

Andrology and fertility

Splenogonadal fusion: A rare case report and literature review

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ARTICLE INFO

Keywords:

Splenogonadal fusion
SGF
Robotic surgery
Testicular cancer

ABSTRACT

Splenogonadal fusion is a rare benign congenital anomaly with few cases described in the literature. It is 16 times more common in males than in females. A 22 year-old healthy male with cryptorchidism presented with pre-operative imaging strongly suggestive of malignancy. Histopathology after left orchiectomy showed mixed splenic and testicular tissue with no sign of malignancy. Splenogonadal fusion is rarely diagnosed preoperatively. It should be included in differential diagnoses in patients presenting with a testicular or abdominal mass. Greater recognition of this rare anomaly may facilitate testis sparing surgery in future cases.

Introduction

Splenogonadal fusion (SGF) is a rare congenital anomaly in which there is abnormal fusion between the spleen and the gonads or deformities of the wolffian duct structures such as the epididymis. Few cases have been reported in the literature since it was first described by Bostroem in 1883.¹ Most cases are diagnosed with postoperative histology as imaging studies are not distinctive.² We present our experience with a rare case of SGF.

Case presentation

A 22 year-old male was referred to our institute with a left undescended testis with suspicion of a malignant lesion (seminoma). The patient had no significant medical or surgical history. He had never felt his left testis in the scrotum, and there was no history of scrotal trauma. His general physical examination was unremarkable. Deep abdominal palpation revealed a mobile left lower quadrant mass. The consistency of the mass could not be evaluated due to its depth. The left hemiscrotum was empty and the right testis was hypertrophied. The left testis was not palpable in the inguinal area. A left undescended testis with malignancy was strongly suspected. Routine blood tests were within normal range. Alpha-fetoprotein, human chorionic gonadotropin (hCG), and lactate

dehydrogenase (LDH) were also within normal range. Scrotal and inguinal ultrasound (US) was unremarkable. Pelvic and abdominal magnetic resonance imaging (MRI) revealed an oval shaped lesion within the left iliac fossa with homogeneous, intermediate high T2 and isointense T1 signal intensities, with diffusion restriction and intense portal venous contrast enhancement, much higher than the splenic enhancement (Fig. 1). The patient underwent robotic left orchiectomy (Fig. 2). Intraoperative findings revealed a left testis with a gonadal vein draining into the left renal vein. A thick, cord like structure connected the superior border of the left testis to the inferior border of the left kidney. In addition, there was continuous fibrous tissue between the testis and the spleen, posteriorly to the left kidney. The procedure was uneventful, and the patient was discharged home on postoperative day 2. Histopathology showed mixed splenic and testicular tissue with no sign of malignancy (Fig. 3A and B). Follow-up examination in our outpatient unit was unremarkable.

Discussion

SGF is a rare congenital malformation with less than 200 cases have been reported since its first description by Bostroem in 1883.^{1,2} The few reported cases are mainly in adolescents and young adults as in our case, with 72% in individuals less than 20 years of age.³ SGF occurs on the left

Abbreviations: SGF, splenogonadal fusion.

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<https://doi.org/10.1016/j.eucr.2020.101307>

Received 8 June 2020; Received in revised form 15 June 2020; Accepted 17 June 2020

Available online 17 June 2020

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Fig. 1. Magnetic resonance imaging (MRI) of the abdomen and pelvis shows an oval shaped lesion within the left iliac fossa.



Fig. 2. Intraoperative findings show the left testis with the gonadal vein draining into the left renal vein, and a thick, cord-like structure arising from the superior border of the left testis.

