Intra-abdominal follicular lymphoma masquerading as severe cardiopulmonary disease in a Vietnam War veteran exposed to agent orange

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DESCRIPTION

A 68-year-old Hispanic male US Army veteran with a medical history of chronic obstructive lung disease, undefined lung scarring, heart failure with preserved ejection fraction, obstructive sleep apnoea, obesity hypoventilation syndrome [on oxygen by nasal cannula 2–4 L, bilevel positive airway pressure (BiPAP) at night] and diabetes mellitus type II, presented to the hospital with dyspnoea and was initiated on treatment for heart failure. The patient had served as an artilleryman in the Vietnam War and had been exposed to Agent Orange—a herbicide used in the US military for defoliation for 10 years prior to discovery of it causing birth defects in laboratory animals. A few studies have not shown that it causes malignancies,¹ but the latest consensus and summary of evidence is that it has been associated with malignancies.²

One month prior to this presentation, the patient was hospitalised for 1 week after experiencing respiratory failure thought to be due to an exacerbation of his heart failure and an upper respiratory tract infection. Within a few weeks after discharge, however, he again started feeling short of breath and gained weight despite doubling his home dose of furosemide as instructed by his heart failure clinic and primary care physician. During this hospitalisation, PaCO₂ was 102 mm Hg. BiPAP did not sufficiently assist and the patient was intubated. CT of the abdomen and pelvis was performed and revealed bulky confluent masses around the pancreatic head, root of mesentery and the abdominal

Figure 2 Massive, tense ascites. On paracentesis, 6.5 L of milky white fluid drained.

aorta measuring approximately 20×18×4 cm with massive ascites (figures 1 and 2).

A paracentesis was performed and 6.5 L of milky white fluid was drained with a triglyceride count of 2265 mg/dL (for comparison, serum triglyceride was only 206 mg/dL), lactate dehydrogenase of 144 units/L, protein of 3.8 g/dL and with cells read as 100% monocytes. Cytology of the chylous ascites was negative for malignancy on two occasions. A subsequent retroperitoneal lymph node core biopsy showed follicular lymphoma grade 1–2. The neoplastic follicles were positive for CD20, CD10, BCL2 and BCL 6. Meshworks of follicular dendritic cells were positive for CD21 and CD23. The neoplasm showed a Ki-67 proliferation fraction of 30%. Flow cytometry was performed on the retroperitoneal biopsy which showed a low cellularity of 35% lymphocytes with a viability of 33%. The lymphocytes analysed were mostly T-cells expressing CD5. Left iliac crest bone marrow biopsy showed lymphoma cells comprising 5% of total marrow cellularity, the involvement of which set the lymphoma at stage IV according to the Ann Arbor staging system. The patient was treated with 5 days of dexamethasone and subsequently started on cycles of rituximab and bendamustine. Over the course of 18 days following peritoneal drain placement, peritoneal fluid drained profusely, and the patient lost 30.2 kg (4.8 stone). The patient was then discharged home with the peritoneal drain, which was removed outpatient.

At the patient's follow-up appointment in clinic, the patient was no longer hypoxic and did not require oxygen at rest nor with activity. He was

Figure 1 Bulky confluent masses around the pancreatic head, root of mesentery, and the abdominal aorta measuring approximately 20×18×4 cm with massive ascites. Cytology ×2 did not show malignancy and diagnosis required a retroperitoneal core biopsy.

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sleeping better, did not critically require diuretics and felt significantly leaner and more robust.

Learning points

- ▶ There is significant evidence that Agent Orange, an herbicide used in the Vietnam War, is associated with non-Hodgkin's lymphoma, Hodgkin disease, chronic lymphocytic leukaemia (including hairy cell leukaemia and other chronic B-cell leukaemias), soft tissue sarcoma and chloracne.
- ▶ Indolent intra-abdominal malignancy can cause cardiopulmonary signs and symptoms and masquerade as other diseases. In patients with risk factors for malignancy and cardiopulmonary signs and symptoms, along with increased abdominal girth, malignancy remains on the differential diagnosis.

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