Lymphoma presenting with spontaneous internal jugular and subclavian venous thrombosis and a large pleural effusion

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Aims

Lymphoma presenting with a simultaneous pleural effusion and thrombosis of the internal jugular and subclavian veins is uncommon. We present the case of a 36-year-old man presenting with a 2-week history of progressive shortness of breath and left-sided chest pain, accompanied by fever and a diffuse swelling over the neck and chest.

A large left-sided pleural effusion on chest X-ray and the clinical presentation led to an initial diagnosis of empyema. However, in view of the unilateral swelling over the supraclavicular region and left side of the chest, a computed tomography (CT) of the chest was performed. This revealed a confluent mediastinal mass abutting the great vessels and a left-sided pleural effusion. Doppler ultrasonography of the left upper limb revealed thrombosis of the internal jugular and subclavian veins. A lymph node (LN) biopsy from the supraclavicular fossa confirmed the diagnosis of a high-grade B-cell non-Hodgkin's lymphoma. The patient was treated with chemo-radiotherapy and remains well and in remission over 18 months since the diagnosis.

Methods

Case report: A 36-year-old man with a background of Crohn's disease (well controlled with 5-aminosalicylic acid) presented with a brief history of worsening shortness of breath and left-sided chest pain to the acute medical unit. He also reported a 2-week history of fever, sweating and swelling over the left side of the neck and had been treated with a course of macrolide antibiotics by his general practitioner, but to no avail.

Examination revealed an unwell and clammy male with a heart rate of 110 beats per minute, temperature of 38.2°C and blood pressure of 177/100 mmHg. His oxygen saturations were 95% on room air. There was a fullness over the left supraclavicular region and over the left side of the chest. He had marked dullness to percussion but no localised tenderness, and absent breath sounds on the left side.

Results

FBC: Haemoglobin 11.5 g/dL, white blood cell 7.6 x 10^9/L, C-reactive protein 92 mg/L, albumin 32 g/L, lactate dehydrogenase (LDH) 459 U/L, alkaline phosphatase 160 U/L. Imaging: ultrasound left upper limb veins revealed an acute thrombus in left internal jugular vein (IJV) and subclavian vein. CT chest abdomen and pelvis demonstrated confluent mediastinal nodal mass with extension into the epicardial fat, right hilum, right paratracheal zone and left supraclavicular fossa suggestive of lymphoma. Left pleural fluid: cytology – lymphoid cells. Total protein 45 g/L, cholesterol 2.0 mmol/L, triglyceride 0.2 mmol/L, LDH 589 IU/L. LN biopsy (Lt SCF): Diffuse large B-cell non-Hodgkin's lymphoma.

Conclusion

Up to 20% of patients with non-Hodgkin's lymphoma and 30% of patients with Hodgkin's lymphoma reportedly have pleural effusions. Pleural effusions in lymphomas are usually exudates and develop by four possible mechanisms: by thoracic duct obstruction by a tumour (chylothorax); by direct pleural involvement of the lymphoma with shedding of cells into the pleural space (malignant); obstructed lymphatic return due to enlarged hilar or mediastinal lymph nodes (paramalignant); and infections (empyema or par pneumonic effusions). Nonetheless, in some cases, especially those with advanced stage low-grade lymphomas with multiple organ involvement, the pleural effusion may be a transudate.

Most lymphoma patients have high serum LDH levels, immunologic defects, numerous immature lymphocytes in peripheral blood, and enlarged hilar or mediastinal lymph nodes. The latter results in limited transportation of fluid from the pleural space back to the veins via the lymphatics.

Our patient had a high protein count (>30) in keeping with an exudate and a high LDH. Triglyceride and cholesterol levels were normal, thereby excluding a chylothorax. The clear straw-coloured fluid was also not macroscopically supportive of the diagnosis. Venous thrombosis was likely secondary to venous stasis caused by extra luminal compression by the lymphoma.

Our patient was treated with monoclonal antibodies against B-cell antigens (rituximab) in combination with cyclophosphamide, doxorubicin, vincristine and prednisone (R-CHOP) followed by radiotherapy, and had an excellent clinical and positron emission tomography (PET) response, and remains well and in full remission. ■

Conflict of interest statement

Authors do not report any conflict of interest.

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