CASE REPORT

A patient with pneumopericardium secondary to extranodal T-cell lymphoma

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SUMMARY

Natural killer (NK)/T-cell lymphomas represent a rare type of lymphoma derived from either activated NK cells or cytotoxic T cells. Most cases with T/NK cell lymphoma present with extranodal manifestations. Primary nodal disease is very rare. The upper respiratory tract is the most common site involved in extranodal disease. Symptoms include epistaxis and nasal obstruction. T/NK cell lymphomas show aggressive and invasive clinical progression, with a poor prognosis. Without treatment, survival is measured in months. We report a case of a 51-year-old male patient with pneumopericardium as a complication of intestinal T/NK cell lymphoma. To the best of our knowledge, this is the first report showing pneumopericardium as a fatal complication of T/NK cell lymphoma. Pneumopericardium should be considered in patients diagnosed with T/NK cell lymphoma presenting with chest pain.

BACKGROUND

This is the first report to show that T/natural killer (NK) cell lymphoma may induce fatal pneumopericardium. Pneumopericardium should be considered in patients diagnosed with T/NK cell lymphoma who present with chest pain.

CASE PRESENTATION

In 2007, a 51-year-old male patient suffering from acute periumbilical pain was admitted to the emergency department. A physical examination revealed involuntary guarding and rebound tenderness. A complete blood count was consistent with polymorphonuclear dominant leucocytosis. The patient was referred to general surgery for consultation and for surgical intervention if necessary. Laparotomy revealed perforation of the jejunum. Segmental jejunal resection and side-to-side anastomosis were performed, and the tissue specimens were sent to the pathology laboratory. Light microscopic examination and immunohistochemical study of the specimens were consistent with T/NK cell lymphoma. A bone marrow biopsy and positron emission tomography (PET)/CT were performed. The results showed neither bone marrow involvement nor extraintestinal tissue involvement. The patient was treated using six cycles of the CHOP protocol consisting of cyclophosphamide, hydroxydaunorubicin (adriamycin), oncovin (vincristine) and prednisolone. His response to the chemotherapy regimen was evaluated with PET/CT, and complete remission was established. After 5 years without any treatment, the patient presented with dyspepsia. An endoscopic examination of his upper gastrointestinal tract

revealed diffuse gastric involvement of T/NK cell lymphoma. The patient was accepted as a case of lymphoma relapse, and he was treated with three cycles of CHOP and three cycles of the DHAP protocol. After the last cycle of DHAP in December 2012, he was hospitalised because of melena and haematemesis. An endoscopic and colonoscopic examination revealed no bleeding. Enteral feeding was stopped, and PPI infusion was started. He became haemodynamically stable following replacements of fluid and blood products. The haematemesis was resolved 2 days after hospitalisation. The patient was being discharged when he developed a cough and a fever of 38.3°C. A chest radiograph revealed a radiolucent halo around the heart compatible with pneumopericardium. A thorax CT was taken which confirmed the diagnosis of pneumopericardium. A pericardial fistula eroding the diaphragm above the stomach secondary to the T/NK cell lymphoma was also observed.

INVESTIGATIONS

The chest radiograph showed a radiolucent halo around the heart compatible with pneumopericardium (figure 1). The thorax CT showed pneumopericardium with left pleural effusion, confirming the diagnosis of pneumopericardium (figure 2).

DIFFERENTIAL DIAGNOSIS

- Post-traumatic pneumopericardium
- Pneumopericardium due to gastropericardial fistula

Figure 1 Chest radiograph showing the pneumopericardium.

To cite: Sener YZ, Babacan T, Sarıca MA, *et al.* *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-201742

Figure 2 Thorax CT showing the pneumopericardium, with left pleural effusion.

TREATMENT

To prevent cardiac tamponade, a pericardial tube was attached by the cardiology department. The patient was referred to the cardiothoracic surgery department for surgical intervention.

OUTCOME AND FOLLOW-UP

The patient died in postoperation follow-up due to septic shock.

DISCUSSION

T/NK cell lymphomas belong to the peripheral T-cell lymphoma family of non-Hodgkin's lymphoma. They are very rare, accounting for just 11.8% of all peripheral T cell lymphomas. T/NK cell lymphomas can be classified as nodal and extranodal, but almost all cases present with extranodal disease. The nose and the upper respiratory tract are the most commonly involved sites. The gastrointestinal tract is the most common site of extranasal T/NK cell lymphoma. The disease frequency is higher in Asia than in Western countries.^{1 2} Epstein-Barr virus (EBV) is

considered a key factor in the pathogenesis of the disease due to virtually all cases containing monoclonal episomal EBV DNA. The disease has a rapid clinical progression without treatment, with survival measured in months; the prognosis is poor even with treatment.² As in this case report, T/NK cell lymphomas can cause tissue wall ruptures, fistulas and perforations because the tumour has a high tendency to grow vertically and invade the tissue walls and vessels. If a patient with a diagnosis of T/NK cell lymphoma presents with abdominal or chest pain, fistula or tissue wall ruptures, and intestinal perforation, pneumothorax and pneumopericardium should be investigated.

Learning points

- ▶ T/natural killer (NK) cell lymphoma may induce pneumopericardium, and this involvement can be fatal.
- ▶ T/NK cell lymphoma shows aggressive and invasive clinical progression, with poor prognosis.
- ▶ T/NK cell lymphoma can rupture the tissue wall and cause fistulas and perforations because of the high tendency of the tumour to grow vertically and invade the tissue walls and vessels.

Contributors All authors were responsible for the conception and design, acquisition of data or analysis and interpretation of data; drafting the article and revising it critically for important intellectual content and final approval of the version published.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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