ULTRASOUND DIAGNOSIS OF FETAL ACRANIA AT 12+6 WEEKS GESTATION WITH A BACKGROUND OF SJOGREN'S SYNDROME

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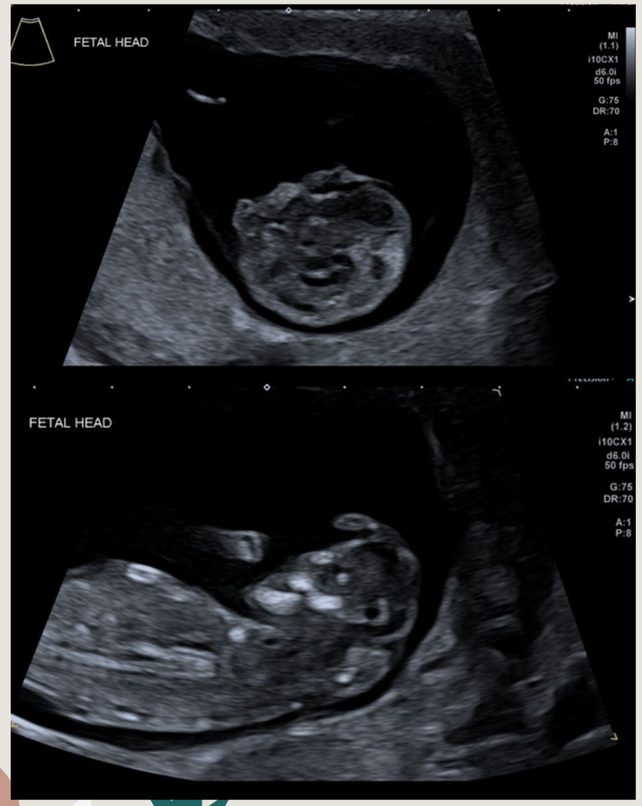

INTRODUCTION

Fetal acrania is a life-limiting malformation in which there is a complete or partial absence of the cranium. It can be diagnosed on ultrasound at 11-13 weeks when the skull is expected to be ossified.

AIM

The aim of this case report is to emphasise the importance of early anatomy as well as a review of the current understanding of causes of fetal acrania and if there is any association with primary Sjogren's Syndrome (pSS).

CASE

This is a 37-year-old gravida 3 para 2 female with unmedicated pSS who presented with an outpatient ultrasound demonstrating a single intrauterine pregnancy demonstrating acrania and distorted brain hemispheres. She had two previous normal vaginal births (1 and 3 years old).

RESULTS

She had a repeat ultrasound which confirmed no cranium with a distorted fetal brain. She proceeded to a medical termination which was complicated by retained products. She underwent an ultrasound guided dilatation and curettage which was uncomplicated. Histopathology confirmed retained products of conception and cord microarray did not reveal a cause.

DISCUSSION

The cause for fetal acrania is not currently well understood. Some factors associated with neural tube defects (NTDs) include hyperthermia, antiepileptics, diabetes and folate deficiency [1]. A few case reports suggest that amniotic band syndrome may be associated with fetal acrania [2]. It is unclear pSS increases the risk of fetal acrania [3]. Regardless, it is important to counsel patients that there is a 2-5% risk of recurrence and to take high-dose folate to reduce this risk in future pregnancies.

REFERENCES

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