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Case Report
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Primary Non-Hodgkin Lymphoma of the Pleura Associated with Bilateral Chylothorax: An Unexpected Diagnosis

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Abstract

Pleural effusion is a common finding in patients with advanced-stage lymphoma. Primary pleural lymphoma, instead, is a very rare entity that diagnosis is often challenging. This is a very interesting case of a 65-years-old immunocompetent woman affected by bilateral chylothorax as the only clinical manifestation of primary pleural small-B-lymphocytes non-Hodgkin lymphoma. The patients underwent to iterative thoracoscopic pleural biopsy since the mediastinal lymph-nodes biopsy and pleural effusion cytological examination were inconclusive. This case highlights the importance of a deep investigation of every pleural effusion, also with iterative pleural biopsy, because even an underestimated bilateral chylothorax can hide a rare neoplasm.

Keywords: Chylothorax; Pleural lymphoma; Pleural effusion; Non-Hodgkin lymphoma; Talc

Introduction

Pleural effusion is a common finding in up to 20% of patients with advanced-stage lymphoma. Primary pleural lymphoma, instead, is a very rare entity, that affects patients suffering from immunodepression, mostly associated with an exudative pleural effusion or pyothorax [1,2]. Diagnosis is often challenging, based on clinical presentation (i.e. dyspnea, cough, thoracic pain), radiological findings (Chest-RX or CT-scan) but also and especially on chemical-physical examination of pleural effusion together with the histological features of the pleural biopsies. Due to its rarity, there are scant data regarding diagnosis, treatment and prognosis of the primary lymphoma of the pleura in literature [3-8].

This is a challenging case of a 65-years-old immunocompetent woman who was referred to our Center for bilateral chylothorax. The diagnosis of primary pleural small-B-lymphocytes non-Hodgkin lymphoma was obtained thanks to iterative thoracoscopic pleural biopsy since the mediastinal lymph-nodes biopsy and pleural effusion cytological examination were inconclusive. The patient underwent exclusive chemo-immunotherapy without any evidence of recurrence at 5-year follow-up.

Background

A healthy 65-years-old woman was referred to our Center in August 2013 because of worsening dyspnea and chest pain. She had no history of smoking, professional asbestos exposition or recent thoracic trauma. The chest X-Ray showed bilateral pleural effusion, greater in the left hemithorax. Left thoracoscopy revealed massive pleural effusion with milky-looking fluid and a diffuse hyperemia of the parietal pleural without nodules.

Cytological and microbiological exams on pleural fluid were negative for presence of bacteria or malignant cells, while its chemical and physical characteristics were suggestive of chylothorax (triglycerides 238 mg/dL, total proteins 2.80 g/dL, specific weight 1,020). Moreover, pleural biopsies revealed pleural inflammation with infiltration of a high number of B- and T-lymphocytes, while the mediastinal lymph-node biopsy showed a reactive hyperplasia.

In 2nd post-operative day, a right thoracentesis was performed, with detection of chylothorax

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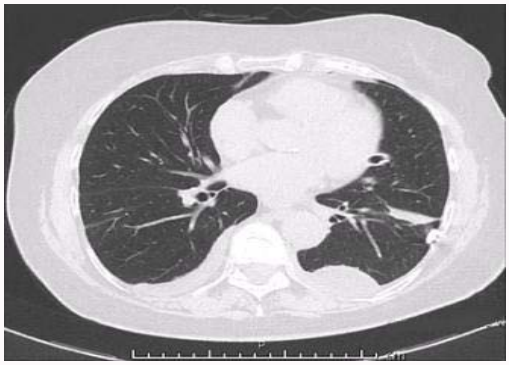

Figure 1: Computed Tomography (CT) image showing bilateral pleural effusion and left pleural thickening.

