Egyptian Journal of Radiology and Nuclear Medicine

REVIEW

Open Access

Splenogonadal fusion- a great clinical masquerader: a case report and review of literature

T. Seetam Kumar[1], Pradosh Kumar Sarangi[2*], M. Sarthak Swarup[3] and Sonia Chhabra[4]

Abstract

Background Splenogonadal fusion (SGF) is a rare developmental choristoma in which ectopic splenic tissue is aberrantly attached to the gonads, mostly in the scrotum. It is a great clinical masquerader, and accurate preoperative diagnosis is often difficult due to the rarity of this entity. Many patients tend to undergo unnecessary surgical explorations and also orchiectomy. Accordingly, this article aims to review the latest literature regarding SGF and a description of the radiological features of this rare entity. Multiple systematic methods were used to find the latest publications on splenogonadal fusion by searching the Scopus, PubMed, and Google Scholar databases online since 2013. The latest comprehensive review of this rare entity was of 61 cases by Malik et al. in 2013. We reviewed all the cases of SGF reported in the literature in the last 10 years between 2013 and 2022 with an emphasis on diverse clinical presentations and radiologic findings. Along with this, a surgical and pathological proved case of SGF will be presented with emphasis on imaging findings. Splenogondal fusion is an uncommon differential diagnosis in patients with scrotal swelling. The current literature review showed the discontinuous type (63%) of SGF to be more common than the continuous (37%) type in contradiction to the previous literature review. Orchiectomy was done in 36% of cases as compared to 24% as described in the latest review by Malik et al.

Conclusions Knowledge of this entity along with familiarization with its imaging features among radiologists is essential for surgical prognostication and avoiding unnecessary orchiectomy. We suggest that the addition of colour Doppler and elastography to routine grey-scale ultrasound can increase diagnostic confidence. Subsequent cross-sectional imaging with magnetic resonance imaging (MRI) helps categorize the subtype and pre-operative planning.

Keywords Splenogonadal fusion, Cryptorchidism, Magnetic resonance imaging, Elasticity imaging techniques, Colour Doppler ultrasonography, Orchiectomy, Scrotum

Background

Splenogonadal fusion (SGF) is a rare benign congenital malformation in which the spleen is aberrantly connected to the gonads or rarely to the mesonephric derivatives such as vas deferens or epididymis [1, 2]. SGF usually presents as a left-sided scrotal swelling/ mass or cryptorchidism and is frequently misdiagnosed as testicular or epididymal tumours [2]. It is a great clinical masquerader, and accurate preoperative diagnosis is often difficult due to the rarity of this entity. Many patients tend to undergo unnecessary surgical explorations and also orchiectomy. Ultrasonography (USG) is often the first-line

*Correspondence:
Pradosh Kumar Sarangi
drpksarangi@gmail.com
[1] Department of Neuroimaging and Interventional Radiology, National Institute of Mental Health and Neurosciences (NIMHANS), Bengaluru, India
[2] Present Address: Department of Radiodiagnosis, All India Institute of Medical Sciences (AIIMS), Deoghar, Jharkhand 814152, India
[3] Department of Radiodiagnosis, VMMC and Safdarjung Hospital, New Delhi, India
[4] Department of Pathology, PT BD Sharma PGIMS, Rohtak, Haryana, India

imaging modality for the evaluation of a scrotal mass. Colour Doppler combined with ultrasound elastography can provide a valuable clue for the diagnosis. Cross-sectional imaging modalities like magnetic resonance imaging (MRI) or computed tomography (CT) further ascertain the diagnosis [3, 4]. We report a histopathologically proven case of SGF in an adolescent boy presented with scrotal swelling and intermittent pain. We reviewed all the cases of SGF reported in the literature in the last 10 years between 2013 and 2022 with an emphasis on diverse clinical presentations and radiologic findings.

Case presentation

A 12-year-old boy presented with progressive discomfort and swelling in the left scrotum for the last 8 months with associated lower abdominal pain radiating to the left testicle. At the onset, there was a transient symptomatic improvement with conservative management, however, the symptoms gradually worsened over the months with no relief from analgesics. The patient had no history of any urinary complaints or genitourinary trauma. Clinical examination revealed two discrete palpable masses in the left hemiscrotum, one almost indiscernible from the left testis and another near the head of the left epididymis. Both the testes could be palpated in the scrotal sac. No signs of scrotal inflammation were noted.

He was referred to our department for an ultrasound evaluation. The grey-scale ultrasound showed two to three well-defined lobulated oval hypoechoic (as compared to normal testis) solid masses in the upper pole of the left testis, with the largest one measuring approximately $35 \times 15 \times 13$ mm^3. On colour Doppler ultrasound, the mass lesions demonstrated higher vascularity in a radiating pattern compared to the normal testis. Strain elastography revealed higher stiffness in the solid mass lesions compared to the normal testicular tissue (Fig. 1). In consideration of the high stiffness and internal branching vascular pattern, the possibility of SGF was raised, and the patient was kept on follow-up to rule out the malignant testicular neoplastic lesion. A repeat ultrasound was performed after 2 months, which showed no interval increase in the lesion size. The patient underwent MR imaging before proceeding with surgical management. This revealed well-circumscribed soft tissue lesions in relation to the upper pole of the left testis. The lesions appear isointense on T1W images, and hypointense on T2W images (as compared to normal testicular parenchyma) with homogenous post-contrast enhancement typically more than the normal testis. Some degree of restricted diffusion was appreciated in the lesions (Fig. 2). No obvious soft tissue strand was noted extending from the lesion into the pelvic and abdominal cavities. With

