

Splenogonadal Fusion Anomaly Associated with Hydrocele in an Adult Patient- A Rare Cause of Scrotal Mass

Erişkin Bir Hastada Hidrozel ile İlişkili Splenogonadal Füzyon Anomalisi- Skrotal Kitlenin Nadir Bir Nedeni

Huseyin Bicer , Ahmet Gur , Cemil Bayraktar , Mert Ali Karadag 

Department of Urology, University of Health Sciences, Kayseri City Hospital, Kayseri, Turkey

Cite as: Bicer H, Gur A, Bayraktar C, Karadag MA. Splenogonadal fusion anomaly associated with hydrocele in an adult patient- a rare cause of scrotal mass. Grand J Urol 2022;2(2):76-9.

Submission date: 31 March 2022

Acceptance date: 05 May 2022

Online First: 09 May 2022

Publication date: 20 May 2022

Corresponding Author: Huseyin Bicer / University of Health Sciences, Kayseri City Hospital, Department of Urology, Kayseri, Turkey / drhuseyinbicer@yahoo.com / ORCID ID: 0000-0001-7703-3618

Abstract

Splenogonadal fusion (SGF) is a very rarely seen congenital anomaly localized usually in the left testis and mimics a testicular tumor. There are two subtypes of SGF, as continuous and discontinuous SGF. Continuous SGF can usually be detected in childhood. The less common discontinuous SGF may not be detected until adulthood, and may be mistaken for testicular tumor and cause unnecessary orchiectomies. In this case report, we aimed to present a patient who underwent orchiectomy due to a left testicular mass associated with hydrocele and was found to have discontinuous SGF in his histopathological evaluation.

Keywords: congenital malformation, radical orchiectomy, splenogonadal fusion

Öz

Splenogonadal füzyon (SGF), genellikle sol testiste, testis tümörünü taklit eden ve oldukça nadir görülen doğumsal bir anomalidir. SGF'nin devamlı (sürekli) ve devamlı olmayan (devamsız, süreksiz) olmak üzere iki alt tipi vardır. Devamlı SGF genellikle çocukluk çağında saptanabilmektedir. Daha nadir görülen devamsız SGF ise yetişkinlik dönemine kadar saptanamayabilir. Testis tümörü zannedilerek gereksiz orşiektomilere neden olabilir. Bu olgu sunumunda, hidrozel ile birliktelik gösteren sol testis kitlesi nedeniyle orşiektomi yapılan ve histopatolojik değerlendirmede devamsız SGF saptanan hastanın sunulması amaçlanmıştır.

Anahtar kelimeler: konjenital malformasyon, radikal orşiektomi, splenogonadal füzyon

Introduction

Splenogonadal fusion (SGF) is one of the very rarely seen fusion anomalies [1]. This anomaly occurs during the splenogonadal convergence that develops between the 5th-8th weeks of the embryonic life [2]. SGF was first described by Bostroem et al. in 1883 [3]. Its continuous and discontinuous types were reported in 1956 [4]. More than 150 cases of SGF have been reported to date, and only 4 cases have been associated with malignancy. For this reason, they are generally considered as benign lesions [1,5].

In continuous SGF, the spleen tissue is continuous on the spermatic cord. Although the discontinuous type is less common, spleen tissue is not observed on the spermatic cord. In these cases, a mass of ectopic spleen or accessory spleen tissue is usually detected on the testis [6].

SGF is usually diagnosed in childhood. However, it is rarely detected until adulthood, and it can be confused with malignant testicular tumors and cause unnecessary orchiectomies [7,8]. Indeed, approximately 35-40% of these patients can only be detected after orchiectomy [1].

In this case report, we aimed to present an adult case who underwent radical orchiectomy due to the suspicion of testicular cancer which was revealed to be discontinuous SGF later on.

