Splenogonadal fusion; a rare histopathology report of a pediatric patient.

Anwar A AlSuwailem. Arwa S AlOlyan. Sarah M AlRashed. Saroona Haroon. Saifaldin M AlZain.

Abstract In 1883, was the first description of splenogonadal fusion (SGF) by Bostroem, which is a rare developmental anomaly of an abnormal connection of splenic tissue with gonads or mesonephric remnants. SGF is of two types; continuous or discontinuous. Almost none of the reported cases have been diagnosed preoperatively, they mainly discovered coincidently during surgeries like orchiopexy and inguinal hernioplasty. We report a case of 12-year old boy presented with history of the absence of testis since birth. Upon physical examination, a mass felt in the left inguinal region raising a possibility of being testicle while the right one was impalpable. Laparoscopic left inguinal exploration and orchiepexy have been done. Histopathology report of a removed mass then revealed ectopic splenic tissue fused within the residual testicle with no increased mitosis or neoplastic features have been identified. Although only 175 SGF reported cases have been found in literature, the condition should be considered in differential diagnosis for every testicular mass. Recommend using such advanced facilities for those patients upon diagnosis like tumor markers, 99 m Tc-sulfur colloid liver spleen scan, inguinal incision and possible biopsy before surgery to avoid unnecessary orchiectomy.

Key words: Spleen; gonads; pediatrics; congenital; fusion; sonographic; laparoscopic.

Introduction:

Splenogonadal fusion (SGF) is a sporadic congenital anomaly. First described in 1883, in which there is an aberrant attachment of spleen to gonads or mesonephric remnants [1]. Very few cases of SGF has been diagnosed preoperatively as it does not have any characteristic clinical or radiological features, and may lead to unavaoidable orchiectomy due to its crucial clinical resemblance to testicular neoplasm. However still it is very important to do excisional biopsy as there are many benign and malignant mimics including hematoma, epidermoid cyst, adrenal rests, nonseminomatous germ cell tumors, and lymphoma. SGF is of two types, continuous or discontinuous. It usually diagnosed coincidently during surgeries like orchiopexy and inguinal hernioplasty.

Case presentation:

A 12-year old boy presented to Paediatric outpatient department in Maternity and Children Hospital with history of the absence of testis; since birth. There was no significant past medical or surgical history, as well as other systems' review did not reveal any abnormality. The family history was also unremarkable. On physical examination, vital signs were normal (including temperature 37°c), weight 33 kg and fairly hydrated. Examination of the genital area exhibited circumscribed penis and poorly developed scrotum. A mass was felt in the left inguinal region was raising possibility of being testicle aand the right one was impalpable. Scrotal ultrasonography (SUG) & Doppler was performed, and it was found that both testes could not be seen in the scrotum. In addition, no hydrocele could be seen bilaterally. The patient then admitted electively and a laparoscopic exploration of testis & orchiepexy were planned.

After left testicular orchiectomy, the pathological gross examination revealed a 3 x 2.5 x 1cm reddish brown, un-encapsulated circumscribed nodule, with spermatic cord measuring 4.5 x 0.7 x 0.5 cm. The nodule was having smooth outer surface and cut section was soft, solid with diffuse brownish areas. No necrosis or any tan whitish solid nodule seen. On microscopic examination, a circumscribed lesion was found, with residual testicular tissue present focally compressed. There were lymphoid follicles and sinusoidal tissue representing white pulp and red pulp (Fig – 2). No intra-tubular germ cell neoplasia was identified. Active and complete spermatogenesis could not be seen. However, only few spermatogonia were identified in the seminiferous tubules (FIG-3). No increased mitosis, significant atypia or atypical lymphoid cells was identified. No other germ line component was identified on multiple sections.

