2. PREGNANCY AND MYOSITIS

Nibha Jain[1], Shilpa Jagadeesh[1], and Arumugam Moorthy[1]
[1]Rheumatology, University Hospital Leicester, Leicester, United Kingdom

Introduction: Inflammatory disorders can appear as a spectrum and pose diagnostic challenges. Inflammatory myositis affects women of childbearing age. This situation presents challenges in management of disease during pregnancy. Myositis specific antibodies are expected to lead to certain clinical presentation but cases outside known information always occur as we learn from this case report.
Case description: A 26-year-old Asian lady was referred to rheumatology for inflammatory arthritis and Raynaud's on a background of scalp psoriasis and family history of psoriatic arthritis. She had hip pain with no spinal inflammation and was commenced on low dose methotrexate. ANA 1:6400 but autoimmune screen negative. She was intolerant to higher doses of methotrexate. She developed severe hip pain which was not septic and eventually developed reduced hip movements and forward flexion of spine. X-ray showed soft tissue calcification around hip methotrexate was stopped due to respiratory symptoms but HRCT and pulmonary function were normal. Unfortunately, she was lost to follow-up.
She was then referred after a year with skin tightening (indurated plaque) over loin and chest. CK was elevated without clinical evidence of muscle weakness and ANA 1:1600 positive with negative ENA and normal DsDNA/Compliments. Dermatologists felt that indurated plaque was morphea as the histology was inconclusive. She developed pelvic girdle weakness along with left hip calcification and progression of skin tightening of fingers and forearm. Although biopsy and MRI thigh were negative for myositis, nerve conduction studies showed severe active polymyositis. Her extended myositis panel now showed Mi-2 antibody.
As she was intolerant to azathioprine with a progressive illness and was keen to have children soon, she was started on IvIG. Despite being on IvIg she developed refractory calcium discharging sinus over her hip. Rituximab was not licensed for myositis then and she was not keen to start on any other medications recommended by the myositis specialist centre. She had a successful pregnancy after 6 months of disease control under joint care of maternal foetal medicine and rheumatology. On repeat autoimmune testing prior to pregnancy, anti-Ro was equivocal (previously negative) hence antepartum surveillance was carried out. The baby had no evidence of congenital heart block.
She has progressive extrarticular calcification with otherwise well controlled disease and prefers to remain on IvIg.
Discussion: We present the first case of psoriasis with Raynaud's developing progressive skin and soft calcification resulting in discharging sinuses. She developed myositis scleroderma overlap with suspected cardiac involvement posing challenge due to intended pregnancy. There was limited data to go by on outcomes of pregnancy in dermatomyositis and no there are similar cases in literature.
In retrospect, her joint symptoms could have stemmed from extra-articular calcification around hip but does not explain skin tightening around fingers. It makes one wonder if resistant scalp psoriasis initially could be related to dermatomyositis and not true psoriasis. She was managed with regular advice from the myositis specialist unit and declined to go on any drugs which could have an impact on fertility or pregnancy. Hence options for treatment were limited and complicated by intolerance to conventional DMARDS. IvIG was selected based on those preferences due to progressive myositis but there were initial reactions to IvIG infusions at which point use of rituximab was considered. The NICE rituximab in myositis guidelines were not present at that time and the individual funding request was declined.
Myositis is an idiopathic inflammatory immune mediated disorder that may be existent in an isolated form or in combination with other autoimmune or connective tissue disorders. It is a T-cell mediated cytotoxic process directed toward unknown muscle antigens. Psoriasis on the other hand is a relapsing skin disease; the diagnosis is of which is made on clinical grounds and can be associated with SpA.

In a retrospective review of psoriasis patients seen at the Mayo Clinic the frequency of pathologically confirmed myopathies or inflammation in muscle in patients with psoriasis was estimated to be 0.13%. However, this could be an overestimate, given potential referral bias. Concomitant autoimmune disorders, psoriatic arthritis, and exposure to anti-TNF-α therapy were the proposed associations with increased risk of developing myopathy in psoriasis patients. Most had inclusion of body myositis.

Key learning points: Evidence suggests that the appropriate treatment with immunosuppressants allows a normal pregnancy without major problems and with no further risk for post-partum relapse. This is presuming the disease is well-controlled for 6 months prior to conception.

There is no definite impact of pregnancy on a well-controlled myositis, although case reports have variable outcomes.

Pregnancy outcomes are better if the disease is fully controlled preconception and there is no cardiac or respiratory involvement. Hence, preconception work up is done in liaison with maternal foetal medicine and includes disease activity measurements, repeat investigations for systemic involvement (commonly ECHO and pulmonary function test), repeat autoimmune screen and individualised preconception counselling.

Poorly controlled disease can increase risk of intrauterine growth retardation, stillbirth or preterm birth. Uncontrolled inflammation is thought to result in poor placental circulation due to inflammatory fibrillin deposition.

Autoimmune disorders are conventionally known to flare postpartum but experiences are variable. We observed a slight CK rise postpartum which settled without needing further treatment.

Treatment options available for women considering pregnancy include glucocorticoids and intravenous immunoglobulin for induction of remission and remission with azathioprine, cyclosporin or tacrolimus.

Data available is from case series and experiences of specialist centres only, hence there is scope for further research.

Owing to limited data on the long-term use of intravenous immunoglobulins in myositis, absence of evidence-based treatment options for calcification in myositis and push for switch to rituximab due to cost implications, further management of patients in similar situations will be challenging.

Conflicts of interest: The authors have declared no conflicts of interest.