Idiopathic pulmonary fibrosis with complication of severe respiratory failure, right heart failure, and steroid induced diabetes – qualification for lung transplantation as a matter of urgency

Idiopatyczne włóknienie płuc powikłane ciężką niewydolnością oddechową, prawokomorową niewydolnością serca i jatrogenną posteroidową cukrzycą – kwalifikacja do przeszczepu płuc w trybie przyspieszonym

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Słowa kluczowe: nadciśnienie płucne, idiopatyczne włóknienie płuc, przeszczep płuc.

Abstract

A 62-year-old woman 146 cm tall and weighing 50 kg, due to idiopathic pulmonary fibrosis, was firstly approved for lung transplantation according to the planned mode. Due to the low height of the patient there were difficulties in the selection of the donor and prolonged waiting time for the surgery. Rapid progression of pulmonary hypertension and steroid-induced diabetes forced us to change the mode of the procedure to urgent. The description of the case shows the difficulties in therapy and choosing the appropriate time for lung transplantation. In this case, the collaboration of specialists from various fields in the decision on transplant is noteworthy.

Streszczenie

Kobieta 62-letnia o wzroście 146 cm i masie ciała 50 kg została początkowo zakwalifikowana do przeszczepu płuc w trybie planowym z powodu idiopatycznego włóknienia płuc. Ze względu na niski wzrost chorej wystąpiły trudności w doborze dawcy i przedłużenie czasu oczekiwania na zabieg. Gwałtowna progresja nadciśnienia płucnego i wystąpienie cukrzycy posteroidowej wymusiły zmianę trybu zabiegu na pilny. Opis przypadku ilustruje trudności w terapii i wybrania odpowiedniego momentu do wykonania przeszczepu płuc. W omawianym przypadku na uwagę zasługuje fakt dobrej współpracy specjalistów z różnych dziedzin w podjęciu decyzji o transplantacji (pulmonologa i kardiologa).

Introduction

Idiopathic pulmonary fibrosis is one of the forms of interstitial pneumonia limited to that organ. The aetiology of this disease is not known. The most probable risk factors to take into account are: tobacco smoking, gastrointestinal disorders, and exposure to wood, dust, and metals. Treatment is difficult and there is no proven advantage of any pharmacological regimen over others. None of the drugs used to date has changed the course of idiopathic pulmonary fibrosis.

Due to the progressive nature of the disease treatment trials are often used with prednisone of 0.5 mg/kg or other glucocorticoids at equivalent dose, with the simultaneous application of cyclophosphamide or azathioprine. The prognosis for this patient group is poor. Survival after diagnosis is on average from 3 to 5 years. Up to 15% of patients develop lung cancer. If the therapy is ineffective some of the patients will be eligible for lung transplantation [1, 2].

In recent years, treatment performed at the Centre in Zabrze (Poland) for patients of lung transplant has resulted in a prognosis of survival comparable to the

best centres in the world. The problem is the shortage of donors, especially for persons who are below 150 cm tall. After obtaining a donor the problem is the need for early arrival to the place of surgery of the patient who is first on the list for lung transplant.

Given the fact that only two centres in Poland are performing such treatments the most comfortable situation is that persons qualified for lung transplant are living at a distance of 100 km or less from the resort. This often requires the help of family and friends and periodic change of residence, but this is one of the factors that facilitate the implementation of transplantation [3].

With the exception of lung transplantation there is no other effective treatment for idiopathic pulmonary fibrosis, and drug therapy can only postpone the moment of transplantation. The worst problems are complications after steroids treatment, including diabetes iatrogenic. Another problem is pulmonary hypertension, which constitutes an obstacle to lung transplantation [4]. In some cases, simultaneous grafting of both the lungs and heart is also required, and the results of such an operation are worse than just the lungs. For the clinician, also a pulmonologist, it is very difficult to assess the qualification mode for lung transplant surgery.

The cardiologist is helpful in such cases, and imaging tests like echocardiography, magnetic resonance imaging of the heart, and cardio pulmonary exercise testing (CPET) can accelerate lung transplant qualification also due to cardiac complications. In case of very rapid progression of pulmonary hypertension the decision to accelerate qualification for lung transplantation in patients with idiopathic pulmonary fibrosis determines the successful outcome [2, 5].

Case report

A 62-year-old patient with idiopathic pulmonary fibrosis with concomitant asthma and steroid-induced diabetes (SID) was admitted to the Department of Internal Disease with severe general condition due to dyspnoea escalating for about a week accompanied by productive cough and subfebrile temperature.

The patient had been hospitalised many times due to similar symptoms, and in the past 6 months it was her fourth stay at the hospital. So far she had accepted methylprednisolone at a dose of 4 mg/day and formoterol inhalation 2 × 1, 4 × 2 breaths ipratropium bromide, and budesonide inhalation 4 × 1. On admission to the Department, in the examination, auscultation over the lung, single quiet crackling was found mainly at the base on both sides. The chest radiograph showed intensification of the stroma of both lungs with numerous visible scattered small maculate changes; the X-ray image was similar to that seen in a study conducted in April 2014.

In laboratory tests, elevated levels of markers of inflammation were encountered (white blood cell count (WBC) 13.00 thousand/l). Enabled oxygen therapy, broad-spectrum antibiotics (piperacillin with tazobactam), and bronchodilator drugs intensively nebulising, gave only a small reduction in dyspnoea and normalisation of inflammatory parameters (WBC 7.72 thousand/l).

Due to the fact that the persistent dyspnoea cleared, despite infection, it was decided to perform echocardiography. The echocardiogram showed marginally thicker muscle of the left ventricle, widening of the right ventricle, mildly increased right atrial cavity, mild tricuspid regurgitation, and features of pulmonary arterial hypertension (RSVP estimated 103 mm Hg, shortened AcT 54 ms). Features of overload of the right ventricle were not present in the previous study carried out three months earlier (AcT 135 ms, VCI 1.4/0.8 cm) [6, 7].

Due to observed progression of the underlying disease the patient consulted with a specialist in cardiology and clinical transplantation and it was decided that the patient qualified for urgent lung transplant (she was previously listed for transplant to non-urgent scheduled surgery). A doctor of the Clinical Department of Silesian Medical University was contacted, and the deadline, on 16.07.2014, for the patient hospitalisation was re-established. In accordance with the recommendations sildenafil was included in the treatment at a dose of 2 × 25 mg [7, 8] and methylprednisolone was discontinued. It was agreed that the patient would visit the Clinic for oxygen therapy in order to qualify for home oxygen therapy and to secure the patient's travel from Warsaw to Zabrze. The patient is going to live with a family in Sosnowiec while waiting for treatment.

Conclusions

In the case described above, the planned pre-qualification transplant of lungs performed by a pulmonologist, towards extremely rapid progression of right ventricular heart failure after cardiac consultation, has been converted into urgent mode.

The reversibility of this process determines the possibility of making the lung transplant without simultaneous transplantation of the heart, which would have a worse prognosis for the patient.

Steroid-induced diabetes required discontinuation of methylprednisolone and the use of sildenafil at a dose of 2 × 25 mg. After contact with the Centre for Transplantation in Zabrze, a time limit has been assigned for completion of specialised research in the hospital within 2 weeks.

In this case, the collaboration of specialists from various fields is noteworthy in the decision about the transplant. The difficulty was the low growth of the

patient and the distance of her place of residence to the Transplantation Centre.

Conflict of interest

The authors declare no conflict of interest.

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