Case Report

Splenogonadal fusion: a rare case report

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Abstract

Splenogon adal fusion is a rare malformation consisting of an abnormal connection between the spleen and the gonad. Splenogonadal fusion has been classified into two types: continuous and discontinuous. The majority of the cases present as scrotal mass, scrotal tenderness or as clinical suspicion of testicular pathology, some cases are found incidentally during herniotomy or orchidopexy procedure, this report tries to highlight a case of discontinuous splenogonadal fusion found incidentally during orchidectomy in a 13-months-old male with tetra Amelia.

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Introduction

Splenogonadal fusion (SGF) is a rare congenital anomaly in which close proximity of spleen and gonad during early embryological development facilitates fusion[1]. Abnormal connections between spleen and the left gonad or the derivatives of the left mesonephros have been observed occasionally, this anomaly was first mentioned by the prominent pathologist Eugen Bostroem in 1883 [2], but the first detailed description of such an instance was published by Pommer in 1889[3] . Putschar and Manion classified SGF as continuous and discontinuous, in the continuous type, the principal spleen is connected to the gonad by a cord like structure that may be fibrous, totally splenic or beaded with multiple splenic nodules. In the discontinuous type there is no connection between the main spleen and the gonad, consisting of gonadal fusion with an accessory spleen or ectopic splenic tissue[4].

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Thirteen months old male who is a known case of congenital ***tetra Amelia syndrome*** presented with undescended left testis (figure 1). On examination of genitalia there was empty left hemiscrotum with normal right hemiscrotum and palpable normal right testis. Apart from absence of all four limbs and absence of left testis there were no other visible anomalies.

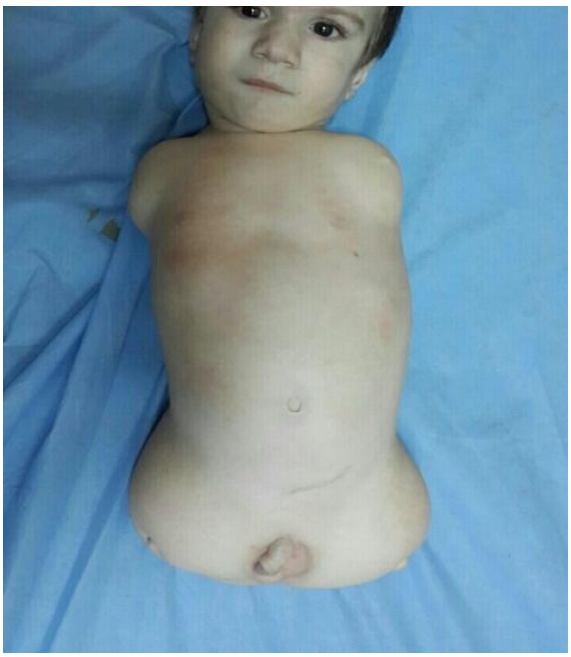

Figure 1: thirteen-month-male with *Tetra Amelia*

Ultrasound showed normal size left testis in the abdomen. CXR and echo were normal. On exploration; left testis was found to be free from attachment to the other abdominal viscera, it delivered outside, a normal looking testis was found with a red oval mass (2.5x1x1 cm) attached to the lower pole, the mass was firm and cannot be separated from testis (figure 2).

