CASE REPORT

Primary mucosa-associated lymphoid tissue lymphoma as a pleural mass

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ABSTRACT. Primary pleural lymphoma is a rare entity that has been described in association with human immunodeficiency virus (HIV) infection or pyothorax. It occurs in only 7% of primary lymphoma cases. We report the case of a 52-year-old male with no history of HIV infection or pyothorax who was diagnosed of obstructive sleep apnoea syndrome (OSAS) and underwent a routine chest X-ray to screen for any organic cause of the symptoms. The radiograph revealed two voluminous masses with extraparenchymatous features but without pleural effusion. A contrast-enhanced CT was performed and confirmed the existence of the pleural masses that showed homogeneous attenuation. Neither mediastinal lymphadenopathy nor pleural effusion were present. A percutaneous CT-guided fine needle aspiration cytology (FNAC) with a 25-G needle was performed (two samples were obtained) and the first diagnosis was of non-Hodgkin's lymphoma. The final diagnosis of primary pleural mucosa-associated lymphoid tissue (MALT) lymphoma was confirmed by a CT-guided core biopsy with a 20-G needle. To the best of our knowledge, no cases of MALT lymphoma presenting as pleural masses without pleural effusion have been reported in immunocompetent patients. In this report, we describe the case of a patient with a primary pleural MALT lymphoma and include a short review of the literature.

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Pleural lymphoma is a rare type of primary lymphoma, accounting for only 7% of primary lymphoma cases. It usually affects patients with human immunodeficiency virus (HIV) infection or chronic pyothorax [1, 2]. However, an extremely rare type of primary pleural lymphoma has been described in immunocompetent patients with no history of chronic tuberculous pyothorax. The radiological findings consisted of pleural effusion associated with a pleural mass, consolidation or inhomogeneous pleural thickening [3–6]. Although pleural effusion is a common manifestation of both secondary and primary pleural lymphoma, in this case there was no pleural effusion and the diagnosis was initially obtained by CT-guided fine needle aspiration cytology (FNAC) of an extraparenchymatous mass and confirmed by a CT-guided tru-cut biopsy.

Case report

A 52-year-old male came to the Radiology Department of the Hospital Universitario La Paz, Paseo de La Castellana, Madrid, Spain, to undergo routine chest radiography to screen for any organic cause of obstructive sleep apnoea syndrome (OSAS) or any other medical

condition related to this pathology. There was no history of cough, chest pain, dyspnoea, haemoptysis, fever, chills, night sweats, weight loss, occupational exposure or recent travel. He was neither a smoker nor an alcoholic.

On clinical examination, there was no evidence of peripheral lymphadenopathy or hepatosplenomegaly. Laboratory findings were normal.

Serological tests were negative for HIV infection and the Mantoux test was also negative.

The chest radiograph showed two voluminous masses with extraparenchymatous features, in the right upper lobe and in the left major fissure. No pleural effusion was noted (Figure 1). Then, a chest CT with intravenous iodinated contrast medium was performed and two pleural masses with homogenous attenuation were seen, as demonstrated in the chest radiograph. Neither pleural effusion nor mediastinal enlarged lymph nodes were present. Lung parenchyma appeared normal, without nodules or consolidations (Figure 2a–c). Initially, a tumour composed of fibrous connective tissue was suspected. In order to rule out a fibroid tumour, the patient underwent MRI. This revealed two homogeneous pleural masses that showed hypointense signal on T_1 weighted sequences and were hyperintense on T_2 weighted sequences, resulting in the rejection of the fibroma diagnosis (Figure 2d–e). There was no evidence of signal loss in the fat-suppressed T_2 weighted sequences, which indicated a fat component. A CT-guided FNAC of

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Figure 1. Chest X-ray, posteroanterior projection. Note a mass over the right hemithorax with extraparenchymatous features and a second mass over the left major fissure. No pleural effusion is present.

the biggest mass located in the right superior lobe was performed. Two passes were carried out with a 25-G $\times$ 9 cm needle. A Diff-Quik stain (Dade Behring, Deerfield, IL) of both samples was made immediately (Figure 3) in order to check that there was sufficient and valid material for the cytological diagnosis. Material for

