Received: 21 March 2023 | Revised: 31 March 2023 | Accepted: 12 April 2023

DOI: 10.1002/ccr3.7264

Clinical Case Reports Open Access WILEY

CASE REPORT

A case of splenogonadal fusion in the left testis

Ting Zhou

Department of Pathology, The Second Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou, China

Correspondence
Ting Zhou, Department of Pathology, the Second Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou, 310000, China.
Email: 2520091@zju.edu.cn

Key Clinical Message

Splenic tissue outside the normal anatomical site can be collectively referred to as ectopic spleen. Clinically, the commonest causes of ectopic spleen include accessory spleen, splenic tissue implantation, and splenogonadal fusion (SGF). Accessory spleen is mostly caused by congenital dysplasia, is mostly located near the spleen, and may be supplied by the splenic artery. Splenic implantation is mostly caused by autologous spleen tissue transplantation caused by trauma or surgery. SGF is the abnormal fusion of the spleen with the gonad or with the mesonephric derivatives. As a rare developmental malformation, it is difficult to make a correct diagnosis preoperatively, and easily misdiagnosed as a testicular tumor cause lifelong harm to patients. An 18-year-old male student who developed left testicular pain without obvious cause that radiated to the perineum 4 months prior to presentation. He was diagnosed with cryptorchidism 12 years ago and underwent orchiopexy without intraoperative frozen section examination. An ultrasound was performed, identifying hypoechoic nodules in the left testis, suggestive of seminoma. During surgery, the testicular tumor revealed a dark red tissue and the diagnosis of a pathological ectopic splenic tissue was made. Because the clinical manifestations of SGF are not specific, misdiagnosis and unnecessary orchiectomy may occur. If a complete preoperative examination which includes biopsy or intraoperative frozen section is performed, unnecessary orchiectomy can be effectively avoided and bilateral fertility can be preserved.

KEYWORDS

case report, correct diagnosis, cryptorchidism, pathology, splenogonadal fusion

1 | INTRODUCTION

Splenogonadal fusion (SGF) is a rare developmental deformity that occurs due to the abnormal fusion of the spleen and the gonads during embryonic development and is usually accompanied by other deformities.[1] It lacks of obvious clinical and pathological features, which makes it difficult to diagnose before surgery. SGF usually presents as a testicular mass or in combination with cryptorchidism, inguinal hernias, etc., and is easily misdiagnosed as a malignant tumor. A recently diagnosed patient with cryptorchidism and SGF is described in this study to increase awareness about the disease.

2 | CASE REPORT

An 18-year-old male patient presented with complaints of a mass in the left testis after being diagnosed with cryptorchidism 12 years ago and underwent orchiopexy. The mass size about 1.0 × 1.0 cm and did not enlarge or shrink during 12 years. There was no obvious discomfort, hematuria, pain, swelling and other discomforts. Four months ago, there was swelling and pain which radiated to the left testis and perineum and there was no obvious cause. However, the size of the mass did not change and there was no associated hematuria. He presented to a local hospital for treatment, and computerized tomography (CT) results showed that his bilateral testicles were of relatively small size and volume with high blood flow to the upper left testis. A seminoma was considered and surgical treatment was recommended. The patient requested for an expert management and presented to our hospital. Upon admission, serum human chorionic gonadotropin (HCG) and alpha-fetoprotein (AFP) levels were normal. On ultrasound, the left testis was normal in size with regular morphology and a clear outline. The envelope was smooth and a class of round hypoechoic nodule could be seen within the parenchyma, measuring about 1.8 × 1.4 cm with a clear boundary and irregular morphology. Abundant blood flow signals were seen on color Doppler flow imaging (CDFI). No abnormalities were observed in the right testis, bilateral epididymis, and bilateral spermatic cord veins (Figure 1). Ultrasound test revealed a left testicular hypoechoic nodule, based on which a seminoma was suspected. Eventually, a clinical diagnosis of testicular malignancy was made. An oblique left inguinal incision at the level of the internal inguinal orifice was made to radically remove the left testicular tumor through the left groin. The general examination of the tumor showed a dissected testicular tissue measuring 3.0 × 2.0 × 1.0 cm and a gray red nodule visible beside the testis measuring 2.0 × 1.0 cm. The cut surface was grayish red in color, soft, and had a complete envelope and a clear boundary. Microscopic findings showed that the ectopic spleen maintained the normal splenic structure, which includes splenic corpuscles, red pulp, presence of medullary cords and sinuses in the red pulp, and dilation of medullary sinuses of varying degrees. The testicular tissue was well demarcated from the ectopic spleen. The testis showed a manifestation of azoospermia, with only supporting cells in the seminiferous tubules and a small number of spermatogenic cells in the lumen of the local region. The spermatogenic cells in the testicular tissue: spermatogonial cells, primary spermatogonial cells, secondary spermatogonial cells, spermatids and spermatozoa were significantly reduced, while supporting cells were increased (Figure 2). The final diagnosis of discontinuous SGF was made. After more than a year of follow-up, the patient had no obvious discomfort.