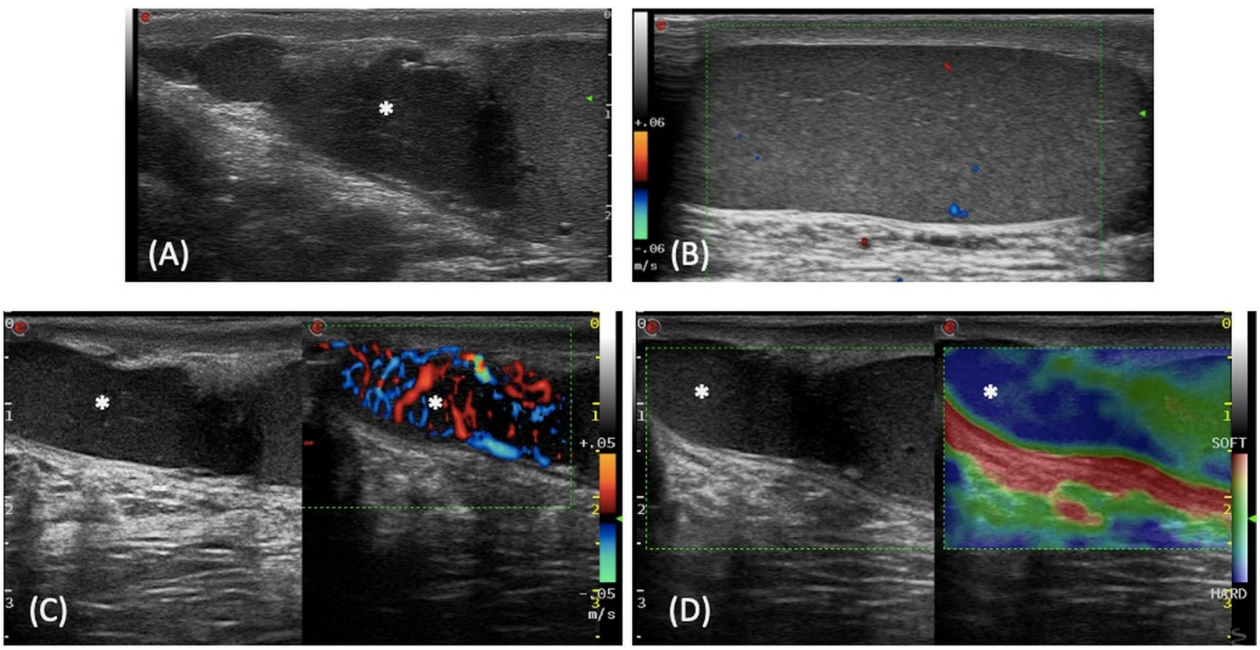

Fig. 1 A 12-year-old boy presented with progressive swelling in the left scrotum for the last 8 months with associated lower abdominal pain radiating to the left testicle. Sonographic images of the left scrotal region show a well-defined lobulated hypoechoic (as compared to testicular parenchyma) solid lesion (white asterisk in **A**) in the upper pole of the left testis with intense vascularity on colour Doppler map (in **C**) as compared to normal testicular vascularity (in **B**). Shear wave elastography map (**D**) demonstrates higher elastic modulus in the mass as compared to adjacent testicular tissue (as blue map–harder tissue)

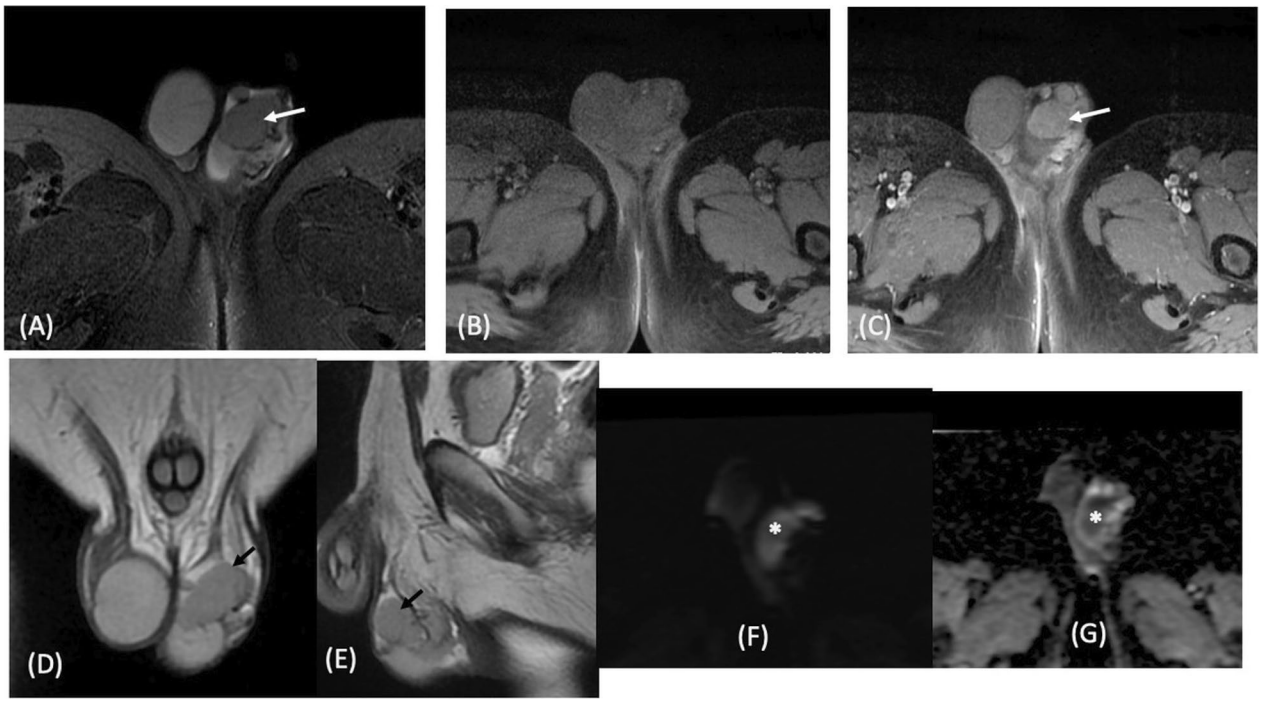

Fig. 2 T2W fat-saturated (**A**), T1W fat-saturated (**B**), and post-contrast T1W fat-saturated (**C**) axial magnetic resonance images reveal two to three (one enlarged) well-defined T2 hypointense (as compared to normal testis) lesions with homogenous enhancement on post contrast study (white arrow), typically more than the adjacent testicular tissue. Coronal (**D**) and sagittal (**E**) T2W images demonstrate the orientation of the lesion (black arrow) in relation to the upper pole of the left testis. The area of restricted diffusion is noted in the lesion (asterisk)—high signal on diffusion-weighted image (**F**) and signal drop in ADC map (**G**)

these clinical pictures and imaging morphology, we prospectively put a high possibility of ectopic splenic tissue in the scrotum as a rare syndrome of discontinuous-type SGF. However, a remote possibility of benign tumour of epididymis was given as differential.

Consequently, surgical exploration was done by a left-sided inguinal incision. Intraoperatively, three dark red, firm, fleshy lesions were identified. The first lesion was within the testicular parenchyma and was connected to two other lesions by a fibrous cord present in the inguinal canal, which terminated distal to the deep inguinal ring. All three lesions were encapsulated by thin white fibrous tissue. These three masses were removed completely with surgical margin clearance, and most of the left testis was preserved (Fig. 3). The post-operative period was uneventful. Histopathological examination of the resected mass lesions revealed a peripheral capsule with multiple cortico-medullary differentiation with white and red pulp. This was consistent with ectopic splenic tissue with no evidence of malignancy. Testicular seminiferous tubules were noted adjacent to the lesions (Fig. 3).

