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Case Report

Splenogonadal fusion in a female: A case report of a rare congenital anomaly ☆

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ABSTRACT

We present a case of an 18-year-old female referred for an MRI pelvis to evaluate suspected uterine anomaly by ultrasound. The MRI showed a complete septate uterus and in addition, an elongated tubular structure (isointense to the spleen) extending from the left ovary in the left retroperitoneal region/left paracolic gutter to the under-splenic surface. CT abdomen and pelvis revealed this to be a similarly enhancing structure as the spleen and appears as a tubular retroperitoneal structure connecting the left ovary to the spleen with associated vasculature joining the splenic vein cranially and the ovarian vessels caudally consistent with splenogonadal fusion.

Introduction

Splenogonadal fusion (SGF) is a rare benign congenital anomaly with few cases described in the literature being 16 times more common in males than in females usually presenting with cryptorchidism or testicular mass and usually on the left side between the left testis and the spleen. It is rarely diagnosed preoperatively.

Case presentation

We present a case of splenogonadal fusion in an 18-year-old female referred for MRI Pelvis to evaluate for uterine anomaly suggested on pelvic ultrasound examination. The MRI revealed preserved outer uterine contour with 2 endometrial cavities separated by midline low T2 signal intensity septum extending inferiorly to the uterine cervix consistent with

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complete septate uterus (Fig. 1). Incidentally a bandlike soft tissue connection between the spleen and the left ovary was partially included in the MRI study (Fig. 2) so further assessment with proper CT abdomen and pelvis was requested. The CT scan redemonstrated the band-like soft tissue connection between the spleen and left ovary associated with vasculature communicating with the splenic and left ovarian vessels in keeping with splenogonadal fusion (Fig. 3).

Our patient and her family were well educated about the condition, and the possibility of related complications. She is under regular follow-up and expecting her first child with no impact on her quality of life up to date (Fig. 4).

Discussion

SGF is a rare congenital malformation with abnormal fusion between the spleen and gonads or mesonephros derivatives, first described by Pommer in 1889 [1]. It is more common in

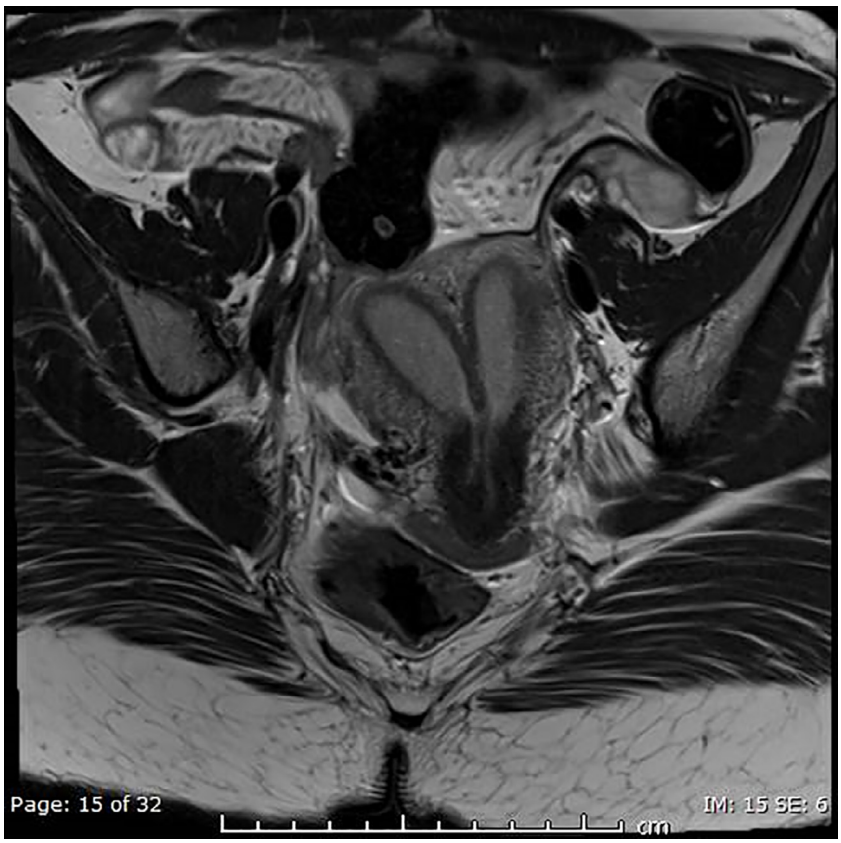

Fig. 1 – MRI Pelvis Coronal T2-weighted image shows low signal septum separating 2 uterine cavities and extending to the endocervical canal. The outer uterine contour is preserved. Appearances consistent with complete septate uterus.