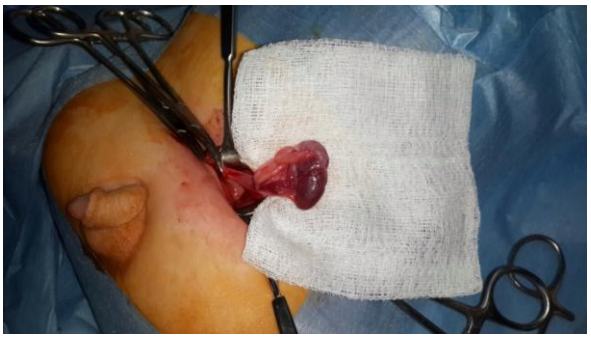

Figure 2: splenogonadal fusion, discontinuous type

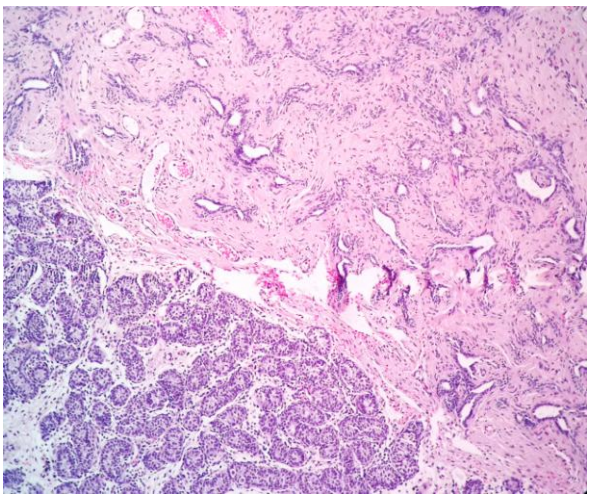

Figure 3: Histologic section showing primitive seminiferous tubules on the left with hypoplastic rete testis on the right (H&E stain, x100).

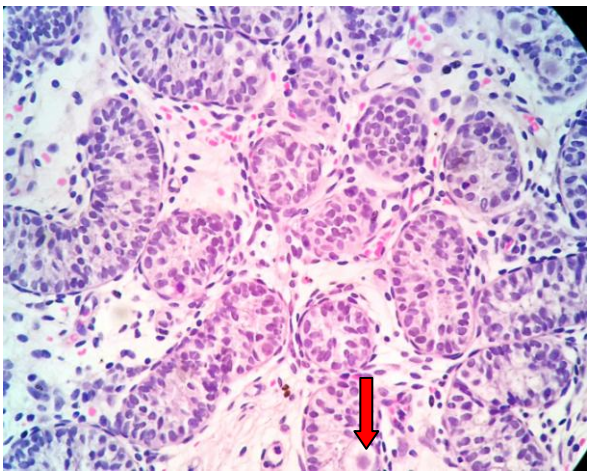

Figure 4: Histologic section showing decreased interstitial Leydig cells with rare germ cells (arrow) (H&E stain, x400).

Both the mass and the testis were excised and sent for histopathological examination, that revealed testicular tissue is composed of primitive small size somnifer-

ous tubules lined by Sertoli cells with absent to rare germ cells, decreased interstitial Leydig cells with hypoplastic rete testis (figure 3,4), normal splenic tissue fused to it and covered by tunica vaginalis (figure 5).

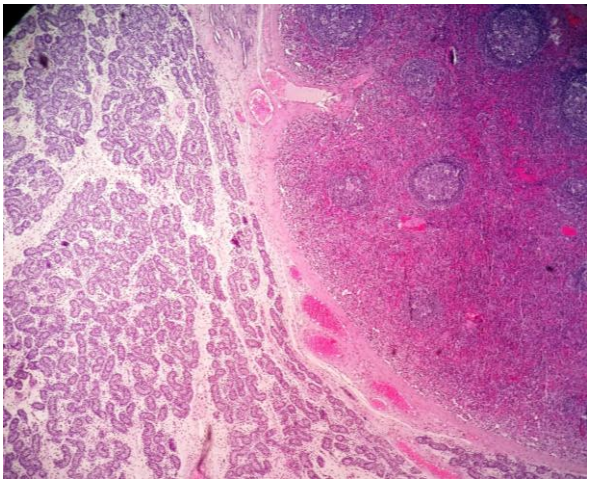

Figure 5: Histologic section showing normal spleen at the right aspect of the image and seminiferous tubules of the testis at the left aspect (H&E, x40).

Discussion

Splenogonadal fusion is rather rare entity with approximately 184 cases reported in the literature[5]. SGF is thought to occur between the splenic-anlage which is within close proximity to the left Urogenital fold until gonadal descent begins at the eighth week [6]. The splenic anlage is formed in the left dorsal mesogastrium in the fifth week, in embryos of 8-10mm length, and consists of multiple, small masses which usually fuse to form one organ[7], it is in intimate topographic relationship to the mesonephros and the gonadal anlage until gonadal descent and mesonephric involution begin at an embryonic length of about 20mm in eighth week [8]. The origin of SGF must be dated in the period of close proximity of these structures. At present the cause remains obscure three major theories have been proposed. Sneath suggested that inflammation between the gonadal ridge and spleen results in an adhesion between the two [9] however this theory does not explain the incidence of right-sided SGF or intraovarian or splenic tissue localized beneath the tunica albuginiea [10, 11]. Von Hochstetter postulated a retroperitoneal pathway that allows communication between the splenic and gonadal tissue [11]. Conversely, Putschar and Manion believed that SGF could be explained by envelopment of splenic tissue by tunica albuginea of the gonad[4].