side in 98% of the cases, and it occurs in males 16 times more frequently than in females.⁴ Few female cases have been reported, in part due to the difficulty of assessing female gonads by physical examination.⁴ SGF is divided into continuous and discontinuous types.^{4,5} In the continuous type, the principle spleen is connected to the gonad by a continuous cord-like structure of fibrous or splenic tissue. Alternatively, the discontinuous type shows a connection between the gonad and an ectopic spleen.^{4,5} While most reported cases were diagnosed incidentally after surgical intervention as in our case, 17% were diagnosed at autopsy.⁴ Cortes reviewed 111 cases of SGF, and cryptorchidism was the most commonly associated anomaly, seen in 31% of cases. Compared to the discontinuous type, continuous SGF carries a five-fold higher risk of associated anomalies such as peromelia, micrognathia, cardiac defect cleft palate, and spina bifida.^{3,4} Our case presented as an intra-abdominal testis with suspicion of malignancy. Several neoplastic and non-neoplastic conditions may mimic testicular seminoma at imaging. Benign mimics other than SGF include segmental infarction, hematoma, infection, epidermoid cyst, adrenal rests, sarcoidosis, and sex cord-stromal tumors.² An association of SGF with some genetic syndromes and mutations has also been described in the literature. SGF is usually recognized after radical orchiectomy due to suspicion of malignancy, as in our case. A lack of conclusive diagnostic studies is a factor in delayed diagnosis, although radio colloid spleen scintigraphy with technetium-99 m has been used to identify accessory splenic tissue to pre-operatively diagnose a very few cases.^{4,5} Due to the preoperative radiographic findings and strong suspicion of malignancy in our case, robotic abdominal orchiectomy was performed after patient counseling.

Conclusion

SGF is a rare congenital anomaly that should be included in the differential diagnosis of solid left lower abdominal or scrotal mass. The preoperative diagnosis of SGF is usually difficult, especially with cryptorchid testis. Radiocolloid imaging may be useful, but the surgeon should use caution as it does not have 100% sensitivity. Most surgeons select radical treatment to decrease the risk of missing malignancy. Urologist awareness of this rare entity may enable testis sparing surgery in future cases.

Consent

The patient has given his consent for the case report to be published. A copy of the written consent is available on request from the authors.

Ethical approval

Case reports are exempted from IRB/ethical approval at the institution.

Source of funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

Declaration of competing interest

None.

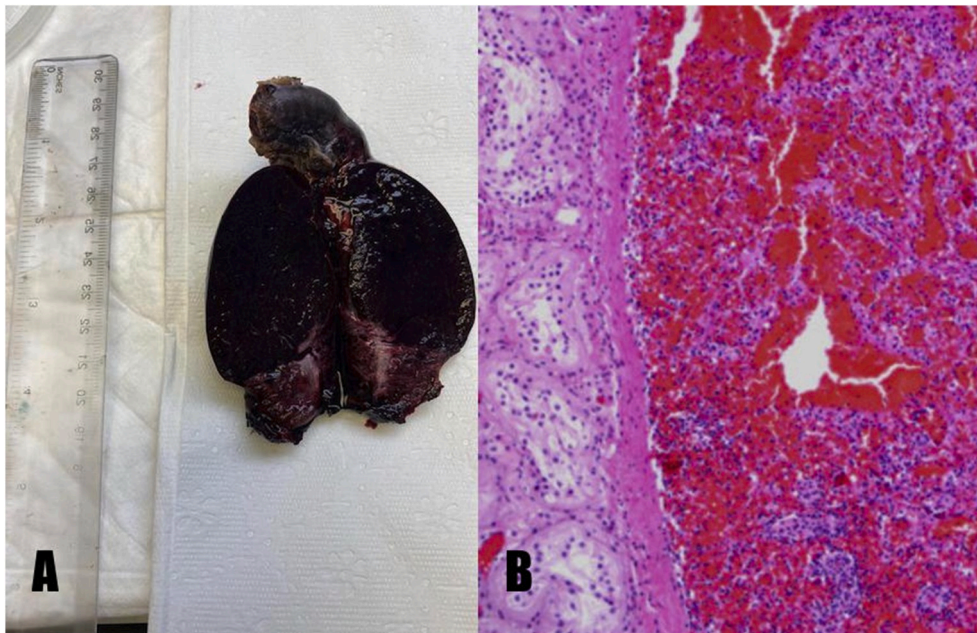


Fig. 3. A: Gross appearance of the 66 g surgical specimen containing a cord-like structure. B: Histopathology shows splenic tissue on the right side, and testicular tissue with a Sertoli cell only pattern.

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