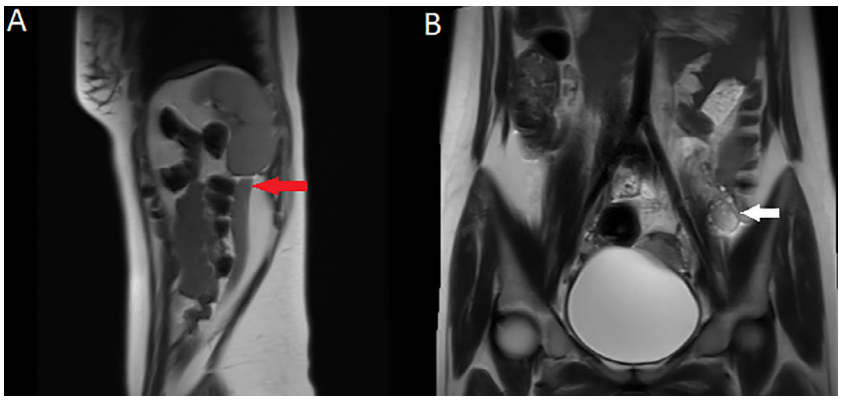

Fig. 2 – MRI Pelvis T2-weighted images, (A) sagittal and (B) coronal planes, demonstrate tubular structure isointense to the spleen in the left retroperitoneal region (red arrow) extending down to a normal appearing left ovary (white arrow).

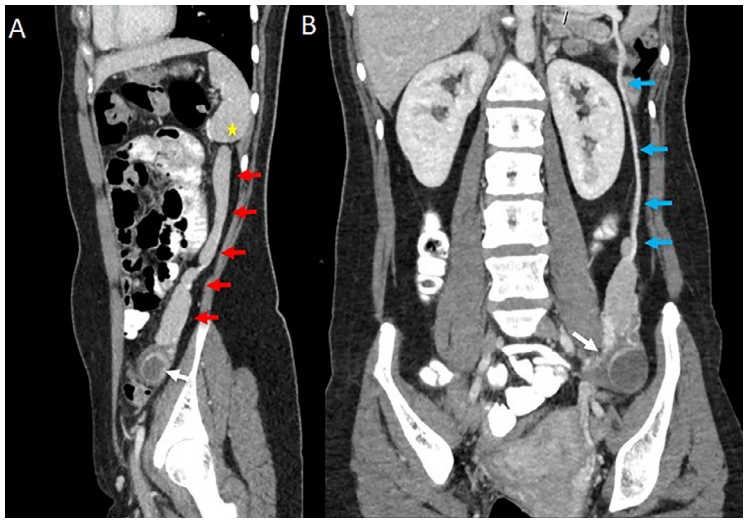

Fig. 3 – Sagittal (A) and oblique coronal and (B) reconstruction of CT Abdomen and pelvis with contrast, confirming a tubular structure (Red arrows) connecting the spleen (yellow star) to the left ovary (white arrow) with vascular connection (Blue arrows).

