Rare disease

Pleomorphic rhabdomyosarcoma with pulmonary tumour embolism

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Summary

Pulmonary tumour embolism is a known complication of cancer disease. To date, pleomorphic rhabdomyosarcoma has been described once with this entity. We report a case of pulmonary tumour embolism diagnosed in the operation room after cardiac arrest of a 30-year-old male patient who had surgical amputation of his right upper limb due to recurrent sarcoma, mandating urgent and successful embolectomy.

BACKGROUND

This case represents a complicated presentation of pulmonary tumour embolism. The patient had a sudden cardiac arrest soon after shoulder ectomy for sarcoma of the right upper limb. The patient had an aggressive resuscitation and successful embolectomy, followed by a challenging hospital course managed by a multidisciplinary team. We report this case to highlight the diagnostic and therapeutic interventions for this rare complication.

CASE PRESENTATION

A 30-year-old male patient was admitted to the orthopaedics ward for amputation of the right shoulder due to extensive gangrene of the upper limb secondary to recurrent rhabdomyosarcoma. He was complaining of numbness, pain and weakness of the right forearm. Review of other systems was unremarkable. The preoperative assessments, including arterial blood gas, ECG and chest x-ray, were normal.

He had amputation of the right shoulder. Soon after surgery, the end-tidal CO₂ trace was not detected, with desaturation down to the lower 80s; he went into sudden cardiac arrest with pulseless electrical activity.

Cardiopulmonary resuscitation was started as per advance cardiac life-support guidelines for around 20 min, and a right side chest tube was inserted empirically for possible pneumothorax or haemothorax until the pulse became palpable and the mean arterial pressure (MAP) was 100 mm Hg.

INVESTIGATIONS

After stabilising the haemodynamics status, a chest x-ray was performed to confirm the position of the endotracheal tube and to rule out intrathoracic abnormality (Figures 1 and 2). After 15 min, another cardiac arrest happened and suspicion of pulmonary embolism was high. Thus, a transesophageal echo (TEE) was performed after stabilising him. The TEE showed right side atrial and ventricular enlargement, severe tricuspid

regurgitation and a large bilobed mobile mass in the right pulmonary artery, with significant proximal narrowing and small pericardial effusion without signs of tamponade (figures 3 and 4).

TREATMENT

The cardiothoracic surgery team was immediately involved. Sternotomy and pulmonary embolectomy were performed successfully (figures 5 and 6). During the surgery, the patient had another cardiac arrest, and was given an intrathoracic massage. Intravenous lidocaine and defibrillation were given for ventricular fibrillation, and after 10 min, he regained sinus rhythm.

Postoperatively, the patient was transferred to the surgical intensive care unit (ICU). He was intubated and mechanically ventilated. Haemodynamically, he was unstable on multiple vasopressors including dopamine, norepinephrine and phenylephrine. Fluid resuscitation was started to achieve a MAP of 65 mm Hg. The initial central venous pressure (CVP) reading was 18, and fluid boluses were adjusted as per the CVP reading. Dobutamine infusion was started due to low mixed venous oxygen saturation of 55%. Broad-spectrum antibiotics including tazocin and vancomycin were given empirically.

Secondary to refractory, hypotension stress dose hydrocortisone was initiated. Laboratory results showed a pH of 7.10, with a high anion gap secondary to lactic acidosis and acute kidney injury. Urgent continuous veno-venous haemofiltration and bicarbonate infusion were initiated because of severe acidosis.

OUTCOME AND FOLLOW-UP

On the second day, the patient started showing signs of responsiveness with significant clinical improvement and less oxygen requirement, less inotropic support and satisfactory neurological assessment after weaning sedation. His course in the ICU was complicated by recurrent transudative pleural effusion on the right side which was drained by means of a pleural pig-tail catheter. Owing to

Figure 1 Preoperational chest x-ray.

prolonged mechanical ventilation, a percutaneous tracheostomy was performed. Haemodialysis was stopped after recovery of his renal function. After 6 weeks, the patient was discharged from the ICU. He underwent a psychological and physical rehabilitation programme, and was discharged home after 4 weeks. Unfortunately, he was reported deceased after 3 months at home.