Case

A 42-year-old male patient presented with the complaint of left scrotal swelling. On physical examination, an appearance compatible with a left hydrocele was observed. The patient who was married and had 2 children, had no history of previous scrotal surgery and his scrotal swelling had been present for about 6 months. In the scrotal ultrasonography (SUSG) performed with the preliminary diagnosis of hydrocele, a 10x9 mm- hypoechoic and homogeneous testicular mass (seminoma?) was reported in the left testicular apex together with a left 67x25 mm hydrocele sac. The patient's total alpha-fetoprotein (AFP) and beta-human chorionic gonadotropin (β -HCG) levels were within normal limits.

Since estimated glomerular filtration rate (GFR) was at the limit (e-GFR: 58 ml/min/1.73m²), contrast-enhanced radiological examination was not applied to the patient at first. Sperm freezing was recommended to the patient because of the possible future pregnancy request, but the patient did not accept the sperm freezing procedure he did not want to have a child. Left inguinal orchiectomy was planned for the patient. During surgery, after the clamp placed around the spermatic cord, the hydrocele sac was opened and orchiectomy was completed after the solid mass near the testicular apex was seen. The patient, whose general condition was good with stable vital signs, was discharged on the postoperative 1st day with the histopathology result, and control visit was recommended. At the 10th day follow-up visit, histopathological evaluation revealed a mass at the apex of the left testis defined as "splenogonadal fusion-ectopic scrotal spleen" (**Figure 1**). No additional treatment or intervention was considered for the patient who was included in the standard follow-up protocol.

Discussion

Discontinuous splenogonadal fusion (SGF) anomalies are confused with testicular cancers and are usually diagnosed as a result of histopathological evaluation performed after radical orchiectomy [1]. Similarly, in our case, testicular cancer was suspected as a result of SUSG performed for another reason, but histopathological evaluation revealed the presence of a mass consisting of a benign spleen tissue due to SGF anomaly.

Information about SGF in the literature is related to case presentations generally detected in childhood [6-7]. Rarely, cases of discontinuous type SGF detected in adults have also been reported [1,5,8].

Karray et al. reported that discontinuous type SGF was detected in a 38-year-old male patient who underwent left radical orchiectomy with the suspicion of left testicular upper pole tumor, similar to our case [1]. The researchers argued that if SGF could be predicted beforehand, testicular sparing approach would be appropriate for their patient. Our patient had no desire for fertility. However, testicular preservation may be important, especially in young men with a desire for fertility.

The majority of testicular cancers are diagnosed when the patient notices a mass in the unilateral testis or when this mass is detected incidentally by SUSG. Contrast-enhanced computed tomography ceCT is very sensitive in staging testicular cancers. Professional guidelines recommend preoperative ceCT scans for staging, but they also indicate that this procedure can sometimes be delayed until the result of histopathological evaluation is obtained [9]. Due to the borderline GFR values in our case, imaging procedures for staging were postponed until after the results of histopathological evaluation were obtained. Contrast-enhanced magnetic resonance imaging ceMRI is more sensitive than SUSG in the diagnosis of intrascrotal masses. However, performing ceMRI procedures routinely is not recommended due to its higher cost.

Instead, it is considered more appropriate to be used in cases where an accurate diagnosis cannot be made with SUSG [9]. In our case, since the tumor was very small and the mass could not be palpated due to the presence of hydrocele, ceMRI might be an appropriate procedure. Thus, we could refrain from performing orchiectomy considering the benign nature of the mass lesion. However, the patient had borderline, GFR values which made us hesitate to perform ceMRI.

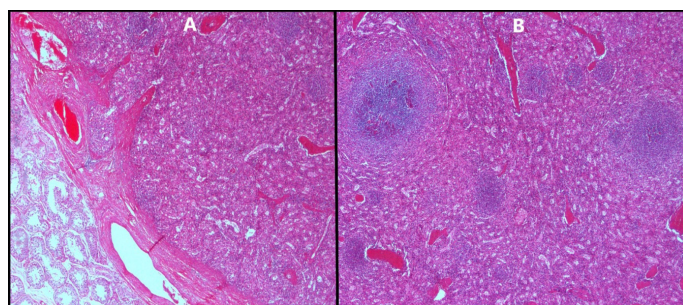


Figure 1a. Ectopic spleen tissue in testis. Seminiferous tubuli in the left, and spleen tissue separated by a clear border in the right testis (H&E x40). **1b.** White and red pulp of the spleen (H&E x40)