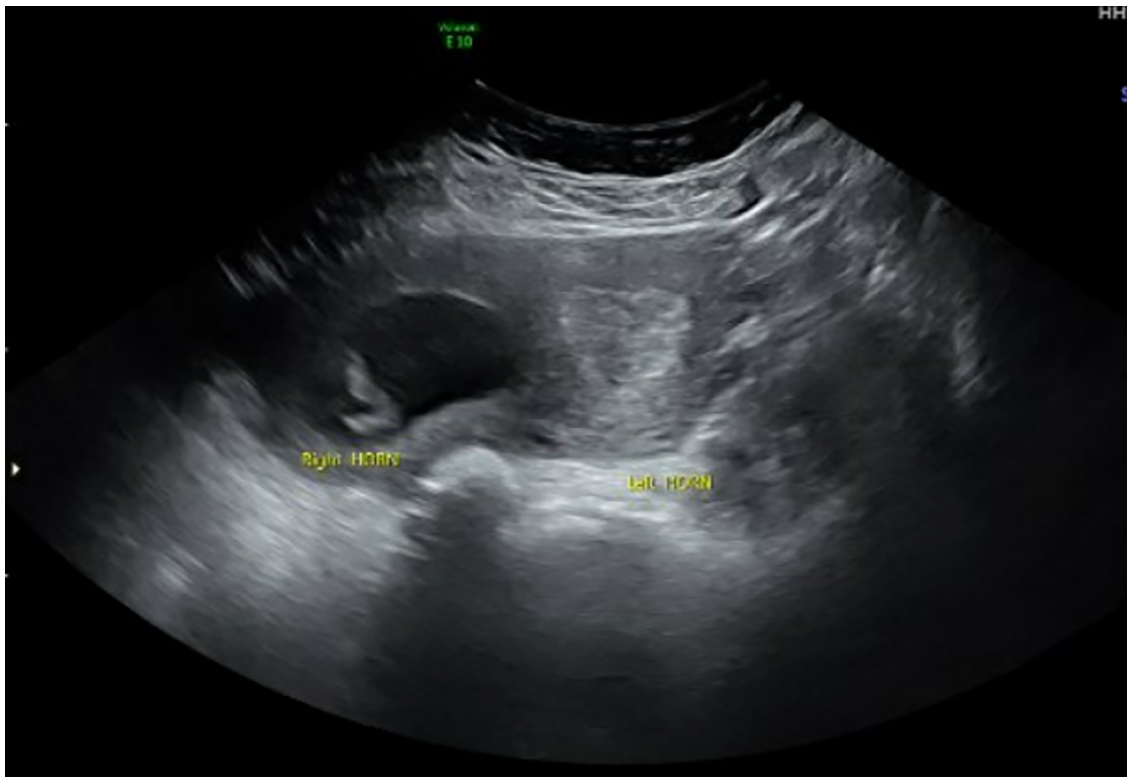

Fig. 4 – Ultrasound of the pelvis (14 months after initial presentation) demonstrating early pregnancy in the right uterine horn of septate uterus.

males (~95%), occurs most commonly on the left (98%), and usually involves the testis (95%). It has been found in patients from birth to 81 years old. There are 2 main types, continuous and discontinuous. In the first type, the gonad is linked to the spleen by a trans- or retroperitoneal cord of splenic tissue or fibrous cord with nodules of splenic tissue; often associated with other congenital anomalies. While in the discontinuous type, ectopic rests of splenic tissue are present in the gonad [2]. Alujevic et al. [3], described the first case of a continuous type splenogonadal fusion in an adult female, and they mentioned that many cases of the continuous type of splenogonadal fusion are associated with congenital malformations.

Meneses [1] described a case of discontinuous type splenogonadal fusion in a female without associated anomalies.

SGF has a clear predilection for males; in a recent literature review conducted by Kumar et al. [6], from 2013, a total of 67 cases were reported, only 1 case was a female (1.4%). Khairat et al. [5], described that most male cases are usually detected incidentally during routine groin surgical exploration for undescended testis or hernia. Although, when symptomatic, painful scrotal swelling is the usual presentation. Various pathologies can affect the ectopic splenic tissue, like rupture from direct trauma. Furthermore, mumps, leukemia, mononucleosis, and malaria can precipitate splenic rest torsion in the scrotum. To our knowledge, only 4 cases were reported with coexisting splenogonadal fusion and malignant testicular neoplasm.

However, in females, SGF is often detected incidentally at abdominal surgery for various abdominal causes and at autopsy. Minority of cases can present symptomatically with abdominal pain relating to gonadal or splenic torsion. Guzman and Wieck [4] described symptomatic SGF with splenic torsion in an adolescent female presenting with left lower quadrant abdominal pain. CT demonstrated a cord of splenic tissue that extended inferiorly to the left lower quadrant and was associated with the left adnexa. A segment of hypoenhancing tissue and surrounding fat stranding was concerning for infarcted splenic or gonadal tissue. Pathology confirmed necrotic splenic tissue.

Interestingly, the literature describes a 5-fold higher risk of associated anomalies in the continuous compared to the discontinuous type, including cryptorchidism (more bilaterally), congenital inguinal hernias, peromelia, micrognathia, cardiac defects, cleft palate, anal malformation among others [5].

Different imaging modalities can aid in the delineation of splenogonadal fusion including ultrasound, CT, MRI, and nuclear studies. Ultrasound is usually the first line of imaging to detect an abnormality related to the spleen or gonads (ovaries and testes); either as an incidental finding or for evaluation of suspected testicular swelling or mass. Our patient was referred for pelvic MRI after an incidental sonographic finding of uterine anomaly. Cross-sectional imaging with CT and MRI is more superior in the characterization of pelvic abnormalities including splenogonadal fusion. In the continuous type of SGF, these modalities clearly identify a cord of splenic tissue connecting the spleen to the gonad. In our case, the MRI demonstrated a complete septate uterus with an incidental finding of a retroperitoneal cord-like structure with similar signal characteristics to the spleen and possible connection to it. Further imaging evaluation by CT abdomen and pelvis confirmed a cord of splenic tissue connecting the spleen to the left ovary consistent with continuous type SGF. In the discontinuous type, however, a mass of similar density and signal intensity to the spleen in CT and MRI respectively resting in or at the vicinity of the gonad could be a clue to the diagnosis. Nuclear imaging with Tc^{99}m-sulfur colloid is a further confirmatory test, particularly in the suspicion of discontinuous type.

Conclusion

SGF is a rare congenital condition in females often detected incidentally for evaluation of suspected anomaly or at autopsy. If symptomatic, it could present with splenic or ovarian torsion. Cross-sectional imaging with CT and MRI is superior in characterization of pelvic abnormalities including SGF, and we recommend the addition of T2 coronal acquisition for the whole abdomen in MRI pelvis requested for the evaluation of suspected uterine anomaly to aid in the detection of other associated anomalies.

Patient consent

Written informed consent was discussed and obtained from the patient for publication.

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