ThinPrep (Hologic, Bedford, MA) liquid-based cytology was also separated and immunocytochemistry techniques (CD20 and CD3), Ig H and T-cell receptor (TCR)- γ polymerase chain reaction (PCR) clonal analyses were made. The smears showed a plentiful lymphoid cellularity of polymorphic appearance with predominance of medium-sized lymphocytes with a basophilic cytoplasm just visible at the edge of the cell, intermingled with a small number of bigger cells probably from the germinal centre and isolated histiocytes. A clear predominance of B-cells (CD20+) over a minority of T-cells (CD3+) was observed. Using PCR analysis, monoclonal rearrangement of the fraction Fr3a of the Ig H was identified. With these findings, the diagnosis of non-Hodgkin's lymphoma was made. Finally, the diagnosis of MALT-type lymphoma was made by means of 2 percutaneous samples of CT-guided tru-cut biopsy with a 20-G $\times$ 15 cm needle and a papanicolaou stain.

Discussion

Primary pleural lymphoma is an extremely rare type of lymphoma. By contrast, pleural involvement by systemic lymphoma is relatively common. Approximately 16% of patients with non-Hodgkin's lymphoma present with pleural disease or subsequently develop pleural involvement during the course of the disease [1, 7]. Most of the primary pleural lymphomas are associated with HIV infection or chronic tuberculous pyothorax [1, 8].

(a)

(b)

(c)

(d)

(e)

Figure 2. Chest enhanced CT, (a) axial, (b) coronal and (c) sagittal images that show two pleural masses with homogeneous attenuation located in right upper lobule and left major fissure without pleural effusion. (d) Chest MR, coronal T_1 weighted sequence demonstrates that both masses are hypointense in this sequence. (e) Coronal T_2 weighted sequence shows a hyperintense signal in the two lesions, rejecting the clinical suspicion of a fibrous tumour.

Figure 3. Diff-Quik stain of a smear obtained from the biggest pleural mass. Abundant lymphoid cell population with polymorphous appearance is shown. Notice the predominance of CD20-positive cell population over a CD3-positive one.

Two forms of primary pleural high-grade lymphoma are described: primary effusion lymphoma (PEL) and pyothorax-associated lymphoma (PAL) [8]. Both are clinically aggressive and have well-characterised clinical and pathological features. Various mechanisms have been implicated for the development of primary pleural lymphoma, including chronic stimulation of B cells by longstanding chronic pleural disease, which occurs in different diseases such as tuberculous pleuritis, Sjögren's syndrome, rheumatoid arthritis, malignant lymphoma, chronic lymphocytic thyroiditis, thyroid lymphomas and Epstein-Barr virus (EBV) infection [3, 8]. PEL has been reported to be strongly associated with human herpesvirus-8 (HHV-8) infection and occasionally with EBV infection [4]. Findings in PEL consist of pleural effusion. The pericardial and peritoneal cavities may also be involved, but there is absence of any pleural mass. On the contrary, PAL usually presents as a homogeneous or inhomogeneous non-enhancing soft tissue mass with or without adjacent bone destruction. However, as we have described, an extremely rare type of primary pleural lymphoma can affect immunocompetent people with no history of tuberculous pyothorax or HIV infection [1, 5, 8]. The histological subtype in these few cases referred to in the literature was extranodal marginal zone lymphoma [4, 5]. The radiological findings of these rare cases were pleural effusion associated with a pleural mass, consolidation or inhomogeneous pleural thickening [3–5].

In a general consideration, primary pleural lymphoma appears normally as a localised, broad-based area of plaque-like pleural thickening [8], a solitary nodule or diffuse nodular pleural thickening [9, 10]. Occasionally, pleural lymphoma can present as an isolated pleural

effusion prior to the development of a demonstrable mass [11]. The diagnosis of primary pleural lymphoma seems to be difficult in most cases. Pleural biopsy and pleural fluid cytological examination are usually the first steps in the diagnostic workup of patients with pleural-based masses [7]. We consider evaluation with CT and MR imaging studies and a CT-guided FNAC to be of great importance.

The above case illustrates an example of MALT-type lymphoma with initial pleural presentation. Although pleural effusion is a common manifestation of both secondary and primary pleural lymphoma, a pleural mass without pleural effusion has not been described previously as a manifestation of any type of primary pleural lymphoma. To our knowledge, this is the first report of a primary pleural MALT-type lymphoma without pleural effusion in an immunocompetent patient without history of tuberculosis or chronic pyothorax.

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