11–12 October 2017

ORAL PRESENTATIONS

PREGNANCY AND RHEUMATIC DISEASE

1. EARLY SCLERODERMA OVERLAP SYNDROME (PULMONARY HYPERTENSION, ILD, POLYMYOSITIS AND INFLAMMATORY ARTHRITIS) AND AN UNPLANNED PREGNANCY

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Introduction: Scleroderma is associated with high risk of poor maternal and fetal outcome in pregnancy and requires careful pre-conception planning, close monitoring in a specialist multi-disciplinary team and appropriate targeted therapy. We present a case of limited cutaneous systemic sclerosis with associated polymyositis and inflammatory arthritis overlap syndrome, during her first pregnancy before disease control had been obtained, managed in a tertiary obstetric medicine department. The case is followed by a discussion of the important issues to consider when counselling scleroderma patients about pregnancy.

Case description: A 32 year old lady was referred to the obstetric medicine clinic for antenatal care. She had a recent diagnosis of limited cutaneous systemic sclerosis with overlap polymyositis and inflammatory arthritis. She had initially presented one year previously with joint pain, symptoms of carpal tunnel syndrome and Raynaud's phenomenon, and was found to have sclerodactyly, myositis, mild interstitial lung disease and borderline raised pulmonary pressures. Blood tests had revealed normal full blood count, kidney, liver and thyroid function, with normal inflammatory markers. ANA was strongly positive (titre 1:640, nucleolar fine speckled staining pattern). ENA was negative, but extended myositis panel revealed a positive anti-PM-Scl antibody. Creatine kinase was raised at 755IU/L. An EMG showed mild bilateral carpal tunnel syndrome, but no evidence of neurophysiological evidence of myositis. A high resolution CT scan of the thorax had demonstrated some suggestion of early NSIP pattern (peripheral ground glass changes) with prominent pulmonary arteries. Pulmonary function tests showed preserved FVC/TLC but reduced gas transfer (KCO c 1.16, 66% predicted and TLCOc 5.3, 52.2% predicted). Echo estimated the pulmonary artery pressure to be 25 to 30 mmHg, and demonstrated a dilated right ventricle. Treatment with pulsed steroids and Hydroxychloroquine had only been partially effective. The patient was seen in the obstetric medicine clinic at 22 weeks gestation, of an unplanned pregnancy. She reported breathlessness walking up two flights of stairs. Clinical examination revealed normal heart sounds with a loud P2 and an ejection systolic murmur. Blood pressure was 94/66 mmHg. Heart rate was 80bpm. Marked facial skin tightening and sclerodactyly was noted, with synovitis of the proximal interphalangeal joints and flexion contractures in the hand with sclerodactyly. There was no finger ulceration. Auscultation of lungs was unremarkable. Urinalysis showed trace of protein and one plus of leucocytes. There was insufficient TR to assess PASP on the repeat echocardiogram, but the RV was dilated, although assessment of RV function was not possible. Booking bloods were unremarkable. CK had normalised to 50 IU/L. Inflammatory markers (ESR, CRP) and immunoglobulin levels were normal. 12 and 20 week fetal scans were normal. TPMT level was normal. Non-invasive prenatal screening revealed a low PAPP-A result, putting her at increased risk of prematurity, growth restriction and pre-eclampsia. A detailed ultrasound in the fetal medicine department at 24weeks gestation was reassuring with normal growth scan and Doppler studies, and serial growth scans were arranged. She was started on high dose oral steroids (Prednisolone 30mg OD), Azathioprine 100mg OD and Omeprazole. The patient was seen every fortnight in the obstetric medicine clinic. She tolerated Azathioprine and had no progression of her scleroderma. Steroid doses were successfully weaned. At time of case submission, she was 28 weeks pregnant.

Discussion: In early pregnancy, several features predicted poor outcomes, including untreated and active scleroderma, multi-organ involvement, an unplanned pregnancy and low PAPP-A results. However, this patient was fortunate as the pulmonary hypertension was only borderline, the interstitial lung disease was very limited and the arthritis and the polymyositis mild, with excellent response to treatment. At diagnosis this patient declined treatment with DMARDS. Therefore at conception, the connective tissue disease was active and only partially treated, with evidence of organ-based complications. However, when there as evidence of on-going disease activity in early pregnancy, there was a reluctance to treat aggressively in pregnancy. This is a common mistake as active disease leads to poor pregnancy outcomes and irreversible progression of disease.

Key learning points: Pre-conception counselling for scleroderma patients is best done within a multidisciplinary team. Pregnancy should be planned once disease control has been achieved without teratogenic medications. A full assessment of organ complications should be made, with particular attention on pulmonary hypertension, which is associated with significant risk of morbidity and mortality. Pregnancies in such patients are high risk, and should be managed throughout pregnancy and the peripartum period in a high-risk obstetric monitoring programme. Patients should be monitored closely for the life-threatening complications of scleroderma renal crisis and pulmonary hypertension. Pre-eclampsia does not appear to be more common in scleroderma but it is difficult to differentiate hypertensive complications (including HELLP). Pregnancy is often complicated by pre-term birth and small for gestational age infants. Non-invasive prenatal screening tests may be useful in identifying those at risk.