Multiple methods were used to find the current research publications on splenogonadal fusion. We started by searching the Scopus, PubMed, and Google Scholar databases online since 2013 with the combination of key terms including 'splenogonadal fusion', 'cryptorchidism', 'accessory spleen', 'scrotal swelling', and 'orchiectomy'. This search strategy recognized the abstracts of published articles, while other research articles were discovered manually from the citations. The initial search returned 427 records from all the databases from where 382 articles were found after removing duplicates. The search results were confined to journal articles written in English. We first reviewed the titles and abstracts for each of the 382 articles to determine their relevance. Following the criteria set out above, 289 studies were eliminated, and 93 studies were retained. These studies were then evaluated by going through the whole article. At least three authors independently reviewed each abstract. Minor disagreements were addressed in a meeting that resulted in an agreement, and finally, 63 articles were retained. The manuscript was drafted based on these final articles. Google spreadsheet was used to capture the data from different studies, and Microsoft Excel spreadsheet was used to tabulate the findings. One of the most comprehensive reviews of this rare entity in the recent literature was that of 61 cases by Malik et al. in 2013 [5]. Since then, from 2013 there were 62 articles with a total of 67 cases reported in the scientific

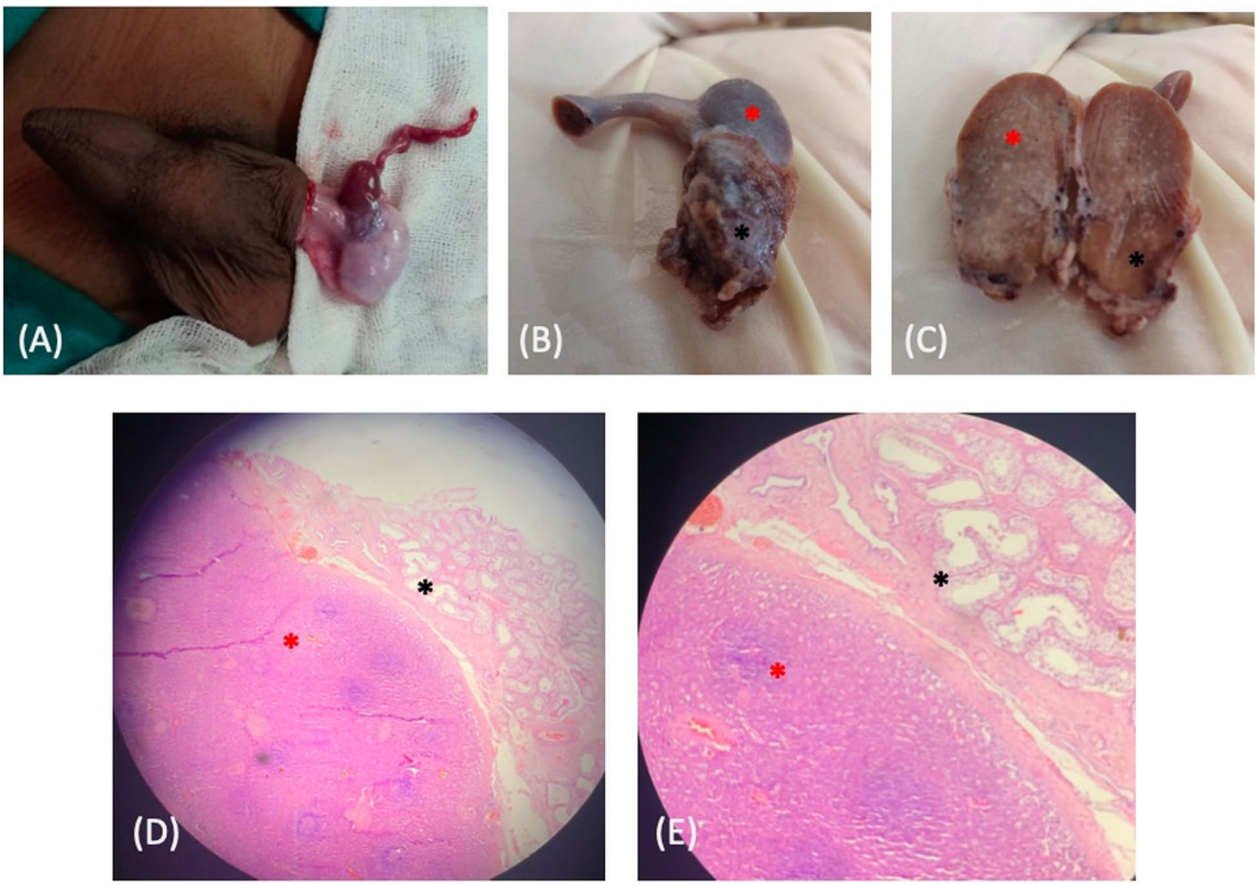

Fig. 3 Intraoperative surgical exploration image (**A**) confirms the radiologic findings. Gross resected specimen image (**B**) and cut section pathological image (**C**) show nutmeg appearance of sectional splenic tissue (red asterisk) and testicular tissue (black asterisk). Low (**D**)- and high (**E**)-power microscopic views of histopathological images demonstrate splenic tissue (red asterisk) and testicular tissue (black asterisk) in the lesion and confirm the diagnosis of splenogonadal fusion

literature till December 2022. We performed a comprehensive review of the literature on these reported cases of SGF in the last 10 years and summarized the details in Table 1 [3, 4, 6–65].

Results

Out of the 67 cases analysed, only one of them was female, while the rest were males. (Table 1). A considerable proportion of patients (71.2%) were observed to be below the age of 20 years (Table 2). The most prevalent clinical presentation among these cases was cryptorchidism, accounting for 28% of the occurrences, followed by painless scrotal swelling, which constituted 23.5% of the cases (Table 3). In terms of the types of SGF observed, the discontinuous type was found to be more prevalent, accounting for 64% of the cases, while the continuous type represented ~ 36% of the cases. Regarding the management approach, orchiectomy, which includes one case of partial orchiectomy, was conducted in 36% of the total cases.

