

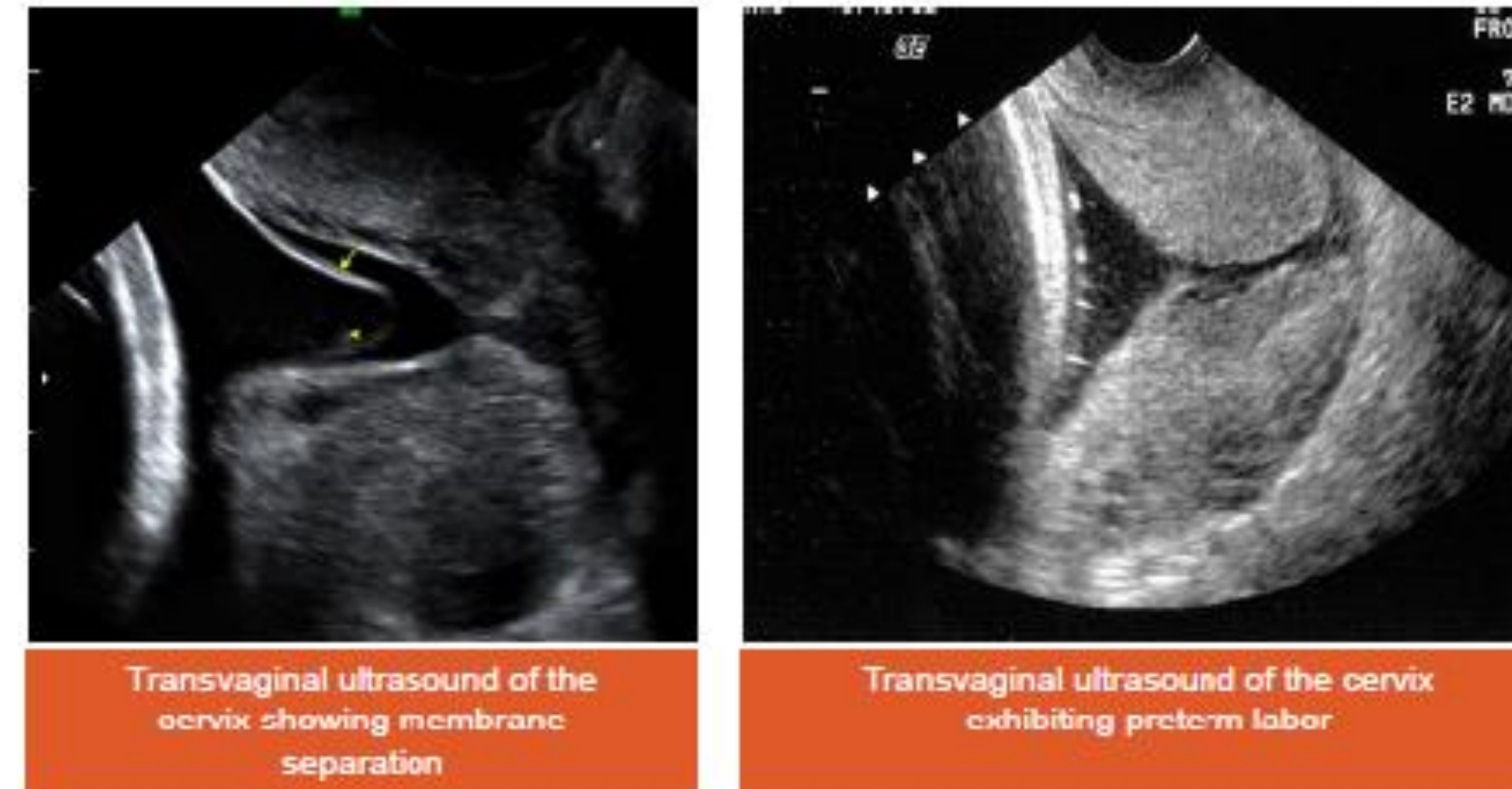
A Case Report on Cervical Insufficiency and Ehlers-Danlos Syndrome

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Introduction

- Cervical insufficiency should be suspected in patients with recurrent second-trimester pregnancy losses. Signs of cervical weakness or insufficiency include: recurrent second-trimester loss, no or minimal symptoms, and cervical dilation with no or minimal uterine contractions. A thorough past medical history should be taken to evaluate for a cause of cervical insufficiency.
- A 28-year-old, G11P0550 female, at an unclear gestational age, 21-24 weeks, was admitted for vaginal bleeding, and intermittent cramping. Patient has poor follow-up to obstetrical care, and stated that she was unsure of the root cause for her ten fetal demises and miscarriages. Patient had no prenatal care in this pregnancy.

