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LETTER TO THE EDITOR

Multiple extranodal organ involvement and increase of serum β -HCG in a case of primary mediastinal large B-cell lymphoma



To the Editor,

Primary mediastinal large B-cell lymphoma (PMLBCL) is a distinct subtype of diffuse large B-cell lymphoma. It is usually diagnosed by pathological findings of a mediastinal mass. Multiple extranodal organs' involvement at the time of diagnosis and β -HCG (beta human chorionic gonadotropin) expression is rare.

A 51-year-old Taiwanese male had been suffering from progressive dyspnea for 2 months. A chest X-ray (Fig. 1A) showed a huge heart with a water bottle sign. Cardiac echo and positron emission tomography scan (Fig. 1C–F) showed tumors involving the anterior mediastinum, thoracic cavity, stomach (Fig. 1E, arrow), and bilateral adrenal glands (Fig. 1F, arrow). Serology tests showed an increased level of lactate dehydrogenase (387 IU/L), β -HCG (10.02 mIU/mL), CA-125 (298.07 U/mL), tissue polypeptide antigen (386.03 U/mL), but normal β 2-microglobulin (160 μ g/dL) and AFP (2.88 ng/mL). He received computed tomography-guided right adrenal gland biopsy and esophagogastroduodenoscopy for tissue proof instead of anterior mediastinal mass. Both the pathology of the adrenal gland and gastric lesion showed an infiltration of large neoplastic B cells compartmentalized by fine delicate bands of fibrosis, and immunohistochemistry was positive for CD20 and LCA (leukocyte common antigen), and negative for CK and CD3. PMLBCL with multiple organs' (heart, stomach, and bilateral adrenal glands) involvement was diagnosed. The patient accepted chemotherapy with an R-CHOP regimen. After six courses of chemotherapy, complete remission was achieved with disappearance of the mediastinal mass on chest X-ray (Fig. 1B) and normalized tumor markers.

PMLBCL, first described by Lichtenstein and colleagues [1] in 1980, is an uncommon and distinct subtype of diffuse large B-cell lymphoma. The case we reported raises several interesting points. First, increasing serum level of β -HCG, a tumor marker of nonhematological tumors, germ-cell tumors, could detract clinicians from making a diagnosis of primary mediastinal lymphoma. We reviewed the related literatures and found that β -HCG is rarely expressed in lymphomas. Only six cases of lymphomas—two PMLBCLs [2], three adult T cell-type malignant lymphomas [3], and one anaplastic large cell lymphoma [4] with β -HCG expression in immunohistochemistry—have been reported. However, only the anaplastic large cell lymphoma reported a raised serum β -HCG level. This is the first reported case of a PMLBCL with raised serum β -HCG level. Second, although PMLBCL seldom presents with extrathoracic structure involvement at the time of diagnosis [5], this case involved multiple extranodal organs including the heart, stomach, and bilateral adrenal glands at the time of diagnosis. A diagnosis of PMLBCL should be kept in mind when dealing with similar cases where the involvement of multiple extrathoracic and extranodal organs is indicated.

In conclusion, PMLBCL may present with multiple extranodal organs' involvement and increased levels of a nonhematologic tumor marker. Although the expression of nonhematologic tumor marker, β -HCG, is rare in lymphomas, one should summarily not rule out the possibility of PMLBCL. Careful diagnosis is necessary to differentiate germ cell tumors and other metastatic malignancies from PMLBCL.

Conflicts of interest: All authors declare no conflicts of interest.

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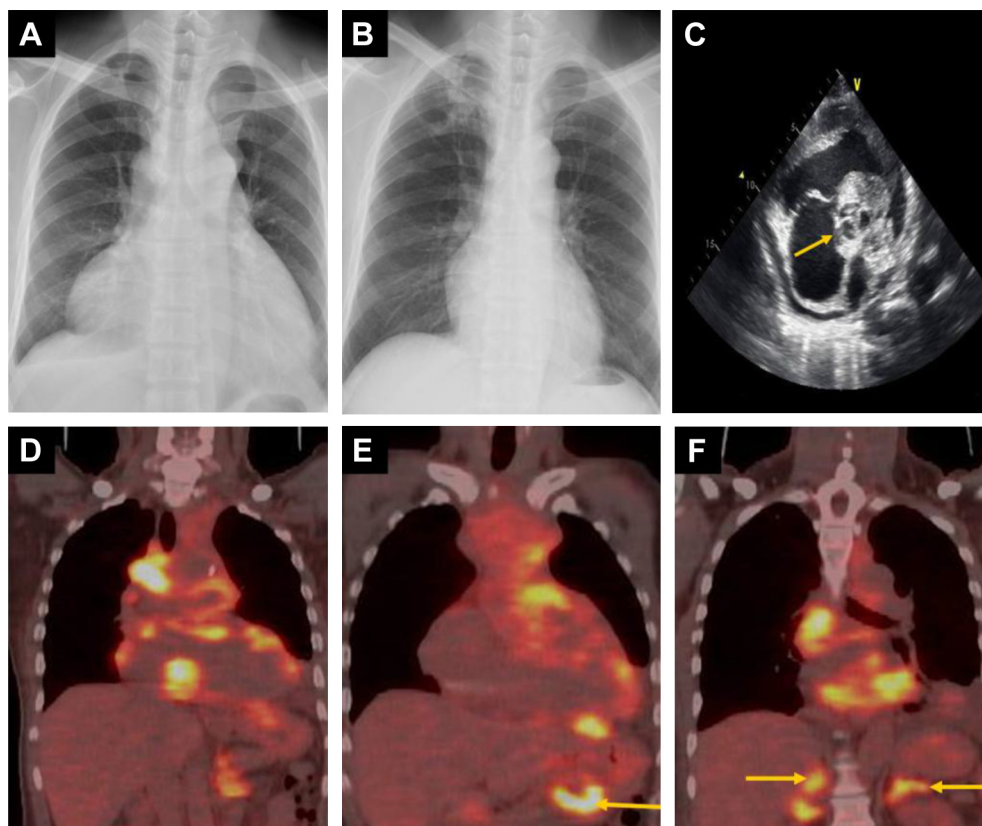


Figure 1. (A) Initial chest X-ray at the time of diagnosis shows a mediastinal mass with water bottle heart sign. (B) Chest X-ray after treatment reveals normal heart size and disappearance of the mediastinal mass. (C) Cardiac echo shows pericardial effusion and a suspected tumor encasing the aorta (arrow). (D–F) Positron emission tomography scan shows lesions over multiple mediastinal lymph nodes, cardiac wall, stomach (E, arrow), and bilateral adrenal glands (F, arrows).

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