CASE REPORT

Primary cardiac angiosarcoma: a rare cause of diffuse alveolar haemorrhage

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SUMMARY

Primary cardiac angiosarcoma is a rare disease with a dismal prognosis. We report a case of a 50-year-old man who presented with haemoptysis, cough and worsening dyspnoea. An intracardiac mass was visualised on echocardiogram. He was treated for diffuse alveolar haemorrhage and acute respiratory distress syndrome but died from refractory hypoxaemic respiratory failure leading to cardiac arrest. The diagnosis of primary cardiac angiosarcoma with haemorrhagic pulmonary metastases leading to diffuse alveolar damage was confirmed on autopsy.

BACKGROUND

Diffuse alveolar haemorrhage (DAH) causing rapidly progressive respiratory failure often presents a clinical challenge. When chest imaging demonstrates bilateral airspace opacities, DAH should always be

considered in the differential diagnosis. A myriad of diseases can cause DAH. Our patient presented with a rare cause of fatal DAH: primary cardiac angiosarcoma with pulmonary metastases.

CASE PRESENTATION

A 50-year-old man with a history of hypertension presented with 3 days of dyspnoea on exertion, scant haemoptysis and cough. He denied tobacco use, recreational drug use and recent travel. He took amlodipine daily and worked in a grocery store. He had no significant family history. Chest imaging demonstrated diffuse bilateral ground-glass opacities (figure 1A, B) and he was treated empirically for community-acquired pneumonia with intravenous antibiotics. On hospital day 3, he was admitted to the medical intensive care unit for progressive hypoxaemic respiratory failure that required endotracheal intubation and mechanical ventilation. Endotracheal intubation was complicated by massive haemoptysis. Flexible bronchoscopy was performed and sequential bronchoalveolar lavage samples were consistent with DAH. He developed severe acute respiratory distress syndrome that required prone position ventilation. High-dose intravenous methylprednisolone was started for DAH due to presumed vasculitis. Transthoracic and transoesophageal echocardiography demonstrated a right atrial mass attached to the interatrial septum (figure 2A, B). The patient was deemed too unstable to undergo cardiac surgical biopsy due to severe hypoxaemia. Bloody secretions were present in the endotracheal tube. Coarse crackles were present bilaterally with diminished breath sounds at the bilateral bases. Cardiac sounds were diminished with no murmur. He was sedated and paralysed on mechanical ventilation.

INVESTIGATIONS

Haemoglobin was 10.5 g/dL, the white blood cell count was $5.8 \times 10^9/L$ and the platelet count was $156 \times 10^9/L$. Prothrombin and activated partial thromboplastin times were within normal limits. Sputum and blood cultures were negative. An extensive work-up for infections, vasculitis and rheumatological diseases was negative.

DIFFERENTIAL DIAGNOSIS

- ▶ Bacterial pneumonia.
- ▶ Hantavirus pulmonary syndrome.
- ▶ Vasculitis.

Figures 1 (A, B) Chest X-ray on admission—bilateral fluffy peripheral lung opacities. Chest CT shows bilateral peripheral lung consolidative and ground-glass alveolar opacities.

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Figures 2 (A, B) Apical four chamber view of transthoracic echocardiogram shows right atrial mass attached to the interatrial septum (arrow). Bicausal view of transoesophageal echocardiogram shows tumour in the right atrium attached to the interatrial septum and extending into the superior vena cava (arrow).

TREATMENT

Despite treatment with empiric antibiotics and high-dose intravenous steroids, the patient died on hospital day 28 from refractory hypoxaemia and cardiac arrest.

OUTCOME AND FOLLOW-UP

Postmortem examination demonstrated a 2 cm × 2 cm × 1 cm poorly differentiated cardiac angiosarcoma in the right atrium with infiltration into the myocardium and endocardium with areas of necrosis and thrombosis. Microscopic examination of his lungs demonstrated extensive multifocal infiltration of metastatic angiosarcoma positive for CD34 with evidence of diffuse alveolar damage (figure 3A–C). Haemorrhagic pericardial and bilateral pleural effusions were also noted. Both adrenal glands had evidence of metastatic angiosarcoma.

DISCUSSION

Common pulmonary diseases can often present with atypical symptoms. DAH is characterised by diffuse blood leakage from the pulmonary microcirculation into the alveolar spaces due to microvascular damage. It is a potentially life-threatening condition that can have numerous causes, including autoimmune diseases, infections, drug toxicities, coagulopathies, malignancies and cardiovascular disorders.¹

Our patient presented with a rare cause of DAH: primary cardiac angiosarcoma with pulmonary metastases. Angiosarcoma is the most common primary cardiac malignancy accounting

Figures 3 (A–C) Low-power view of tumour mass infiltrating in lung parenchyma. Areas of diffuse alveolar damage with hyaline membranes. Tumour cells under high power show hyperchromatic nucleus and prominent nucleoli.

for 33% of all primary cardiac tumours.² The symptoms are commonly unspecific and may range from arrhythmia to right-sided heart failure due to pericardial tamponade.

To date, less than 200 cases of primary cardiac angiosarcoma have been reported, with the lungs being the most common metastatic site. In a fraction of these cases, patients initially present with pulmonary symptoms such as dyspnoea and cough. Several

Patient's perspective

'I expect a short hospital stay for pneumonia and am eager to return to work soon'.

Overall, we were unable to obtain the direct patient perspective as the patient died during the hospitalisation.

Learning points

- ▶ Diffuse alveolar haemorrhage (DAH) should be considered in the differential diagnosis of patients with new bilateral pulmonary opacities on chest imaging.
- ▶ Primary cardiac angiosarcoma may rarely present with fatal DAH due to pulmonary metastases.
- ▶ Diagnosis is often made in retrospect on autopsy due to difficulty to obtain antemortem pathological specimen in these patients.

patients have initially presented with haemoptysis and a few reports of DAH as the initial presentation have been published. The prognosis is extremely poor with a median survival time of 3.8 ± 2.5 months without surgical resection.³ Due to the rarity, studies to establish the optimal treatment of cardiac angiosarcomas are scarce. Surgery is the treatment of choice, dependent on the location of the tumour and metastases. Although total resection is the treatment of choice, tumour recurrence is very common. Chemotherapy and radiation may be used as adjuvant therapy in selected cases.^{4 5}

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