Images



Discussion

- Ehlers-Danlos syndrome (EDS) is a connective tissue disorder that is characterized by skin hyperextensibility, joint hypermobility, and tissue fragility. The pathophysiology of EDS involves genes, usually autosomal dominant, that affects the synthesis of collagen. The cervix, being predominantly composed of collagen is affected by Ehlers-Danlos syndrome. Uterine prolapse has also been incited by EDS. Cervical insufficiency is directly correlated to EDS by the cervical weakness that this syndrome causes. By understanding the underlying cause of the patient's recurrent second trimester loss, early steps can be taken to prevent another loss. Cervical cerclage in the twelfth to fourteenth week of gestation can prevent shortening of the cervix to cause preterm labor.
- Symptoms of cervical insufficiency usually begin between fourteen to twenty weeks gestation, and present with pelvic pressure. Braxton-Hicks like contractions, premenstrual cramping, backaches, or change in vaginal discharge. Tocodynamometry shows no or infrequent contractions at irregular intervals. Membranes may be prolapsed, or in extreme cases, ruptured. In most cases, cervical length is shortened, less than 25 mm. In this case, the patient had been experiencing intermittent uterine cramping for the past week, most likely inciting complications of cervical insufficiency. Details of the patient's previous cerclage are unknown; and therefore, the patient's best outcome for a delivery in a subsequent pregnancy is an ultrasound, transabdominal or laparoscopic changes.

Case Summary

- On presentation, patient was complaining of intermittent vaginal spotting and uterine cramping for the past week. Associated with good fetal movement, and no loss of amniotic fluid.
- **Past OB history:** Patient has a poor prenatal history of multiple losses, previous cerclage, classical Cesarean delivery at 26 weeks, resulting in neonatal demise, and spontaneous vaginal delivery at 36 weeks, resulting in neonatal demise secondary to cystic kidneys.
- **PMH:** Major Depressive Disorder, Borderline Personality Disorder, Post-traumatic Stress Disorder, Tourette Syndrome, polysubstance use disorder on Methadone 170 mg PO daily, and homelessness
- **Imaging:** Bedside ultrasound with Maternal Fetal Medicine showed a single, viable fetus in the breech presentation, with a heart rate of 153 beats per minute. Placenta was anterior. Amniotic fluid index was 13.5. No structural gross or anatomical abnormalities. Cervix was dilated at 2.4 cm with prolapsed membranes about 3.2 cm beyond the external cervix os, with fetal feet in the prolapsed membranes, within the vagina. Estimated weight of the fetus was 406g. Approximate gestation 20 3/7 weeks. Rescue cerclage was forgone due to uterine tenderness.
- **Hospital Course:** Patient was admitted to Labor and Delivery to rule out chorioamnionitis, started on IV fluids, and received IV Ampicillin and IV Azithromycin. Patient and father of the baby elected not to induce labor, as contractions were absent, and patient wanted all protective measures taken to allow for a viable fetus. Fetal heart monitoring was appropriate for gestational age.
- Patient remained afebrile, with no signs of infection. After a further, thorough discussion of the patient's PMH, patient revealed she had a prior diagnosis of Ehlers-Danlos syndrome. Patient remained on inpatient bedrest until 22 6/7 weeks. At this time, bedside ultrasound revealed a 598g fetus in the transverse lie. Cervix dilated to 2 cm, with prolapsed membranes. No contractions. Patient stable to be transferred for a higher level of care, now that the fetus had reached a viable, gestational age. At 23 0/7 weeks, patient was transferred to a Level III NICU for a higher level of care.

Conclusion

- Despite the common risk factors for cervical insufficiency, there are rare diagnoses that can impact a woman's gestation. A thorough history taking should be elicited to rule in or out these rare diagnoses, when common causes are noncontributory. In this case, Ehlers-Danlos syndrome most likely contributed to this patient's extensive history of cervical insufficiency.

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