Discussion

SGF is a rare congenital malformation first observed by German pathologist Bostroem in 1883. This can be associated with other congenital anomalies, such as cleft palate, cardiac defects, and micrognathia [2, 5]. Splenogonadal fusion was first categorized by Putschar and Manion in the year 1956 into two types, the continuous and the discontinuous form. The continuous form was reported to be slightly more common (58%) than the discontinuous type according to the previously reviewed literature [5]. However, in our 67 reviewed cases, only 24 cases (36%) have a continuous type. In the continuous type of SGF, a cord-like structure attaches the orthotropic spleen to the gonad. This cord-like structure can be fibrous, entirely splenic, or can have a beaded appearance containing intervening

Table 1 Details of the reported cases of SGF from 2013 till December 2022

S. No.	Published cases	Age (years), Sex (M/F)	Clinical presentation	Imaging done or not*	Orchiectomy	Type of SGF
1.	Li et al. [6]	6, M	Painless scrotal swelling	No	No	Discontinuous
		7, M	Inguinal hernia	No	yes	Continuous
		2, M	Bilateral cryptorchidism	No	Yes	Discontinuous
		12, M	Bilateral cryptorchidism	No	Yes	Discontinuous
2.	Mahalakshmi et al. [7]	6, M	Inguinal hernia	Yes	No	Continuous
3.	Ferrón et al. [8]	2, M	Painless left scrotal mass	Yes	No	Discontinuous
4.	Lui et al. [9]	6, M	Painless scrotal swelling	Yes	No	Continuous
5.	Fernandez et al. [10]	2, M	Inguinal hernia	Yes	No	Discontinuous
6.	Duhli et al. [11]	28, M	Painless scrotal swelling	No	Yes	Discontinuous
7.	Chiaramonte et al. [12]	12, M	Inguinal mass	Yes	No	Discontinuous
8.	Lakshmanan et al. [13]	6, M	Inguinal hernia	Yes	No	Continuous
9.	Sountoulides et al. [14]	31, M	Infertility with mass	Yes	No	Continuous
10.	Bal et al. [15]	20, M	Testicular mass	No	Yes	Discontinuous
11.	Kocher et al. [16]	35, M	Painless scrotal swelling	yes	yes	discontinuous
12.	Kumar et al. [17]	25, M	Infertility	Yes	Yes	Continuous
13.	Bosnali et al. [18]	7, M	Inguinal hernia	No	No	Continuous
14.	Foellings et al. [19]	16, M	Painless scrotal swelling	Yes	Yes	Discontinuous
15.	Shadpour et al. [20]	17, M	Bilateral cryptorchidism	No	Yes	Continuous
16.	Trottman et al. [21]	20, M	Painless scrotal swelling	Yes	No	Discontinuous
17.	Croxford et al. [22]	18, M	Testicular mass	Yes	Yes	Discontinuous
18.	Falaha et al. [23]	1.1, M	Inguinal mass	No	Yes	Discontinuous
19.	Uglialoro et al. [24]	45, M	Painless scrotal swelling	Yes	No	Discontinuous
20.	Jakkani et al. [25]	16, M	Bilateral cryptorchidism	Yes	Yes	Discontinuous
21.	Harris et al. [26]	55, M	Testicular mass	Yes	Yes	Discontinuous
22.	Babu et al. [27]	21, M	Inguinal hernia	No	No	Continuous
23.	Celik et al. [28]	1.5, M	Cryptorchidism	No	No	Continuous
24.	Preece et al. [29]	18, M	Cryptorchidism and retractile testis	Yes	No	Discontinuous
25.	Akama et al. [30]	76, M	Inguinal hernia	No	No	Discontinuous
26.	Abokrecha et al. [31]	1.5, M	Bilateral cryptorchidism	No	No	Discontinuous
27.	Huang et al. [32]	4, M	Bilateral cryptorchidism	Yes	No	Continuous
28.	Shakeri et al. [33]	4, M	Painless scrotal swelling	No	Yes	Discontinuous
29.	Karray et al. [34]	38, M	Testicular mass	No	Yes	Discontinuous
30.	Srinivasa Rao et al. [35]	6, M	Strangulated inguinal hernia	No	No	Discontinuous
31.	Zhou et al. [36]	9, M	Painless scrotal swelling	Yes	No	Discontinuous
32.	Grosu et al. [2]	53, M	Scrotal infection	Yes	No	Discontinuous
33.	Xiang X et al. [3]	0.5, M	Incarcerated inguinal hernia	Yes	No	Continuous
34.	Marwah et al. [37]	15, M	Painless scrotal swelling	Yes	No	Discontinuous
		11, M	Bilateral cryptorchidism	No	Yes	Discontinuous
35.	Chen et al. [38]	1.5, M	Cryptorchidism, hernia	No	No	Discontinuous
36.	Mann et al. [39]	22, M	Painful scrotal swelling	Yes	Yes	Discontinuous
37.	Navas et al. [40]	3, M	Inguinal swelling /hernia	No	No	Discontinuous
38.	Patil et al. [41]	14, M	Traumatic scrotal swelling	No	Yes	Discontinuous
39.	Hmidi et al. [42]	1.6, M	Testicular torsion-acute scrotum	Yes	No	Discontinuous
40.	Seager et al. [43]	25, M	Painless scrotal swelling	Yes	No	Discontinuous
41.	Guney et al. [44]	0.5, M	Bilateral cryptorchidism	Yes	No	Discontinuous
42.	Biben et al. [45]	0.5, M	Bilateral cryptorchidism	no	yes	discontinuous
43.	Xiao-xi et al. [46]	4, M	Painless mass for 3 years in the left scrotum	No	No	Discontinuous

Table 1 (continued)