Male to female ratio is 16:1, the male preponderance may be due to the fact that the male sex gonad is located superficially. Female gonads are inside the body and have fewer complications than their male counterparts. SGF is often discovered accidentally during gynecological surgery, with a higher prevalence in Caucasian individuals, followed by African descent and other ethnicity.[12]

SGF has been reported to have multiple associations with major congenital abnormalities; the majority of anomalies are seen in patient with continuous SGF, with a fivefold increase in occurrence in comparison with patients with discontinuous SGF[13]. In a fetus of 17mm in greater length the buds of the extremities are differentiating, Meckl's cartilage (the mold for the bony mandible) is forming and the spleen is in intimate contact with mesonephric-gonadal anlage[4]. commonly reported anomalies include peromelus and craniofacial abnormalities, specifically micrognathia, of a total of 184 cases in the literature 26% were noted to have an association with one or more congenital abnormalities [5] such as micrognathia, anal atresia, asymmetry of the skull, peromelus, abnormal fissure of lung and liver, spina bifida diaphragmatic hernia and hypospadius.[1,6,10,14,15]

Mushett [16] has shown experimentally on chick embryos that short periods of anoxia produce peromelus combined with malformations of eye, brain and hydromyelia of the spinal cord in addition to SGF and micrognathia.

SGF interferes with closure of processus vaginalis, and it is frequently associated with ipsilateral inguinal hernia and cryptorchidism [17] 31% of SGF patients are diagnosed with cryptorchidism or inguinal hernias, and in 59% of cases the cryptorchidism is diagnosed as bilateral[1]. Regarding cryptorchidism, 80% of the cryptorchidic testicles can be palpated while 20% are impalpable[18].

The most common locations for the cryptorchidic testicle are: inguinal canal 63%, ectopic location 11% external inguinal ring 9% and abdominal only 2%.[18]

The diagnosis of SGF prior to surgery is challenging, SGF typically presents as an asymptomatic testicular mass and other manifestations may include acute testicular pain and swelling caused by ectopic splenic tissue infection[19] but the actual disease itself lacks characteristic features.

The lack of awareness of SGF is a major factor in it is misdiagnosis. Imaging methods, including B-type ultrasonography, computed tomography CT, magnetic resonant image MRI, ^{33}Tcm spleen scanning aid with the diagnosis of SGF[16,20, 21, 22] however; laproscopies have achieved improved diagnosis and management of SGF [23]. The diagnostic evaluation of patients with an abnormal gonad is complex due to multifactorial etiopathogenesis and the rarity of the condition. The primary aim of diagnosis is to rule out malignancy,

three steps that should be considered when diagnosing and treating SGF. Firstly, a mass found at birth growing slowly for several years in a benign condition should be considered. Secondly, various imaging techniques should be used to investigate the nature of the mass. Thirdly, in doubtful cases, a biopsy should performed during surgery or, preferably prior to making an incision, for example, by needle biopsy, punch biopsy or a classical bivalve biopsy and regional node evaluation. If the mass proves to be malignant, a radical resection should be performed immediately. If the malignancy is unconfirmed, an orchiectomy is sufficient for further pathological study. In cases where the mass has been identified to be benign, but the organ has been opened, removal of splenic tissue may be performed.[12] With SGF, there may be no need for surgery. However, diagnosis and surgical treatment by a diagnostic laparoscopy are suggested. Laparoscopy is a safe and reliable approach that is highly accurate in the diagnosis and treatment of non palpable testis.[12]

Conclusion

SGF is a rare condition and diagnostic evaluation of patients with abnormal gonad is complex due to multifactorial etiopathogenesis and the rarity of the condition, although rare, the splenogonadal fusion should be considered in the differential diagnosis of scrotal masses in children, and orchiectomy should be avoided.

