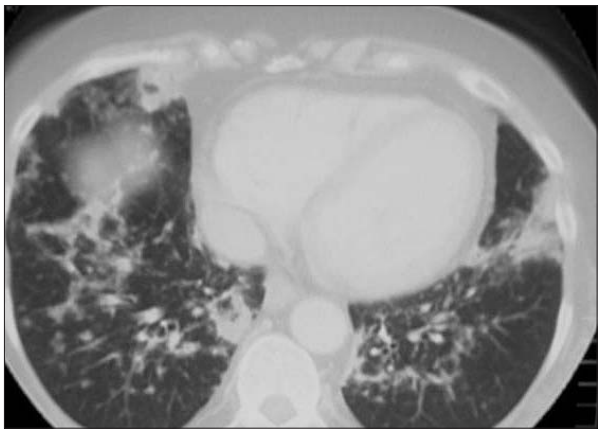

Fig. 1A. - Chest CT (February 2003) bilateral multiple nodules and alveolar consolidation areas.

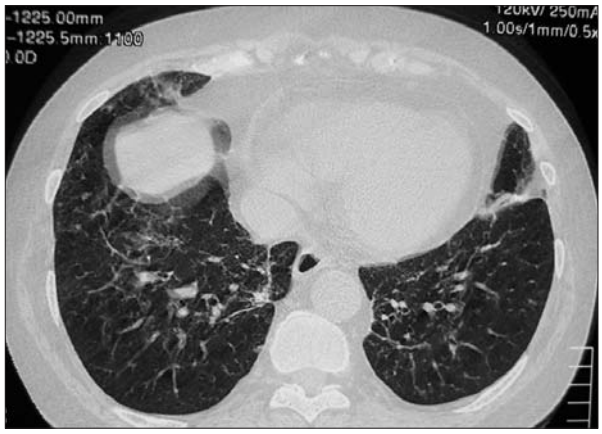

Fig. 1B. - Chest CT (November 2003) no nodules and reduction of alveolar consolidation.

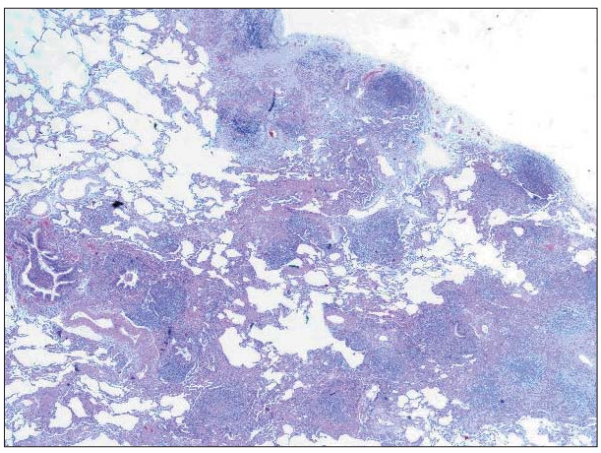

Fig. 2A. - Videothoracoscopic biopsy, showing lymphoid aggregates along the lymphatic routes (Hematoxylin-Eosin, 20x).

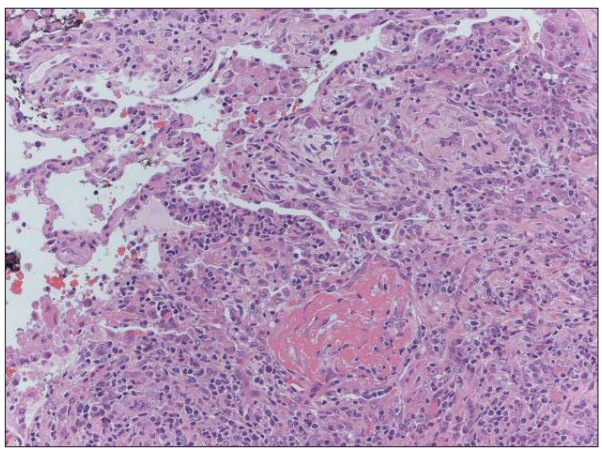

Fig. 2B. - Tiny foci of organising pneumonia with fibrin and intra-alveolar macrophages were present. (Hematoxylin-Eosin, 200X)

pulmonary lymphoid tissue is formed into poorly-organised aggregates predominantly located at bronchial divisions and adjacent to distal respiratory bronchioles [1]. Additionally, small numbers of lymphoid cells are occasionally found scattered beneath and between bronchial epithelial cells [2]. Approximately 60% of lung lymphoid cells are B cells, and the rest are T lymphocytes [3]. The constellation of non capsulated lymphoid follicles, cellular collections, and loosely distributed mucosal lymphocytes constitute the bronchus-associated lymphoid tissue (BALT) or "pulmonary microtonsils" [4].

The acquired benign diffuse proliferations of BALT includes follicular bronchitis/bronchiolitis (FBB); diffuse pulmonary lymphoid hyperplasia (DPLH); and lymphocitic interstitial pneumonia (LIP) [4].

DPLH is characterised by lymphoid infiltrates localised in the pleura, in the interlobular septae and along bronchovascular bundles, and differs from LIP because a significant alveolar septal involvement is lacking; however, a morphological overlap exists between DPLH, FBB and LIP [5]. Moreover, the differential diagnosis with low-grade MALT lymphoma can be difficult and requires immunohistochemical tests.

DPLH is a rare clinical-pathological entity generally associated with connective tissue diseases, especially Sjögren's syndrome or rheumatoid arthritis [5], immunodeficiency syndrome [4] and infections [4].

In our patient's case, her history and laboratory tests have excluded any association between her condition and the diseases listed above. Therefore, we have diagnosed our case as an idiopathic DPLH. To the best of our knowledge, this is the first example of idiopathic DPLH reported in medical literature.

References

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