S. No.	Published cases	Age (years), Sex (M/F)	Clinical presentation	Imaging done or not*	Orchiectomy	Type of SGF
44.	Bhutani et al. [47]	12, M	Painless scrotal swelling	Yes	No	Discontinuous
45.	Qadeer et al. [48]	29, M	Painless scrotal swelling	Yes	No	Discontinuous
46.	Jonuzi et al. [49]	13, M	Unilateral cryptorchidism	Yes	No	Discontinuous
47.	Kadouri et al. [50]	45, M	Painful scrotal swelling	Yes	No	Discontinuous
48.	Alsunbul et al. [51]	22, M	Left cryptorchidism	Yes	Yes	Continuous
49.	Burki et al. [52]	22 months, M	Severe penoscrotal hypospadias, bilateral impalpable testes	Yes	No	Continuous
50.	Viteri et al. [53]	2-year- and 9-month-old male	Moebius and Poland syndromes left inguinal hernia	No	No	Continuous
51.	Aoyagi et al. [54]	11, M	Bilateral absence of the testes together with hypospadias	Yes	No	Continuous
52.	Masmoudi et al. [55]	3.5, M	Left-sided vaginal hydrocele	Yes	No	Continuous
53.	Chen et al. [56]	8 months, M	Bilateral cryptorchidism	Yes	No	Continuous
54.	Kulkarni et al. [57]	16 months, M	Bilateral cryptorchidism	Yes	Yes	Continuous
55.	Durmuş et al. [58]	1.5, M	Bilateral undescended testis	Yes	No	Continuous
56.	Fadel et al. [59]	31, M	Rapidly growing left-sided testicular mass	Yes	Yes	Discontinuous
57.	Bicer et al. [60]	42, M	Left scrotal swelling-hydrocele	Yes	Yes	Discontinuous
58.	Alkukhun et al. [61]	15, M	Painless left testicular mass was found on routine physical exam	Yes	Yes, partial left orchiectomy	Discontinuous
59.	Kerkeni Y et al. [62]	8, M	Asymptomatic three-centimetre oval left scrotal mass mistaken for a spermatic cord cyst	No	No	Continuous
60.	Lazreg et al. [63]	5, M	Bilateral testicular cryptorchidism	yes	no	discontinuous
		9month, M	Follow-up case of an infant for the disorder of sex differentiation, at 9-month celioscopy -bilateral intra-abdominal testes	Yes	No	Continuous
61.	Guzman et al. [64]	Young adolescent female (F), Age not mentioned	1 day of left lower quadrant abdominal pain	Yes	No	Continuous
62.	Chen et al. [65]	5, M	Reversible mass in the left inguinal region, diagnosed with a left inguinal hernia before surgery	No	No	Continuous

M- Male, SGF- Splenogonadal fusion, * Imaging includes at least one of these modalities- ultrasonography, computed tomography, and magnetic resonance imaging

Table 2 Age at the time of diagnosis in reported cases of SGF

Age (in years)	Number of cases (%)
0–9	33 (50.0)
10–19	14 (21.2)
20–29	9 (13.6)
30–39	4 (6.0)
40–49	3 (4.5)
50–59	2 (3.0)
60–89	1 (1.5)
Total	66

Numbers are calculated from 66 cases as described in Table 1. One case is excluded as the exact age was not mentioned

fibrous and splenic nodules similar to a 'rosary bead' [20]. The discontinuous type of SGF usually presents as a firm scrotal mass, and no connection between the spleen and the ectopic splenic tissue can be established and is considered a special variant of accessory spleen. In this type, the ectopic splenic tissue is adherent to the testis or rarely other mesonephric derivatives like the epididymis. Most cases of continuous-type SGF have been associated with syndromic congenital anomalies, and the cases of discontinuous-type SGF have been associated with isolated inguinal swellings [6, 41].

The exact aetiology remains uncertain, however, numerous theories have been proposed to explain its etiopathogenesis and association with other

Table 3 Presenting clinical symptoms in reported cases of SGF

S. No.	Clinical presentation	Number of cases (%)
1.	Painless scrotal swelling	16 (23.5)
2.	Painful scrotal swelling	2 (3.0)
3.	Traumatic scrotal swelling	1 (1.5)
4.	Scrotal infection	1 (1.5)
5.	Testicular mass	6 (8.9)
6.	Inguinal hernia	10 (14.9)
7.	Incarcerated/ strangulated inguinal hernia	2 (3.0)
8.	Inguinal mass	2 (3.0)
9.	Cryptorchidism	19 (28.3)
10.	Cryptorchidism with hernia	1 (1.5)
11.	Cryptorchidism with retractile testis	1 (1.5)
12.	Infertility	2 (3.0)
13.	Acute scrotum/torsion	1 (1.5)
14.	Left hydrocele	2 (3.0)
15.	Left abdominal pain	1 (1.5)
Total		67

malformations. If there is inflammation of the peritoneal surfaces adjacent to the spleen and gonadal ridges during the development process, resulting in adhesion, this can initiate the fusion of the two organs before their descent. Between the 5th and 8th gestational weeks, the developing splenic tissue becomes fused to the gonad and is subsequently pulled caudally with the descent of the gonad into the pelvis during the 8th to 10th weeks of gestation. It is hypothesized that a teratogenic event occurring during the aforementioned gestational period causes limb deformities, micrognathia, and other congenital anomalies in addition to SGF [8, 11, 32, 57].

Although SGF can occur in both sexes, it is predominantly symptomatic in males. This disparity is mainly due to the superficial location of the testis, which allows for readily frequent palpation and detection vis a vis the abdominal location of ovaries in females. SGF is commonly diagnosed in females as an incidental finding during surgery for a different diagnosis or on autopsy [5, 41]. It is typically noted on the left side in up to 97% of cases [5]. Even in our review as well, all cases were males with SGF on the left side with the exception of only one case presented on the right as reported by Marwah et al. [37]. Guzman et al. [64] reported a case of symptomatic SGF with splenic torsion in an adolescent female who underwent operative detorsion and partial splenectomy. SGF mostly presents before 20 years of age, with a reported value of 68% in the review done by Malik et al. [5]. In our review as well, 47 out of 67 cases (71.2%) presented below 20 years of age. Clinical presentation varies from simple inguino-scrotal swelling to strangulated inguinal hernia. Varied clinical presentations are tabulated in Table 3. A wide variety of clinical presentations makes it a great clinical masquerader. Cryptorchidism was the most common presentation (28%) in our reviewed cases, followed by painless scrotal swelling (23.5%).

SGF is mostly a retrospective diagnosis based on histological features of a surgical or autopsy specimen, and the radiologic literature is limited. This stresses the need for a high index of suspicion and awareness for a potential SGF preventing unnecessary orchiectomies. Previously before the era of cross-sectional imaging and sonography, technetium-99m sulphur colloid scan was the only diagnostic clue towards a diagnosis of SGF [5, 29]. A definitive diagnosis of SGF may not be made solely based on sonographic findings. However, a well-defined lobulated capsulated homogenous hypoechoic mass close to the testis with a branching/ radiating vascular pattern resembling the vascular pattern of normal splenic tissue in a patient with left-sided scrotal swelling along with normal tumour markers for testicular malignancies should raise the suspicion for SGF. A disorganized branching criss-cross vascular pattern from the testicular mass lesion on colour Doppler may indicate testicular malignancy [66]. Microflow imaging (MFI) is a recently developed ultrasound technique by Philips (Bothell, WA, USA, a similar technique known as superb microvascular imaging [SMI]; Toshiba Medical Systems, Tokyo, Japan) different than conventional power Doppler technique, which is very sensitive for detecting slow and weak blood flow from tissue with high spatial resolution [67]. In MFI, the artefact reduction technique is applied to separate slow flow signals from tissue motion artefacts without the use of contrast agents. Studies on the utility of MFI in breast, thyroid, testicular, and hepatic lesions are available. However, to the best of our knowledge, no literature on the application of MFI in SGF is available. This technique may be useful in detecting the pattern of tissue vascularity in SGF and testicular tumours. With the advent of ultrasound elastography, testicular tissue can be distinctly differentiated from other tissue like the spleen having higher stiffness [3, 21]. Contrast-enhanced ultrasonography (CEUS) can facilitate the evaluation process as the ectopic splenic tissue shows avid atrial phase enhancement. In the continuous type of SGF, computed tomography (CT) can demonstrate the connection with the spleen and delineate vasculature originating from the splenic hilum [25]. MR evaluation of the pelvis and abdomen helps better delineation of SGF, which can differentiate between continuous from discontinuous types owing to the better soft tissue spatial resolution. Signal intensities and enhancement patterns of the ectopic