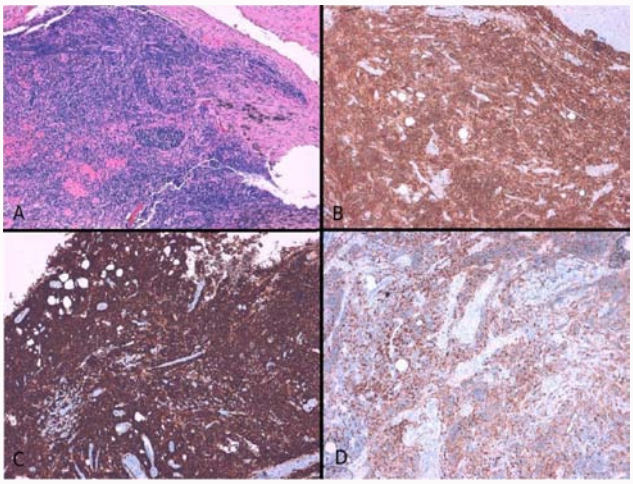

Figure 2: Immunohistochemistry panel 10x: A) H&E stain; B) Bcl-2 IHC Antibody; C) Anti-CD20 Antibody; D) Anti-CD5 Antibody.

again (triglycerides 201 mg/dL, total proteins 5.20 g/dL, specific weight 1,028).

The patient was initially treated conservatively with total parenteral nutrition with alipidic solution, intravenous corticosteroid (Methylprednisolone 20 mg per day) and intramuscular octreotide (0.1 mg in three administrations per day). She was observed for 15 days, during which the fluid leakage was superior to 200 ml daily. A chest-CT scan showed off moderate bilateral pleural effusion, thickening of the costo-vertebral and diaphragmatic pleura bilaterally with mediastinal lymphadenomegaly (Figure 1).

After 20 days, the patient underwent left thoracoscopy again, to obtain costovertebral pleural and mediastinal nodes biopsies and induce a chemical pleurodesis with sterile talc. For the second time the lymph-node histological examination was irrelevant; nevertheless, the pleural specimen revealed small B-cell non-Hodgkin lymphoma with the following Immunohistochemistry: CD20, CD5, Bcl2, CD10, CD43 positive; Bcl6, CD23, CD21, Cycline D1 negative, Ki-67: 5% to 10% (Figure 2).

The postoperative chest X-ray showed the almost complete resolution of the left pleural effusion (Figure 3).

The patient was addressed to the Division of Hematology of our Hospital to start the chemotherapy with Rituximab and Bendamustine (6 cycles). Since then, the patient had a regular follow-up with chest

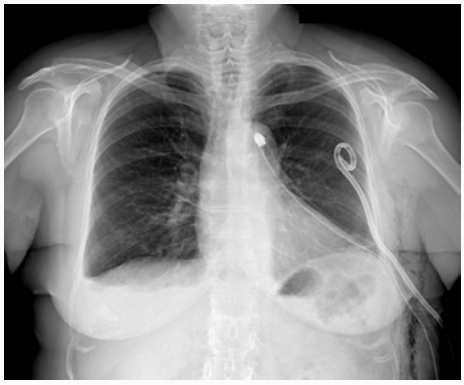

Figure 3: Post-operative chest X-Ray showing resolution of the pleural effusion.

and abdomen CT scans every 6 months with no signs of recurrence.

Discussion

Non-Hodgkin Lymphomas (NHL) are associated with pleural effusion in up to 20% of patients. In most cases, pleural effusion is exudative; nevertheless, it may occur as chylothorax because of lymphatic vessels' neoplastic infiltration or ab-extrinseco compression of the major thoracic ducts by a lymphomatous mass [6,9]. Pleural involvement is generally considered a sign of advanced disease (Ann-Arbor stage III or more), moreover only few cases of primary pleural lymphoma (mostly a large B-cell NHL) are reported in literature and, in those cases, represent an early stage (namely stage I). This distinction is fundamental in establishing treatment and prognosis of patient affected by NHL and the diagnosis is achievable only by pleural and lymph nodes histological examination [10,11].

In the reported case, the patient was affected by a primary pleural small B-cell NHL, which manifested only with bilateral chylothorax associated to pleural thickening and with no evidence of other disease's localization.

Bilateral chylothorax is a rare condition and, in absence of trauma, a malignancy should be considered [6,9,12-14]. As suggested by guidelines for spontaneous chylothorax, we initially treated the patient conservatively with fasting and medical therapy (cortisone and somatostatin analogue) [15]. After two weeks of fluid leakage >200 ml daily we decided to undergo the patient a surgical pleurodesis and thanks to this intervention the diagnosis came out.

