BEYOND COINCIDENCE? AN UNEXPECTED PAIRING OF DEVELOPMENTAL ANOMALIES IN A 3 YEAR OLD FEMALE

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Introduction:

The Canal of Nuck is an anatomic phenomenon in females that occurs when the *processus vaginalis* fails to obliterate. This patency allows for herniation of abdominal or pelvic structures such as ovaries, fallopian tubes and peritoneum into the inguinal canal and down to the labia majora. These hernias can present as classical hernias, mimic Bartholin cysts, lymphadenopathy, or ovarian torsion.

Craniosynostosis is the premature fusion of cranial sutures which can lead to impaired cerebral development, neurologic deficits, and may be associated with syndromic conditions such as FGFR or TWIST1 mutations that affect other body systems. Given this correlation, it is essential to consider other anomalies when craniosynostosis is present. We present a case of a patient with concomitant craniosynostosis and a hernia into the Canal of Nuck, highlighting the diagnostic challenges and consideration of syndromes when craniosynostosis is involved in an atypical anatomic presentation.

To our knowledge, the coexistence of these conditions in the same patient has not been previously described.

Case presentation:

A 3-year-old female with a history of craniosynostosis, chronic constipation, ventriculoperitoneal shunt and right inguinal hernia repair presented with one week of reduced appetite, increased straining with defecation, and a new left inguinal bulge. Parents were concerned about a second hernia.

Examination showed a well-appearing, afebrile child with moist mucous membranes. The abdomen was moderately distended with a tender palpable bulge in the left inguinal region without overlying skin changes. The right inguinal region had a well-healed surgical scar.

Ultrasound images showed an abnormal appearance of the left inguinal region that was interpreted by the radiologist as an encysted hydrocele of the Canal of Nuck or an incarcerated inguinal hernia without bowel involvement. (Fig 1) Pediatric surgery was consulted, however

prior to the surgical assessment, the bulge had spontaneously reduced and the decision was made to pursue elective repair in the outpatient setting.

An open surgical repair was performed successfully under general anesthesia 1 month after presentation which the patient tolerated well. Her post operative course was uncomplicated.

Discussion:

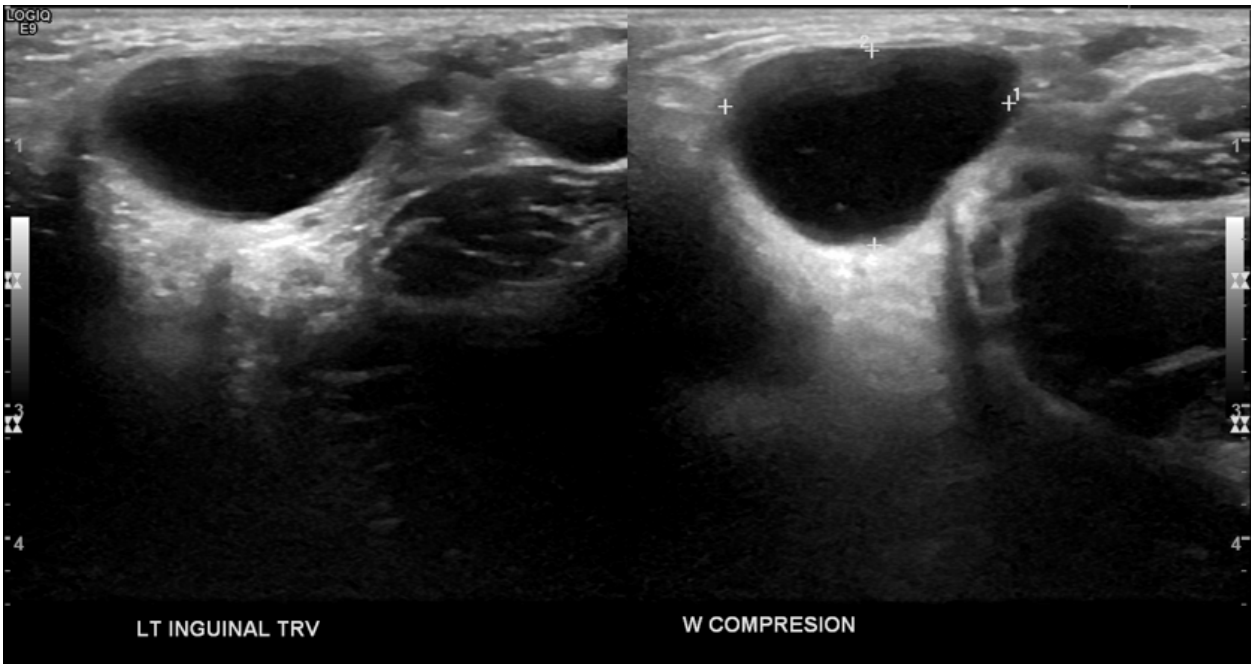
Craniosynostosis is often associated with syndromes affecting multiple organ systems, and this case highlights the need for early recognition of atypical hernias in syndromic patients as they carry risks for complications such as incarceration or strangulation.

In the Emergency Department, Canal of Nuck hernias may pose a diagnostic challenge due to their anatomic origin. Presentation can vary depending on the structures involved, for example these hernias may contain the ovary and cause symptoms more consistent with ovarian torsion,[6] a diagnosis that is rarely considered in this patient's age group but may cause significant morbidity. A prompt sonographic assessment is indicated, and the Canal of Nuck hernia may only be discovered when the affected ovary cannot be visualized in the typical location on ultrasound. This is helpful in the diagnosis of the patient's condition, and may assist in surgical planning.[7] A multidisciplinary approach to evaluation and treatment is recommended, with consideration of both gynecologic and surgical input depending on the structures involved. Patients and their families should also be educated on the anatomic anomaly to allow for better communication with surgical or emergency department teams should an acute abdominal process arise later.

Conclusion:

Canal of Nuck hernias are rare and pose a diagnostic challenge. The clinician must maintain a high index of suspicion and obtain ultrasound imaging to guide management and prevent complications. The case also highlights a potential relationship between craniosynostosis and hernias into the Canal of Nuck and emphasizes the importance of considering atypical presentations in other organ systems when craniosynostosis is present. Further research is needed to fully characterize this relationship.

Figure 1:



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