splenic tissue in SGF will be similar to the normal orthotopic splenic tissue. MR imaging is also useful to differentiate SGF from other malignancies as ectopic tissue usually adheres to testicular tissue with maintained fat planes. Among our 67 reviewed cases, many cases do not have any radiologic data, and MR imaging is available in less than 10 cases [3, 7, 9, 17, 19, 51, 56, 58, 63]. Hence, we emphasize using radiological modalities in the diagnostic evaluation of this entity and their accurate interpretation for correct clinical guidance and management.

Tissue diagnosis is the gold standard in establishing the diagnosis of this entity. The ectopic splenic tissue in SGF shows the normal splenic architecture on histopathology and demonstrates capsule, cortex, and medulla with red pulp and white pulp with sinusoids. Ectopic splenic tissue may sometimes show thrombosis, calcification, fat degradations, and hemosiderin depositions. Few cases had been reported with an intermingling of splenic and gonadal tissue histologically [2, 5]

Unnecessary interventions can be avoided if a confident diagnosis using ultrasound and cross-sectional imaging modalities is made. A testis-preserving surgical procedure should be planned in a patient with SGF if however, performed [5, 41, 51, 57]. There are few cases in the literature reporting an association between SGF and testicular malignancy [64]. SGF associated with cryptorchidism increases the risk of malignancy and hence orchiectomy is a better option [41]. A complete orchiectomy is often not required because the fused splenic tissue can be dissected safely off the tunica albuginea. Even if SGF is almost always a benign condition, the cord attached to the ectopic splenic tissue is liable to undergo torsion as well as associated with congenital hernias as described in a few case reports [3, 7]. Orchiectomy was done in 24 cases out of 67 male patients (36%) in our review, and it was about 37% and 24% in the cases reviewed by Carragher [1] and Malik et al. [5], respectively. The symptomatic adolescent female reported by Guzman et al. [64] underwent laparoscopic detorsion and partial splenectomy with preservation of the left ovary.

Conclusions

Splenogondal fusion is an uncommon differential diagnosis in patients with scrotal swelling. Its typical imaging findings should raise suspicion. We suggest that the addition of colour Doppler and elastography to routine grey-scale ultrasound can increase diagnostic confidence. Subsequent cross-sectional imaging with MR helps categorize the subtype and pre-operative planning. In our case, unnecessary radical orchiectomy was avoided as a differential diagnosis of SGF was prospectively suggested based on imaging findings. Knowledge of this entity along with familiarization with its imaging features among radiologists is essential for surgical prognostication and avoiding unnecessary orchiectomy.

Acknowledgements
Not applicable

Author contributions
TSK, PKS, and MSS conceived of the need of the study. TSK and SC did the imaging and pathological evaluation of the case, respectively. All authors contributed to the literature search. TSK drafted the first article. All authors critically reviewed further revisions. All authors have read and approved the final manuscript.

Funding
No funding was obtained for this study.

Availability of data and materials
Not applicable.

Declarations

Ethics approval and consent to participate
Ethical clearance number or waiver is not required for review article from our institute. This article followed all ethical standards for carrying out research.

Consent for publication
Written consent has been obtained from the parents of the patient to include their data and accompanying images in this publication.

Competing interests
The authors declare they have no competing interests.

Received: 12 June 2023 Accepted: 30 August 2023
Published online: 10 October 2023

References

1. Imperial SL, Sidhu JS (2002) Nonseminomatous germ cell tumor arising in splenogonadal fusion. Arch Pathol Lab Med 126(10):1222–1225. https://doi.org/10.5858/2002-126-1222-NGCTAI
2. Lin CS, Lazarowicz JL, Allan RW et al (2010) Splenogonadal fusion. J Urol 184(1):332–333. https://doi.org/10.1016/j.juro.2010.04.013
3. Grosu S, Rübenthaler J, Knösel T et al (2019) Splenogonadal fusion evaluation using contrast-enhanced ultrasound and elastography—a case report. Med Ultrason 21(3):356. https://doi.org/10.11152/mu-1897
4. Xiang X, Jiang Y, Liu JX et al (2019) A rare content of congenital inguinal hernia: a case report of splenogonadal fusion. BMC Pediatr 19(1):422. https://doi.org/10.1186/s12887-019-1807-x
5. Malik RD, Liu DB (2013) Splenogonadal fusion: an unusual case of an acute scrotum. Rev Urol 15(4):197–201
6. Li WF, Luan MX, Ma Z et al (2013) Splenogonadal fusion: report of four cases and review of the literature. Exp Ther Med 6(3):816–818. https://doi.org/10.3892/etm.2013.1207
7. Mahalakshmi VN, Barathi SD (2013) An unusual content in a congenital hernia—complete spleno–gonadal fusion band. Indian J Surg 75(S1):110–112. https://doi.org/10.1007/s12262-012-0409-8
8. Ferrón SA, Arce JD (2013) Discontinuous splenogonadal fusion: new sonographic findings. Pediatr Radiol 43(12):1652–1655. https://doi.org/10.1007/s00247-013-2730-1
9. Liu W, Wu R, Guo Z (2013) The diagnosis and management of continuous splenogonadal fusion in a 6-year-old boy. Int Urol Nephrol 45(1):21–24. https://doi.org/10.1007/s11255-012-0349-z
10. Fernandez Atuan R, Burgues Prades P, Gonzalez Esgueda A et al (2013) Splenogonadal fusion: case report and review of literature. J Pediatr Surg Case Repo 1(10):375–377. https://doi.org/10.1016/j.epsc.2013.10.004