AFP and β -HCG are the most commonly used tumor markers in the diagnosis of testicular tumors [10]. Seminoma was suspected in the preoperative SUSG evaluation of our case. However, β -HCG positivity is reported in only 30% of pure seminomas, whereas AFP is usually within normal limits [11]. However, since the tumor markers were within normal limits, we could not make a precisely accurate diagnosis.

It has been reported that in patients with a small tumor size, negative tumor markers, a single testis, and a desire for fertility, the option of testicular-sparing surgery may be offered to the patient [12,13]. However, frozen section studies are generally not recommended due to the higher rates of inconsistencies between the frozen section results and the final histopathology [14]. Testis-sparing surgery was not recommended for our case because the other testis was completely normal, the patient had 2 children, and no desire for fertility.

It is known that SGF is frequently associated with cryptorchidism [8]. Lopes et al. reported that SGF was detected in a 36-year-old infertile patient with a history of bilateral cryptorchidism [5]. In our case, unlike this case, cryptorchidism and infertility were not accompanied by SGF, but our patient had a left-sided hydrocele. Hydrocele may be a complication of pathologies such as epididymitis, epididymo-orchitis, testicular tumor, or it may coexist incidentally with testicular tumors. Hydrocele may interfere with correct palpation of the testis and tumors may be overlooked [15]. In our case, the small size of the tumor and the presence of hydrocele prevented testicular palpation in genital examination and prevented the detection of the tumor. However, the presence of tumor was detected by SUSG. To the best of our knowledge, this is the first case of SGF with accompanying hydrocele in the literature.

In conclusion, discontinuous SGF anomalies, which are very rare, can be confused with testicular tumors and cause unnecessary orchiectomies. It is very difficult to detect these anomalies in the preoperative or intraoperative period. However, in case of doubt, the diagnosis can be confirmed by ceMRI. and Tc-99m sulfur colloid liver-spleen scanning, which can be performed preoperatively [16]. Measurement of GFR is important for the decision to perform ceMRI. because renal clearance of gadolinium is markedly prolonged in patients with moderate (GFR: 30-60 ml/min) and severe renal impairment (GFR: 15-30 ml/min) [17]. These conditions may restrict the use of preoperative ceMRI. However, unnecessary orchiectomies can be prevented with such preoperative examinations and testicular sparing surgeries or conservative follow-up protocols may be applied. In addition, they contribute to the preservation of fertility.

Ethics Committee Approval: N / A.

Informed Consent: An informed consent was obtained from the patient.

Publication: The results of the study were not published in full or in part in form of abstracts.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Any contribution was not made by any individual not listed as an author. Concept – H.B.; Design – H.B.; Supervision – C.B.; Resources – A.G.; Materials –

H.B.; Data Collection and/or Processing – C.B.; Analysis and/or Interpretation – H.B.; Literature Search – C.B.; Writing Manuscript – H.B.; Critical Review – M.A.K.

Conflict of Interest: The authors declare that they have no conflict of interest.

Financial Disclosure: The authors declare that this study received no financial support.