3 | DISCUSSION

Splenogonadal fusion was originally described in 1883 and its type were described in 1956.[1] In the continuous type of SGF, an ectopic spleen in the testicles or epididymis possesses a cord-like structure, which could be fibrous, splenic tissue, epididymal tissue,[2] or testicular blood vessels, that is connected to the main spleen. The other type is the discontinuous type in which the intra-gonadal ectopic spleen is not contact with the main spleen. Commonly, it occurred in the left in both unilateral or bilateral cryptorchidism.[3] Meanwhile, the continuous type accounted for about 55% of all reported cases and was associated with higher congenital malformations, which included cryptorchidism, micrognathia, hypospadias, etc.[4] Three hypotheses have been proposed previously to explain the occurrence of SGF: the inflammation hypothesis, retroperitoneal pathway of the splenic cells contact with the gonadal anlage, and wrapping of splenic tissue in the white membrane.[4] It has been suggested that cryptorchidism and continuous SGF may be caused by abnormal development or lack of degeneration of the cranial suspensory ligament (CSL) in the testis.[5] Most scholars believe that SGF occurs due to a developmental malformation. Within 5–8 weeks of embryonic development, the spleen rotates close to the gonad due to the rotation of the stomach. If the descending gonads adhere to the spleen, it leads to the fusion of the two organs, and the spleen is found anywhere along the descended gonads, including the groin and scrotum.

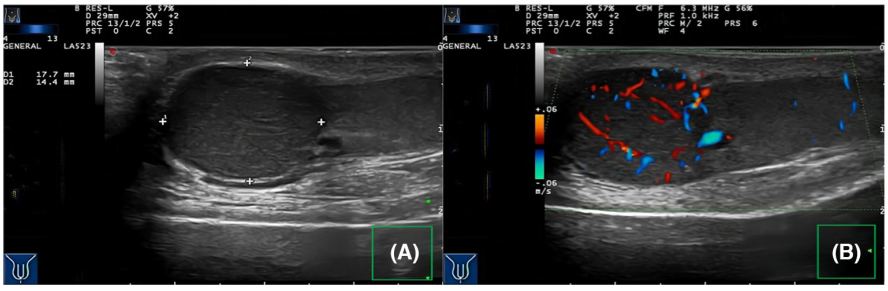

FIGURE 1 Ultrasound shows hypoechoic nodule within the parenchyma of the left testis with clear boundaries (A); CDFI shows an abundant blood flow signal in the nodule (B).