11. Duhli N, Venkatramani V, Panda A et al (2014) Splenogonadal fusion: pathological features of a rare scrotal mass. Indian J Pathol Microbiol 56(4):474–476. https://doi.org/10.4103/0377-4929.125387
12. Chiaramonte C, Siracusa F, Li Voti G (2014) Splenogonadal fusion: a genetic disorder—report of a case and review of the literature. Urol Case Repo 2(2):67–69. https://doi.org/10.1016/j.eucr.2014.01.003
13. Lakshmanan PM, Reddy AK, Nutakki A (2014) A surprising content of congenital hernia: complete splenogonadal fusion band. BMJ Case Repo 2014:bcr2014203640. https://doi.org/10.1136/bcr-2014-203640
14. Sountoulides P, Neri F, Bellocci R et al (2014) Splenogonadal fusion mimicking a testis tumor. J Postgrad Med 60(2):202. https://doi.org/10.4103/0022-3859.132350
15. Bal K, Ermete M, Balci U et al (2014) Splenogonadal fusion: a very rare congenital anomaly in the differential diagnosis of a testicular mass. Turk J Urol 40(1):62–64. https://doi.org/10.5152/tud.2014.90757
16. Kocher NJ, Tomaszewski JJ, Parsons RB et al (2014) Splenogonadal fusion: a rare etiology of solid testicular mass. Urology 83(1):1–2. https://doi.org/10.1016/j.urology.2013.09.019
17. Kumar S, Jayant K, Agrawal S et al (2014) A rare case of continuous type splenogonadal fusion in a young male with primary infertility. Case Rep Urol 2014:796761. https://doi.org/10.1155/2014/796761
18. Bosnalı O, Cici İ, Moralıoğlu S et al (2014) Continuous-type splenogonadal fusion: report of a rare case. Turk J Pediatr 56(6):680–683
19. Foellings K, Baker S, Walker D (2014) Sonographic and magnetic resonance imaging characteristics associated with Splenogonadal fusion. J Diagnost Med Sonography 30(5):256–260
20. Shadpour P, Rezaimehr B (2015) "Rosary of Testes": Splenogonadal fusion in association with bilateral abdominal testes presenting as polyorchidism. Case Rep Urol 2015:317189. https://doi.org/10.1155/2015/317189
21. Trottmann M, Marcon J, Mai V et al (2015) Characterization of splenogonadal fusion by contrast-enhanced ultrasound (CEUS) and elastography. Ultraschall Med 36(2):97–100. https://doi.org/10.1055/s-0034-1369759
22. Croxford WC, Pfistermuller KLM, Scott F et al (2015) Splenogonadal Fusion presenting clinically and radiologically as a seminoma. Urol Case Repo 3(6):204–205. https://doi.org/10.1016/j.eucr.2015.06.007
23. Falah KM, Hama MA, Yasseen HA et al (2015) Splenogonadal fusion: a rare case report. Annals Pathol Lab Med 02(01):C39-41
24. Uglialoro AD, Goltzman ME, Niazi M et al (2016) Splenogonadal fusion presenting as an asymptomatic testicular mass. Urology 97:1–4. https://doi.org/10.1016/j.urology.2016.07.019
25. Jakkani R, Alhajri F, Alteriki A et al (2016) Discontinuous splenogonadal fusion diagnosed on computed tomography. Indian J Radiol Imaging 26(4):506. https://doi.org/10.4103/0971-3026.195776
26. Harris AM (2016) Splenogonadal fusion: a rare benign testicular mass in a 55-year-old male. Urol Case Repo 9:41–42. https://doi.org/10.1016/j.eucr.2016.08.013
27. Babu N, Kumar P (2016) Rare content in congenital inguinal hernia: Splenogonadal fusion band. Int Surg J 3(1):394–396. https://doi.org/10.18203/2349-2902.isj20160267
28. Celik A, Tiryaki S, Darcan S et al (2016) Splenogonadal fusion-limb deformity syndrome: a rare but important cause of undescended testis. World J Pediatr 12(2):246–248. https://doi.org/10.1007/s12519-015-0046-y
29. Preece J, Phillips S, Sorokin V et al (2017) Splenogonadal fusion in an 18-month-old. J Pediatr Urol 13(2):214–215. https://doi.org/10.1016/j.jpurol.2016.06.005
30. Akama Y, Shimanuki K, Asahi S et al (2017) Incidentally detected splenogonadal fusion in a laparoscopic transabdominal preperitoneal hernia repair operation: a case report. Inter J Surg Case Repo 35:29–32. https://doi.org/10.1016/j.ijscr.2017.04.004
31. Abokrecha A, Almatrfi A (2017) Discontinued splenogonadal fusion and bilateral empty scrotum in an 18-month-old boy. European J Pediatr Surg Rep 5(1):e1–e3. https://doi.org/10.1055/s-0037-1598624
32. Huang G, Huang Y, Zeng L et al (2017) Continuous-type splenogonadal fusion: a case report. Exp Ther Med 13(5):2019–2021. https://doi.org/10.3892/etm.2017.4198
33. Shakeri A, Shakeri A, Rasolmali R et al (2018) A case of splenogonadal fusion accompanied by accessory spleen in a 4-year-old boy. Urol Ann 10(4):406–408. https://doi.org/10.4103/ua.ua_194_17
34. Karray O, Oueslati A, Chakroun M et al (2018) Splenogonadal fusion—a rare cause of scrotal swelling: a case report. J Med Case Reports 12(1):172. https://doi.org/10.1186/s13256-018-1712-1
35. Srinivasa Rao RC, Radhakrishna V, Rao N et al (2018) Torsion of a splenule in a case of splenogonadal fusion mimicking a strangulated inguinal hernia. J Indian Assoc Pediatr Surg 23(2):100–102. https://doi.org/10.4103/jiaps.JIAPS_191_17
36. Zhou L, Muthucumaru M, Stunden R et al (2018) Splenogonadal fusion: a rare scrotal mass in a 9-year-old boy. ANZ J of Surg 88(1):81–82. https://doi.org/10.1111/ans.13250
37. Marwah N, Bhutani N, Kalra R et al (2019) Accessory spleen within the scrotum. J Pediatr Surg Case Repo 42:19–23. https://doi.org/10.1016/j.epsc.2018.12.009
38. Chen CJ, Kavoussi N, Jacobs MA (2019) Splenogonadal fusion: a rare finding during routine orchiopexy. Urol Case Repo 27:100904. https://doi.org/10.1016/j.eucr.2019.100904
39. Mann JA, Ritchie E (2019) Splenogonadal fusion: a case of two lesions. Urol Case Repo 24:100878. https://doi.org/10.1016/j.eucr.2019.100878
40. Navas FJ, Castro M, Arredondo GP (2019) Splenogonadal discontinuous fusion as a transoperative finding in inguinal hernia surgery in children (Granado's Hernia). Clinical Case Mathews J Case Rep 4(1):1–4. https://doi.org/10.30654/MJCR.10044
41. Patil SV, Patil PS, Sharma VD et al (2020) Splenogonadal fusion—a rare anomaly. J Indian Assoc Pediatr Surg 25(2):115–117. https://doi.org/10.4103/jiaps.jiaps_6_19
42. Hmidi N, Makhlouf D, Laamiri R et al (2020) Splenogonadal fusion discovered by testicular torsion. Iranian J Pediatr Surg 6(1):50–53. https://doi.org/10.22037/irjps.v6i1.31705
43. Seager MJ, Alexander S, Muneer A et al (2020) Splenogonadal fusion—a rare paratesticular lesion and how to recognise it on ultrasound. Ultrasound 28(1):54–57. https://doi.org/10.1177/1742271X19876085
44. Guney IB, Temiz A, Oksuzler M et al (2020) Splenogonadal fusion in children: a rare entity mimicking inguinal tumor. Clin Nucl Med 45(6):453–454. https://doi.org/10.1097/RLU.0000000000002989
45. Biben E, Pugach J (2020) Splenogondal fusion with accessory spleens identified with laparoscopy. Cureus 12(7):9017. https://doi.org/10.7759/cureus.9017
46. Xiao-xi LI, Hua XI, Yi-chen HU et al (2020) Continuous splenogonadal fusion: a case report and review of literature. J Shanghai Jiaotong Univ (Med Sci) 12:1002–1004
47. Bhutani N, Kajal P, Sangwan V (2020) Splenogonadal fusion. J Pediatr Surg Case Rep 59:101514. https://doi.org/10.1016/j.epsc.2020.101514
48. Qadeer A, Quincey C, Gill K et al (2020) Splenogonadal fusion—a radiologic-pathologic correlation and review of the literature. Radiol Case Rep 15(10):1817–1822. https://doi.org/10.1016/j.radcr.2020.07.033
49. Jonuzi A, Popović N, Zvizdić Z et al (2020) A rare case of discontinuous type splenogonadal fusion in a 13 year old boy: a case report and review of literature. Int Surg J 7(10):3438–3441. https://doi.org/10.18203/2349-2902.isj20204150
50. Kadouri Y, Carnicelli D, Sayegh HE et al (2020) Pathogenesis, Diagnosis, and management of splenogonadal fusion: a literature review. Case Rep Urol 2020:8876219. https://doi.org/10.1155/2020/8876219
51. Alsunbul A, Alsuhaibani S, Al Ali H et al (2020) Splenogonadal fusion: a rare case report and literature review. Urol Case Rep 33:101307. https://doi.org/10.1016/j.eucr.2020.101307
52. Burki T, Kanfar S, Jamalalail Y et al (2021) Continuous type of intra-abdominal splenogonadal fusion: a case report with review of literature. Pediatr Urol Case Rep 8(4):79–82. https://doi.org/10.14534/j-pucr.2021267554
53. Viteri MC, Farina DA, Mayorga HM et al (2021) Splenogonadal fusion associated with Moebius and Poland syndromes: first case reported. Cir Pediatr 34(4):219–222
54. Aoyagi C, Matsuoka H, Gunge N et al (2021) Successful fowler-stephens orchiopexy for an abdominal testis associated with continuous type of splenogondal fusion: a case report. Urology 156:e137–e140. https://doi.org/10.1016/j.urology.2021.03.040
55. Masmoudi M, Echaieb A, Georges P et al (2021) Splenogonadal fusion presenting as a multiple nodular paratesticular masses: a case report. EC Paediatr 10:89–94. https://doi.org/10.31080/ecpe.2021.10.01050
56. Chen G, Wang X, Zhao Y et al (2021) Splenogonadal fusion: a case report and review of the literature. BMC Urol 21:1–10. https://doi.org/10.1186/s12894-021-00781-z
57. Kulkarni AA, Kanbur AS, Kamat NV et al (2022) Lumbar cryptorchidism with splenogonadal fusion: the odd couple. J Gastrointest Abdom Radiol 5(01):049–054. https://doi.org/10.1055/s-0041-1740477

