Familial Mediterranean fever in pregnancy: A case study

Dr Evelyn Smith Romero, Dr Aye Htun

Obstetrics and Gynecology, Canterbury Hospital, Sydney Local Health District, NSW

Background

Familial Mediterranean fever (FMF) is an autosomal recessive, inherited, auto-inflammatory disorder. FMF presents as sporadic, self-limiting and recurrent episodes of fevers and serosal inflammation and can progress to amyloidosis and nephropathy.

The acute bouts present as several days of pyrexia with severe chest, joint and/or abdominal pain. Episodes may be triggered by emotional or physical stress, menstruation or surgery and may worsen in pregnancy.

Most individuals experience their first attack before or during the second decade of life, thus affects women of reproductive age [1].

Case

A 29-year-old female was diagnosed with FMF at 7 years of age and had been stable on colchicine for several years. She had a spontaneous and planned pregnancy. Though remaining on colchicine, her FMF flares became more frequent antenatally; including requiring an admission to hospital for severe abdominal pain. However, her renal function remained normal, as did the foetal wellbeing.

Her child was delivered via an elective caesarean section. She opted for this delivery mode as she was concerned that the emotional distress of labour would trigger painful attacks. She delivered a live infant at term in good condition with an uncomplicated operation and puerperium.

Tel Hashommer Diagnostic Criteria for Familial Mediterranean Fever [12]

Major Criteria	Minor Criteria
Recurrent febrile episodes with serositis (peritonitis, synovitis or pleuritis)	Recurrent febrile episodes
Amyloidosis of type AA without a predisposing disease	Erysipelas-like erythema
Favorable response to regular colchicine treatment	FMF in a first-degree relative
Diagnosis of FMF can be made if 1) two or more major symptoms or 2) one major plus two minor symptoms are present.	

Discussion

FMF is a rare disease that affects women in their reproductive years. The natural history of FMF in pregnancy is shown to worsen in a third of pregnancies, improve in a third and remain unchanged in a third.

Attacks in pregnancy can lead to peritonitis and eventual miscarriage or preterm birth [2]. Women with renal amyloidosis may experience a deterioration of renal function during pregnancy, as well as abortion and stillbirth [3, 4].

Standard treatment is oral colchicine to prevent attacks and the progression to amyloidosis. Colchicine is a category D drug, due its properties as a mitotic inhibitor and is permeable through the blood-placenta barrier.

Large observational studies have found that long term colchicine use in pregnancy does not increase the risk of cytogenetic or congenital pathology, nor of preterm labour [1, 5-7]. Prior to introduction of colchicine, the rate of abortions and miscarriages was 25-30% higher in women with FMF [8, 9].

Colchicine should be continued in pregnancy, as its therapeutic benefit outweighs the risks to the foetus. Regular colchicine use should not preclude breastfeeding for informed women [10, 11].

Individuals with FMF may have identifiable triggers for attacks including emotional or physical stress. Therefore, mode and timing of delivery should be patient's decision with multi-disciplinary support.

Pre-conception planning is recommended to gage severity of the disease and attack frequency, obstetric and family history, as well as renal function.

Conclusions

- ❑ FMF is a rare rheumatological, inherited disease that presents a challenge to antenatal healthcare providers.
- ❑ Women require close surveillance and multi-disciplinary input.
- ❑ The decision on mode and timing of delivery should be individualized and determined with regards to disease progression and patient needs.
- ❑ Colchicine has shown to be safe in pregnancy and breastfeeding and reduces risks of adverse pregnancy outcomes.

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