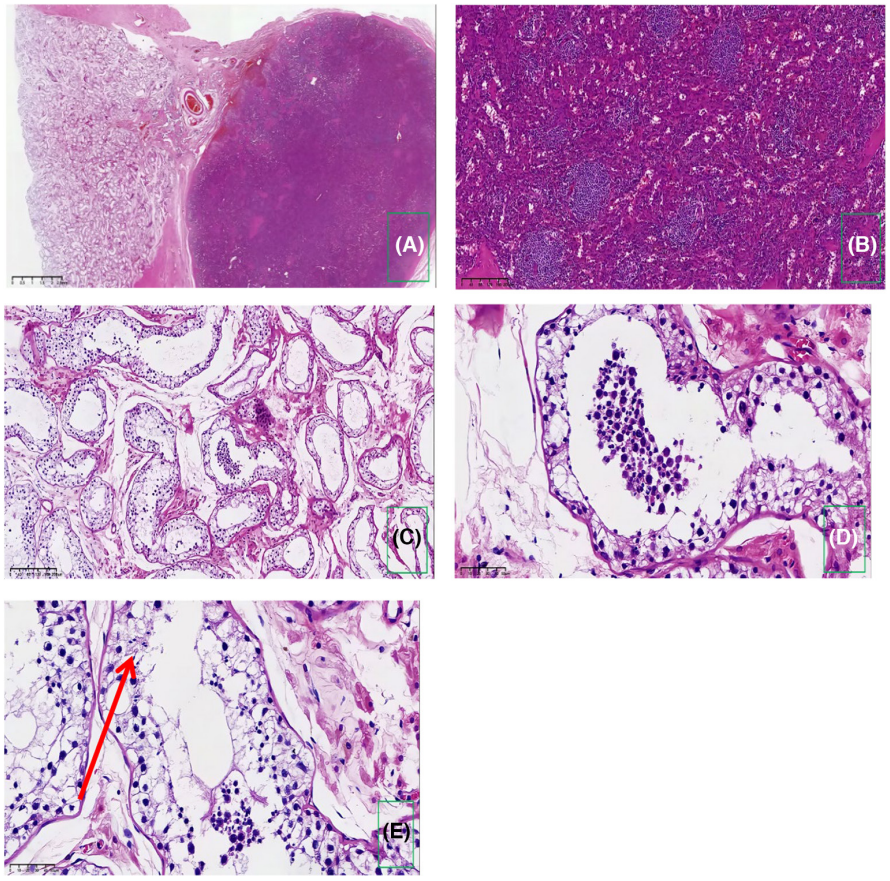

FIGURE 2 The appearance of SGF under the microscope. Digitally scanned sections show testicular tissue on the left, ectopic splenic tissue on the right, and a small testicular mediastinal mesh in the middle. The testicular tissue was well demarcated from the ectopic spleen ((A), H&E ×40). The ectopic spleen consisted of splenic corpuscles, red pulp, medullary cords and sinuses in the red pulp, and dilation of medullary sinuses of varying degrees ((B), H&E ×100). Some seminiferous tubules contained a small number of seminiferous cells ((C), H&E ×40). The seminiferous tubules contained a small number of seminiferous cells (no obvious mature sperms were found in this seminiferous tubules; (D), H&E ×400). The seminiferous tubules contained a small number of seminiferous cells. (a small number of spermatozoa could be seen as indicated by the red arrows; (E), H&E ×400).

Therefore, clinicians should pay close attention to the groin, iliac fossa, and paracolic sulci during surgery to avoid omission ectopic spleen.

Splenogonadal fusion can occur in both males and females, and the ratio is nearly 16:1.[6] The female reproductive organs are located in the abdomen, meanwhile, it is not easy to detect by palpation or observe without symptoms.[7] It may be because there are few reports on female patients.

Previous studies have reported only four patients who had cryptorchidism and testicular tumors.[1] Their testes were located in their abdomen, and this increased the risk of testicular tumors. It was reported that 64% of the patients underwent unnecessary orchiectomy.[3] Therefore, for patients with cryptorchidism, clinicians should also think of the possibility of SGF. A complete preoperative auxiliary examination which includes tumor markers, B-mode ultrasonography, computerized tomography (CT), enhancement CT (ECT) or magnetic resonance imaging (MRI), etc. should be performed as it can help to diagnose the SGF. Ultrasonography is very popular because it is affordable and can display the location and morphology of lesions during initial screening. Contrast-enhanced ultrasound (CEUS) and elastography are useful tools that can help to differentiate testicular malignancies from benign tumors such as ectopic splenic tissue.[8] The location, extent, morphological features, and tissue signatures of the mass can be observed by CT and MRI, which are helpful in evaluating paratesticular lesions and narrows the scope of differential diagnoses. MRI has no ionizing radiation, is safer, has a high soft tissue resolution, and may be more suitable for children.[9] In addition, using the ^{99m}Tc-nanocolloid splenic scintigraphy can improve the detectability of the lesions[10] and clearly diagnose SGF, so that a mass resection surgery without damage to the testes can be performed.[11] Laparoscopy is also a safe and effective method of diagnosing and treating SGF.[12] If permitted, we strongly recommend that clinicians perform