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Competing Interests

None declared.

References

1. Cortes D, Thorup JM and Visfeldt J: The pathogenesis of cryptorchidism and splenogonadal fusion: a new hypothesis. Br J Urol 1996;77: 285-290.
2. Hock BC, Spleno-gonadal fusion.2011;7:101-3
3. Pommer, G. Verwachsung des linken kryptorchischen Hodens und Nebenhodensmit der Milz in einer Missgeburt mit zahlreichen Bildungsdefecten. Ber. d. naturw.-med. Ver. in Innsbruck, I887/I889;I7-19:I44-8.
4. Putschar WG, Manion WC. Splenicgonadal fusion. Am J Pathol. 1956;32:15-33.
5. Rena DM and Dennis BL.Splenogonadal Fusion: An unusual case of an acute scrotum.Rev Urol. 2013; 15:197-201
6. Carragher AM. One hundred years of splenogonadal fusion. Urology. 1990;35:471-5.
7. Sabin, F. R. The Development of the Spleen. In: Manual of Human Embryology, Keibel, F., and Mall, F. P. (eds.). J. B. Lippincott Co., Philadelphia,I9I2, 2, 745-5I
8. Felix, W. The Development of the Urinogenital Organs. In: Manual of Human Embryology, Keibel, F., and Mall, F. P. (eds.). J. B. Lippincott Co., Philadelphia, I912, 2, 752-979.
9. Sneath WA. An apparent third testicle consisting of a scrotal spleen. J Anat Physiol.1913;47:340-342.
10. Gouw AS, Elema JD, Bink-Boelkens MT, et al. The spectrum of splenogonadal fusion. Case report and review of 84 reported cases. Eur J Pediatr. 1985; 144: 316-23.
11. Von Hochstetter A. Milzgewebe im linken Ovarium des linken Individualteiles eines menschlichen Thoracopagus. Virchows Arch. 1953;324:36-54.
12. Li WF, Luan MX, Ma Z and Chen YJ. Splenogonadal fusion:Report of four cases and review of the literature.2013;6:816-818.
13. Khairat AB, Ismail AM. Splenogonadal fusion: case presentation and literature review. J Pediatric Surg. 2005;40:1357-60.
14. Guarin U, Dimitrieva Z, Ashley SJ. Spleno-gonadal fusion. Brunei Int Med J.2011;7(2):101-103
15. Lipsonn AH. Amelia of the arms and femur/fibula deficiency with splenogonadal fusion in a child born to a consanguineous couple. Am J Med Genet 1995;55: 265-8
16. Mushett, C. W. Elektive Differenzierungsstorungen des Zentralnervensystems am Hiihnchenkeim nach kurzfristigem Sauerstoffmangel. Beitr. z. path. Anat. u. z. aUlg. Path., I953, II3, 367-387.
17. Varma DR, Sirineni GR, Rao MV, et al. Sonographic and CT features of splenogonadal fusion. Pediatr Radiol. 2007;37:916-919.
18. Schneck FX, Bellinger MF. Abnormalities of the testis and scrotum and their management. In: Wein AJ, Kavoussi LR, Novick LR, Partin AW, Peters CA, editors. Campbell-Walsh urology. 9th ed. Philadelphia: Saunders Elsevier; 2007. p. 3761-98.
19. Stewart VR, Sellars ME, Somers S, Muir GH and Sidhu PS: Splenogonadal fusion: B-mode and color Doppler sonographic appearances. J Ultrasound Med. 2004; 23: 1087-1090.
20. Netto JM, Pérez LM, Kelly DR, Joseph DB and Royal SA: Splenogonadal fusion diagnosed by Doppler ultrasonography. Scientific World Journal.2004; 4: 253-257.
21. Li YH: Preoperative detection of splenogonadal fusion by CT. Surg Radiol Anat. 2009;31:733-735.
22. Alalayet YF, Mansoor K, Shiba NA, Khan AM and Al Kasim F: Splenogonadal fusion. Eur J Pediatr Surg. 2008;18: 342-344.
23. Papparella A, Nino F, Coppola S, Donniacono D and Parmeggiani P: Laparoscopy in the diagnosis and management of splenogonadal fusion: case report. Eur J Pediatr Surg. 2011; 21: 203-204.