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Localized Intravascular Coagulopathy of Venous Malformation Involving the Labia as a Mimic of Child Sexual Abuse: A Case Report

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Localized Intravascular Coagulopathy of Venous Malformation Involving the Labia as a Mimic of Child Abuse: A Case Report

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INTRODUCTION

- Child abuse is a significant public health challenge in the United States.
- The National Child Abuse and Neglect Data System (NCANDS) estimated 674,000 victims of child maltreatment in the United States in 2017 with almost 45,000 children affected by sexual abuse.[1, 2]
- Ano-genital injuries in children are rare in children and often prompt concern for maltreatment.
- Medical literature identifies a wide spectrum of injury severity attributed to mechanisms including accidental falls, motor vehicle accidents and less commonly child abuse.
- Indicators of physical injury are often not identified in the evaluation of child sexual abuse.[3, 4]
- It is essential for providers to identify abuse and to accurately unique non-abusive findings and avoid unnecessary protective services interventions and criminal proceedings.[5]
- Mimics of sexual abuse are reported, including urethral prolapse, rectal prolapse, lichen sclerosis, vulvar ulcers, and erythema, inflammation and fissures of the perianal or vulvar tissues due to non-sexually transmitted infections.[5]
- We present a case of localized intravascular coagulopathy as a complication of venous malformation, which sufficiently mimicked trauma to prompt consultation with a child abuse pediatrician.

CASE PRESENTATION

- Five-year-old female presented to a pediatric urgent care clinic, accompanied by her mother, with complaint of vaginal pain while bathing.
- Her mother visually inspected her genitals and described a new raised, blue-purple bump on the right labia that was painful to touch.
- Mother voiced concern for the possibility that the physical finding could be the result of sexual abuse as the patient had recently stayed with her father, from whom mother is separated. The mother reported remote history of sexual abuse on the paternal side of the family but did not report that the patient had contact with individuals of concern.
- There was no report of accidental trauma such as a fall or straddle injury. The patient specifically denied inappropriate sexual contact.
- Her past medical history was significant for a venous malformation on the right buttock that had been present since birth.
- She was seen by Pediatric Dermatology at the age of three at which time there was low clinical suspicion for deeper involvement of the venous malformation. Nd:YAG laser ablation therapy was offered at that time, which mother declined due to need for general anesthesia. The child did not return for additional pediatric dermatology follow-up prior to her acute presentation due to new labial finding.
- She had no prior history of urinary tract infections, and denied dysuria, hematuria, vaginal discharge or bleeding.

Physical Exam:
- Tender 1 x 0.5 cm blue-purple firm nodular mass on the right labia with discoloration and engorgement extended to the clitoris and right labia minora sparing the clitoral hood.
- Collection of multiple non-tender violaceous reticular blanching lesions approximately 15 cm in length visible from the lateral margin of the right buttock and thigh medially to the gluteal cleft consistent with venous malformation (Figs. 1a-c).

Consultation:
- Obstetrics/Gynecology recommended obtaining sexually transmitted infection (STI) screening for concerns of hematoma secondary to vaginal trauma.
- Child Abuse Pediatrics was less suspicious for trauma based on the patient's history of venous malformation, the unique features of the new lesion, and consistent denial by the patient of physical trauma or inappropriate sexual contact.
- Multi-disciplinary Vascular Malformations Program recommended laboratory analysis and a genitourinary/pelvic MRI with contrast.

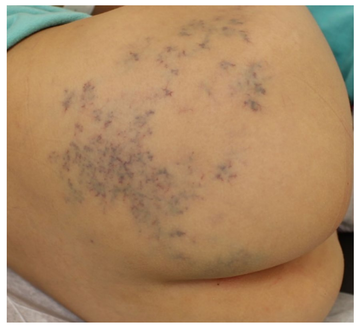

Fig 1c. Known venous malformation of the right buttock.

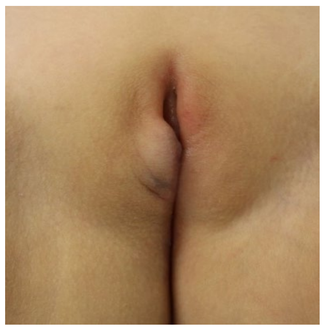

Fig 1a. Fullness and minimal discoloration of right labia majora.