a preoperative biopsy or intraoperative freezing prior to surgery after confirmed benign lesions, laparoscopic exploration, testicular descent, and fixation are performed. As to whether the whole testis should be removed or if a gonad-sparing mass resection should be performed, some scholars believed that it depends on whether the gonads are normal and if there are adhesions between the gonads and the spleen. Surgery is not required when the diagnosis is clear and the gonad are free of complications.[10] Some authors have also performed surgery to retain the spleen, as completed removal of the spleen affects the blood supply to the patient's testis.[1] Overall, more clinical data is needed on whether splenic tissue should be removed or not. Perhaps with further understanding of the disease, more clinicians will choose gonad-preserving mass resection. Our patient presented with discontinuous type SGF after cryptorchidism surgery. Although it was unknown why SGF was not found during the previous operation, the serum tumor markers were normal. Only an ultrasound examination was performed and no further detailed examination was carried out. Therefore, our case was misdiagnosed as a malignancy preoperatively and a radical surgery was performed. On microscopy, a clear, thick capsule existing between the spleen and testicular tissue was found. This capsule separated the spleen and testicular tissue without damaging the testicular tissue. If not misdiagnosed before surgery, this finding would have allowed for a testicular-sparing mass resection. However, the patient's seminiferous tubules were atrophied and only a small number of spermatogenic cells were seen. If the contralateral testis is normal, an orchiectomy of the affected side will not have much impact on the patients' fertility.

4 | CONCLUSION

Splenogonadal fusion is a rare congenital anomalies. Clinicians, imaging physicians, and pathologists should be aware of the likelihood of this benign lesion occurring. Performing other procedures such as adequate examination, biopsy or intraoperative freezing, can effectively prevent misdiagnosis and unnecessary orchiectomy, so as to retain the fertility of testis of the affected side of the patient. As for whether to remove the testis, more data is needed to accumulate experience and establish standards.

AUTHOR CONTRIBUTIONS

All of the work was done independently by the author.

FUNDING INFORMATION

None.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONFLICT INTEREST STATEMENT

The authors declare no conflicts of interest.

INFORMED CONSENT

Written informed consent was obtained from the patient and her next of kin to publish this report in accordance with the journal's patient consent policy.

ORCID

Ting Zhou https://orcid.org/0009-0005-2637-294X

REFERENCES

1. Huang G, Huang Y, Zeng L, Yuan M, Wu Y, Huang L. Continuous-type splenogonadal fusion: a case report. *Exp Ther Med*. 2017;13:2019-2021.
2. Zhu MB, Li XJ. One case report of splenogonadal fusion malformation. *J Harbin Med Uni*. 1986;(3):103-104.
3. Qadeer A, Quincey C, Gill K, Paddock M. Splenogonadal fusion: a radiologic-pathologic correlation and review of the literature. *Radiol Case Rep*. 2020;15(10):1817-1822.
4. Chen G, Wang X, Zhao Y, Zhu L, Tang D. Splenogonadal fusion: a case report and review of the literature. *BMC Urol*. 2021;21(1):16.
5. Kaya C, Koca O, Karaman IM, Radmayr C. Splenogonadal fusion in a 13-year-old boy with contralateral displaced intra-abdominal testis. *Urology*. 2010;75(1):173-175.
6. Abokrecha A, Almatrfi A. Discontinued Splenogonadal fusion and bilateral empty scrotum in an 18-month-old boy. *Eur J Pediatr Surg Rep*. 2017;5(1):e1-e3.
7. Steinmetz AP, Rappaport A, Nikolov G, Priel IE, Chamovitz DL, Dolev E. Splenogonadal fusion diagnosed by spleen scintigraphy. *J Nucl Med*. 1997;38(7):1153-1155.
8. Trottmann M, Marcon J, Mai V, et al. Characterization of splenogonadal fusion by contrast-enhanced ultrasound (CEUS) and elastography. *Ultraschall Med*. 2015;36(2):97-100.
9. Li YH. Preoperative detection of splenogonadal fusion by CT. *Surg Radiol Anat*. 2009;31(9):733-735.
10. Guney IB, Temiz A, Oksuzler M, Alkan M. Splenogonadal fusion in children: a rare entity mimicking inguinal tumor. *Clin Nucl Med*. 2020;45(6):453-454.
11. Preece J, Phillips S, Sorokin V, Herz D. Splenogonadal fusion in an 18-month-old. *J Pediatr Urol*. 2017;13(2):214-215.
12. Li WF, Luan MX, Ma Z, Chen YJ. Splenogonadal fusion: report of four cases and review of the literature. *Exp Ther Med*. 2013;6(3):816-818.

How to cite this article: Zhou T. A case of splenogonadal fusion in the left testis. *Clin Case Rep*. 2023;11:e7264. doi:10.1002/ccr3.7264