Small B-cell lymphocytic lymphoma is mainly a disease of elderly people, the prognosis is defined by the International Prognostic Index (IPI), based on five parameters: age (<=60 years *vs.* >60 years), LDH serum concentration (normal *vs.* abnormal), performance status, Ann Arbor stage (I/II *vs.* III/IV) and number of extranodal sites involved (<=1 *vs.* >1) [16]. In accord to this, our patient was in the first risk group (5-years survival 73%), having only one poor prognostic feature (age >60 years). After diagnosis her performance status was good, so she could receive chemo-immunotherapy.

Conclusion

Chronic pleural effusion may be, sometimes, a great pretender, tough to figure it out. We wanted to focus on such a rare entity as a primary pleural lymphoma, slightly known by scientific community because of its rarity and of its unspecific clinical features. This case highlights the importance of a deep investigation of every

pleural effusion, also with iterative pleural biopsy, because even an underestimated bilateral chylothorax can hide a rare neoplasm.

References

1. Oikonomou A, Giatromanolaki A, Margaritis D, Froudarakis M, Prassopoulos P. Primary pleural lymphoma: plaque-like thickening of the pleura. Jpn J Radiol. 2010;28(1):62-5.
2. Steiropoulos P, Kouliatsis G, Karpathiou G, Popidou M, Froudarakis ME. Rare cases of primary pleural hodgkin and non-hodgkin lymphomas. Respiration. 2009;77(4):459-63.
3. Karadeniz C, Güven MA, Ruacan Ş, Demirbilek S, Sağbil Ş, Akhan O. Primary pleural lhymphoma: an unusual presentation of childhood non-Hodgkin lymphoma. Pediatric Hematol Oncol. 2000;17(8):695-9.
4. Ravikumar G, Tirumalae R, Das K. Primary pleural non-Hodgkin lymphoma in a child - an exceedingly rare disease. J Pediatr Surg. 2012;47(3):E29-31.
5. Kumar A, Harris K, Roche C, Dhillon SS. A 69-year-old woman with lymphoma and chylothorax. Looking beyond the usual suspect. Ann Am Thorac Soc. 2014;11(9):1490-3.
6. Daly MM, Kunkler IH. Chylothorax in non-Hodgkin's lymphoma managed by mediastinal radiotherapy. Clin Oncol (R Coll Radiol). 1995;7(6):397-9.
7. Mares DC, Mathur PN. Medical thoracoscopic talc pleurodesis for chylothorax due to lymphoma. A case series. Chest. 1998;114(3):731-5.
8. Mohamed M, Tan J, Kalpurath KK. Non-Hodgkin lymphoma manifesting as massive malignant chylothorax: successful management with chemotherapy and ambulatory drainages using indwelling pleural catheter. Intern Med J. 2015;45(9):980-3.
9. O'Callaghan AM, Mead GM. Chylothorax in lymphoma: mechanisms and management. Ann Oncol. 1995;6(6):603-7.
10. Vega F, Padula A, Valbuena JR, Stancu M, Jones D, Medeiros LJ. Lymphomas involving the pleura. A clinicopathologic study of 34 cases diagnosed by pleural biopsy. Arch Pathol Lab Med. 2006;130(10):1497-502.
11. Wang Z, Wu YB, Xu LL, Jin ML, Diao XL, Wang XJ, et al. Diagnostic value of medical thoracoscopy in malignant pleural effusion induced by non-Hodgkin's lymphoma. Oncol Lett. 2017;14(6):8092-9.
12. Podder S, Mora M, Patel V, Sivamurthy S. A rare case of bilateral chylothorax: a diagnostic challenge--follicular lymphoma versus primary effusion lymphoma. BMJ Case Rep. 2015;2015.
13. Doerr CH, Allen MS, Nichols FC 3rd, Ryu JH. Etiology of Chylothorax in 203 Patients. Mayo Clin Proc. 2005;80(7):867-70.
14. Teng CL, Li KW, Yu JT, Hsu SL, Wang RC, Hwang WL. Malignancy-associated chylothorax: a 20-year study of 18 patients from a single institution. Eur J Cancer Care (Engl). 2012;21(5):599-605.
15. Bender B, Murthy V, Chamberlain RS. The changing management of chylothorax in the modern era. Eur J Cardiothorac Surg. 2016;49(1):18-24.
16. Shankland KR, Armitage JO, Hancock BW. Non-Hodgkin lymphoma. Lancet. 2012;380(9844):848-57.