References

- [1] Karray O, Oueslati A, Chakroun M, Ayed H, Bouzouita A, Cherif M, et al. Splenogonadal fusion- a rare cause of scrotal swelling: a case report. *J Med Case Rep* 2018;12:172. <https://doi.org/10.1186/s13256-018-1712-1>.
- [2] Chen S-L, Kao Y-L, Sun H-S, Lin W-L. Splenogonadal fusion. *J Formos Med Assoc* 2008;107:892-5. [https://doi.org/10.1016/S0929-6646\(08\)60206-5](https://doi.org/10.1016/S0929-6646(08)60206-5).
- [3] Bostroem E. Demonstration eines Präparates von Verwachsung der Milz mit dem linken Hoden. *Gesellschaft deutscher Naturforscher und Ärzte Verhandlungen der 56 Versammlung*. Freiburg, 1883:149.
- [4] Putschar WG, Manion WC. Splenic-gonadal fusion. *Am J Pathol* 1956;32:15-33. <https://pubmed.ncbi.nlm.nih.gov/13275562/>.
- [5] Lopes RI, de Medeiros MT, Arap MA, Cocuzza M, Srougi M, Hallak J. Splenogonadal fusion and testicular cancer: case report and review of the literature. *Einstein (Sao Paulo)* 2012;10:92-5. <https://doi.org/10.1590/s1679-45082012000100019>.
- [6] Zhou L, Muthucumar M, Stunden R, Lenghaus D. Splenogonadal fusion: a rare scrotal mass in a 9-year-old boy. *ANZ J Surg* 2018;88:E81-82. <https://doi.org/10.1111/ans.13250>.
- [7] Chiaramonte C, Siracusa F, Li Voti G. Splenogonadal Fusion: A Genetic Disorder? -Report of a Case and Review of the Literature. *Urol Case Rep* 2014;2:67-9. <https://doi.org/10.1016/j.eucr.2014.01.003>.
- [8] Sountoulides P, Neri F, Bellocci R, Schips L, Cindolo L. Splenogonadal fusion mimicking a testis tumor. *J Postgrad Med* 2014;60:202-4. <https://doi.org/10.4103/0022-3859.132350>.
- [9] European Association of Urology. Testicular Cancer Guidelines. <https://uroweb.org/guideline/testicular-cancer/#5> [Accessed: 30 Apr 2022].
- [10] Barlow LJ, Badalato GM, McKiernan JM. Serum tumor markers in the evaluation of male germ cell tumors. *Nat Rev Urol* 2010;7:610-7. <https://doi.org/10.1038/nrurol.2010.166>.
- [11] Gilligan TD, Hayes DF, Seidenfeld J, Temin S. ASCO Clinical Practice Guidelines on uses of serum tumor markers in adult males with germ cell tumors. *J Oncol Pract* 2010;6:199-202. <https://doi.org/10.1200/JOP.777010>.

- [12] Bieniek JM, Juvet T, Margolis M, Grober ED, Lo KC, Jarvi KA, Jarvi KA. Prevalence and Management of Incidental Small Testicular Masses Discovered on Ultrasonographic Evaluation of Male Infertility. *J Urol* 2018;199:481-6. <https://doi.org/10.1016/j.juro.2017.08.004>.
- [13] Scandura, G, Verrill C, Protheroe A, Joseph J, Ansell W, Sahdev A, et al. Incidentally detected testicular lesions <10 mm in diameter: can orchidectomy be avoided? *BJU Int* 2018;121:575-82. <https://doi.org/10.1111/bju.14056>.
- [14] Matei DV, Vartolomei MD, Renne G, Tringali VML, Russo A, Bianchi R, et al. Reliability of Frozen Section Examination in a Large Cohort of Testicular Masses: What Did We Learn? *Clin Genitourin Cancer* 2017;15:e689-96. <https://doi.org/10.1016/j.clgc.2017.01.012>.
- [15] Roy CR, Peterson NE. Positive hydrocele cytology accompanying testis seminoma. *Urology* 1992;39:292-3. [https://doi.org/10.1016/0090-4295\(92\)90310-s](https://doi.org/10.1016/0090-4295(92)90310-s).
- [16] Malik RD, Liu DB. Splenogonadal fusion: an unusual case of an acute scrotum. *Rev Urol* 2013;15:197-201. <https://pubmed.ncbi.nlm.nih.gov/24659917/>.
- [17] Tavernaraki E, Skoula A, Benakis S, Exarhos D. Side Effects and Complications of Magnetic Resonance Contrast Media. *Hospital Chronicles* 2012;7:208-14. <https://core.ac.uk/download/pdf/229445989.pdf>.