DISCUSSION

Pulmonary tumour embolism was first described by Schmidt in 1897 in a young man with primary gastric malignancy.¹ Antemortem diagnosis is rarely recognised, and the autopsy series demonstrates the presence of pulmonary embolisation in 26% of the patients who die of cancer.¹

The neoplastic cells spread to systemic circulation either through invasion of small veins or release of fragments into the neovasculature. Most of the tumour cells become trapped within the pulmonary capillaries.² Many of them can trigger the coagulation cascade, and obstruction of the pulmonary vessels, usually from the presence of both tumour cells and associated clot.³

The most common presentation of pulmonary tumour embolism is subacute dyspnoea. Pleuritic chest pain is

frequently reported, while other symptoms like cough and haemoptysis are rare. Physical examination of these patients usually reveals tachypnoea and tachycardia. Signs of pulmonary hypertension and cor pulmonale may be present.¹

Massive pulmonary tumour embolism rarely occurs as a leading cause of circulatory collapse or imminent death. Few cases have been reported in the literature where urgent embolectomy with the aid of the cardiopulmonary bypass machine without evidence of ischaemic brain injury has been successful.⁴ Efficient and effective cardiopulmonary resuscitation in the operation room, instant help of the cardiothoracic surgeon and aggressive effort in the ICU may all contribute to saving a patient's life.

Pulmonary tumour embolism was reported mostly with mucin-secreting adenocarcinoma originating from the breast, gastric, colon and lung. However, sarcoma and mesothelioma are less frequent causes of this complication.^{5 6}

Imaging studies may support the diagnosis of pulmonary tumour embolism. Chest radiograph and CT are usually unremarkable, but can occasionally show cardiac silhouette and prominent pulmonary arteries. Ventilation-perfusion lung scans may demonstrate multiple small, peripheral, subsegmental perfusion defects. Pulmonary

Figure 2 Postoperational chest x-ray immediately after the first attempt at cardiopulmonary resuscitation. It shows normal lung fields, no pneumothorax or haemothorax. The right chest tube is placed in a good position. Right upper extremity amputation.

Figure 3 Transesophageal echo: The right pulmonary artery (RPA) is obstructed by a mobile bilobed mass (arrow).

angiography is insensitive to tumour emboli; cytological examination of blood aspirated from a wedged pulmonary artery catheter has a high degree of accuracy for the diagnosis of lymphangitic carcinomatosis. However, negative pulmonary wedge cytology does not exclude the diagnosis, and surgical lung biopsy should be pursued in patients in whom a definitive diagnosis is critical.¹

The decision as to whether to pursue aggressively the management of tumour embolism often depends on the nature of the underlying tumour. Confirming the diagnosis of tumour embolism in a patient with a chemotherapy-resistant tumour is thus unlikely to alter the patient's therapy or outcome.⁴

The therapy of tumour embolism is directed at the primary tumour.⁵ Embolectomy and inferior vena cava filter placement have been employed in unusual patients suffering from large, central emboli from infra-diaphragmatic tumours. Chemotherapy does not generally

affect prognosis unless the patient has a very responsive neoplasm. Surgical resection of renal cell carcinoma may lead to a cure for tumour embolism syndrome.⁵ Thrombolysis or anticoagulation is not helpful as pulmonary hypertension is predominantly the result of fibrocellular intimal proliferation.⁷

Tumour emboli do not represent true metastases, but survival after the onset of symptoms is generally limited due to concomitant lymphangitic or metastatic carcinoma. The overall survival time in patients in whom the diagnosis is established antemortem typically is 4–12 weeks.³

Our patient had pleomorphic rhabdomyosarcoma which is resistant to chemotherapy and radiotherapy complicated massive pulmonary tumour embolism and successful embolectomy. He was discharged after a rehabilitation course in the hospital; however, the prognosis is very poor.

Figure 4 Transesophageal echo: The right atrium (RA) and right ventricle (RV) are significantly dilated with severe tricuspid regurgitation (TR) and small pericardial effusion.

Figure 5 Gross appearance of tumour embolus after removal from the right pulmonary artery.

Figure 6 Microscopic appearance of the malignant tumour embolus; as cellular spindle cell proliferation arranged in fascicles. The tumour cells show mild-to-moderate pleomorphism in this picture (H&E stain, magnification $\times 20$).

Learning points

- ▶ Pulmonary vascular occlusion by malignant cells should be considered in cancer patients when they present with acute dyspnoea, pulmonary hypertension, cor pulmonale or sudden cardiopulmonary arrest.
- ▶ Despite difficulty of diagnosis, clinical suspicion should be high.
- ▶ Loss of end-tidal CO₂ trace for intubated patients indicates either endotracheal tube dislodgment or pulmonary embolism.
- ▶ Treatment should be focused on the underlying malignancy; however, surgical embolectomy may be the choice.

Competing interests None.

Patient consent Obtained.

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