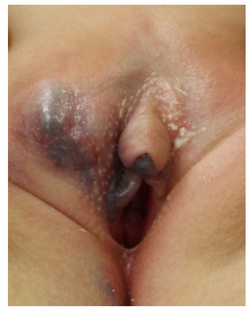

Fig 2b. Painful nodular purple mass on inner right labia majora. Engorgement of the right labia minora, clitoris, with sparing of the clitoral hood.

DISCUSSION

- Localized intravascular coagulopathy (LIC) is a well-described occurrence in individuals with venous malformations, occurring in approximately 40% of patients with these lesions.
- LIC occurs due to relatively slow flow of blood through dilated vessels with associated disruption of the basement membrane, which triggers activation of the coagulation cascade.[6]
- The presence of LIC is supported by evaluation of the fibrin split products D-dimer and fibrinogen.[7]
- A significantly elevated D-dimer level with an associated low fibrinogen level (<150 mg/dL) defines severe LIC and carries risk for progression to disseminated intravascular coagulopathy (DIC).
- LIC has also been associated with increased risks of bleeding, deep vein thrombosis, and thromboembolic disease.

- Recent studies have shown that there is an increased risk of LIC in patients in the following circumstances: larger venous malformations, the presence of phleboliths (either on imaging studies or when palpable during physical examination), in association with intramuscular lesions, with lesions present on the trunk as opposed to extremities, and with treatments such as surgical intervention or sclerotherapy.[8]
- Early therapeutic interventions such as low molecular weight heparin have been used to prevent LIC from progressing toward these potential complications.[9]

- This case represents an example of the importance of objective assessment in unusual case presentations to ensure diagnostic accuracy, and appropriate direction of medical and child protection resources.
- Thorough history, included open-ended interview of the patient, full physical examination, and appropriate ancillary studies are essential in evaluating cases of genital abnormalities, particularly where concern for abuse is raised.
- This practice ensures thorough evaluation and treatment of medical conditions while avoiding unnecessary escalation of social investigation.

INTERVENTION/TREATMENT

Laboratory analysis revealed:
- Elevated D-Dimer of 840 ng/mL (reference range 0-500 ng/ml)
- Normal PT/INR, PTT, and fibrinogen
- Complete blood count (CBC) revealed a normal platelet count, slightly elevated Hemoglobin of 14.7, and Hematocrit of 43.
- Urine STI screen was negative for gonorrhea and chlamydia.

MRI findings:
- Ill-defined T1 hypointense and T2 hyperintense lesion within the medial aspect of the right gluteal soft tissues and gluteal musculature, measuring approximately 10.7 cm in craniocaudad extent with heterogeneous enhancement.
- Additional larger T2 hyperintense cystic lesions within the right gluteus maximus musculature (at the posteromedial aspect) demonstrated no significant enhancement. The largest fusiform lesion measured 2.4 x 1.1 cm (Fig 2a).
- Ovoid heterogeneously T2 hyperintense 1.4 x 0.9 cm lesion within the medial right labial soft tissues with T2 hyperintensity extending along the medial pelvic sidewall with peripheral mild enhancement of this lesion (Fig. 2b). Normal ovaries and uterus.

Findings reported by Pediatric Radiology were consistent with a venous malformation within the right gluteal soft tissues and musculature, and low-density clot/thrombus within the medial right labial soft tissues with final diagnosis of localized intravascular coagulopathy (LIC) of venous malformation with vulvar involvement.
The patient was discharged to follow up with the Multidisciplinary Vascular Lesion Clinic. Acute symptoms were managed with Sitz baths and ibuprofen; the family ultimately pursued combined laser and sclerotherapy of the malformation without recurrence of symptomatic LIC within one year of initial diagnosis.

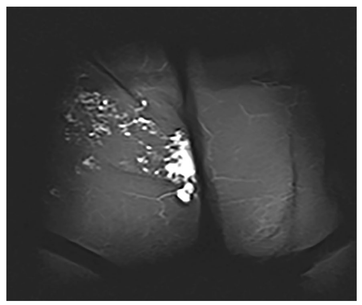

Fig 2a. MRI with contrast demonstrating gluteal involvement of venous malformation.

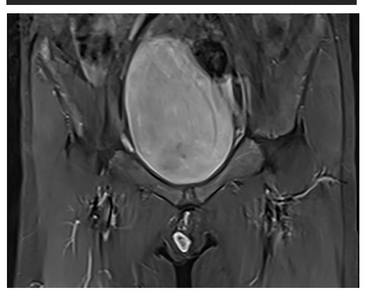

Fig. 2b. MRI with contrast demonstrating right labial LIC of venous malformation.

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DISCLOSURES

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