58. Durmuş G, Boybeyi-Turer O, Özcan HN et al (2022) Continuous hepatogonodal and splenogonogal fusion: a rare cause of bilateral intra-abdominal testis in an 18-month-old boy. European J Pediatr Surg Rep 10(1):e80–e83. https://doi.org/10.1055/s-0042-1747671
59. Fadel MG, Walters U, Smith A et al (2022) Splenogonadal fusion: aiding detection and avoiding radical orchidectomy. Ann R Coll Surg Engl 104(2):e32–e34. https://doi.org/10.1308/rcsann.2021.0080
60. Bicer H, Gur A, Bayraktar C et al (2022) Splenogonadal fusion anomaly associated with hydrocele in an adult patient—a rare cause of scrotal mass. Grand J Urol 2(2):76–79. https://doi.org/10.5505/GJU.2022.14622
61. Alkukhun L, Ionno A, Stanton M et al (2022) Splenogonadal fusion presenting as a testicular mass, a case report and literature review. Radiol Case Rep 17(6):2191–2194. https://doi.org/10.1016/j.radcr.2022.01.065
62. Kerkeni Y, Hannachi W, Zouaoui A et al (2022) Splenogonadal fusion misdiagnosed as spermatic cord cyst. Tunis Med 100(10):730–732
63. Lazreg KB, Helal I, Khanchel F et al (2022) Splenogonadal fusion: a case series of two challenging diagnoses. Clin Case Rep 10(8):e6250. https://doi.org/10.1002/ccr3.6250
64. Guzman J, Wieck MM (2022) Symptomatic splenogonadal fusion with splenic torsion in an adolescent female. J Pediatr Surg Case Rep 86:102452
65. Chen J, Li M (2023) Continuous splenogonadal fusion: a rare case report. Asian J Surg 46(4):1848–1849. https://doi.org/10.1016/j.asjsur.2022.10.065
66. Bushby LH, Sriprasad S, Hopster D et al (2001) High frequency color Doppler US of focal testicular lesions: crossing vessels (criss-cross pattern) identifies primary malignant tumors. Radiology 221:449
67. Microflow Imaging | Philips Healthcare [Internet]. Philips. [cited 2023 Mar 26]. Available from: https://www.usa.philips.com/healthcare/resources/feature-detail/ultrasound-microflow-imaging.

Publisher's Note