Medical Expertise

"Development of the European Network in Orphan Cardiovascular Diseases"
„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych"

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CASE SUMMARY

The article "Idiopathic pulmonary artery hypertension" constitutes a very interesting case regarding unusual clinical presentation and course of patient with PAH. There are some critical points for the final outcome. The first is related to familiar character of PAH – son of the presented patient died for PAH. There was no data on screening for the disease in the family. Active screening should always be performed.

Unfortunately, Authors had local / national limitations in reimbursement of specific treatment for PAH that obviously had impact on further follow-up of the disease.

The next important point was lung disease coexisting with PAH. Patient suffered from chronic lung disease – on chest CT emphysema, bronchiectases and fibrotic changes were observed, however, any final diagnosis was made on this step of the disease. It could be both COPD and tuberculosis, especially, that in the explanted lungs caseating granulomas (active tuberculosis) were find (see slides). Thus, we should never forget about tuberculosis as a latent or overt cause of lung problems.

The next steps of patient's treatment – heart and lung transplantation, control biopsies and tuberculosis pharmacotherapy were done perfectly. Regardless of all efforts the clinical deterioration was observed as a result of chronic lung rejection – bronchiolotis obliterans syndrome. Finally, the cause of death was myocardial infarction due tobliterating coronary

vasculopathy. Probably, immunosuppressant therapy was a reason for rapid progression of coronary after transplantation.

DISCUSSION

Post-lung transplant survival ranges from 59%-93% at 1-year. Data on heart-lung transplant are limited (130 cases in Pittsburgh registry).

Cardiac allograft vasculopathy (CAV) is an important cause of morbidity and mortality after heart transplantation. CAV occurs in approximately 30% of patients by 5 years. Early detection of CAV may allow alterations in medical therapy before progression to the stage that revascularization is required. Thus, a routine screening for CAV in transplant recipients is sufficient, traditionally by invasive coronary angiography (ICA). Recent advances in invasive (intravascular ultrasound) and non-invasive imaging technology (MSCT) also permit detection of subangiographic CAV.

The treatment options for established CAV are limited; coronary revascularization is available only for some patients because of the diffuse character of CAV. According to some authors retransplantation is the only definitive therapy for CAV. The most promising development in the recent years is the use of mTOR inhibitors, which are regarded as effective in preventing CAV in de novo patients; their role in the treatment of established CAV is still uncertain.

EXPERT'S OPINION

In my opinion, the revised study is of high value and should be published in JRCD.

Minor concern – I suggest some change of the title to be more informative, i.e. "Idiopathic pulmonary arterial hypertension with subsequent complications of implemented therapy"

CONCLUSION

The revised manuscript presents real-life case of PAH with coexisting lung problems requiring differential diagnosis and the data on clinical complications of heart and lung transplantation with subsequent immunosuppressant therapy. It is of high value and suitable for publication.

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