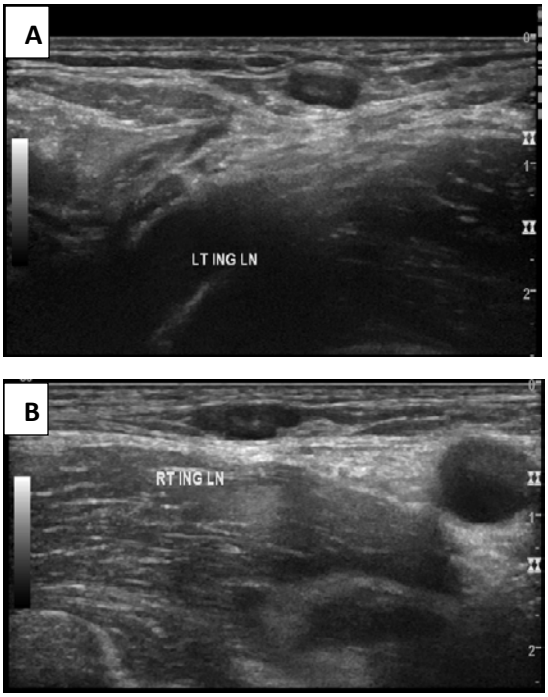

Fig. 1. [A, B]

Both testes could not be seen in scrotum nor inguinal canal at time of scan. Multiple bilateral small sized inguinal LNs are seen. No hydrocele could be seen bilaterally.

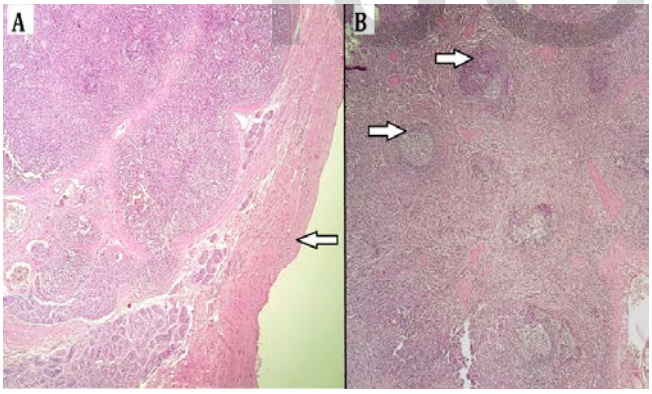

Fig. 2.

Histological examination of the mass. Hematoxylin and eosin staining shows that it was surrounded by a capsule (FIG 1 A – Arrow) and splenic tissuecomprised red pulp, white pulp (FIG 1 B – Arrows) and splenic trabeculae. (x10)

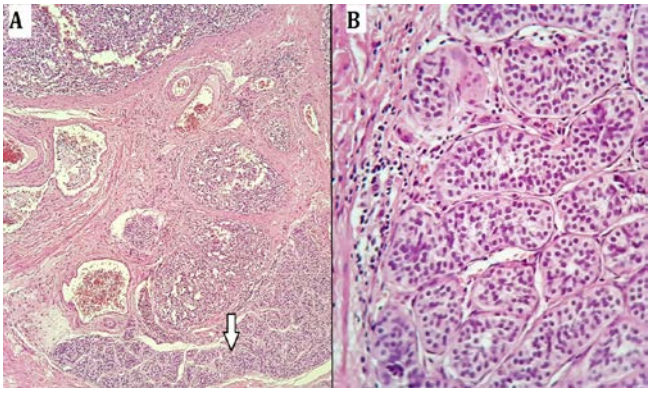

Fig. 3.

Higher power reveals compressed seminiferous tubules in the surrounding (FIG 2 A (20x) – Arrow). The seminiferous tubules do not show any convincing proper spermatogenesis (FIG 2 B (40x)

Discussion:

Splenogonadal fusion is a rare congenital anomaly. About only 175 cases have been described in the literature to date, 50% of them present before 10 years of age, however our patient was 12 years old [2]. The left testis is almost always to be affected, because of the anatomical proximity of the left gonad to the spleen, as in our case [3]. In 1956, Putschar and Manion classified SGF into two types, continuous and discontinuous depending on whether an anatomical connection is detected between the principle spleen and gonad or not, respectively [4]. Associated congenital disorders have been reported in the mostly continuous-type SGF like cryptorchidism, limb defect, micrognathia, or less commonly cardiac, lung, intestinal defect. While the discontinuous-type is rarely associated with other congenital anomalies, however some of the cases are reported with infertility. Our patient was having no other developmental defect \ discontinuous type SGF.[3] SGF usually presents by concerning about cryptorchidism and discovered incidentally during groin exploration. Also it may present as a scrotal mass.[3] Or as a painful scrotal lump due to testicular torsion.[4] Such a clinical presentation has limited preoperative diagnostic measures. Almost impossible to establish the diagnosis before surgery [6], but it's recommended to do tumor markers (a-fetoprtein, lactate dehydrogenase, B-human chorionic gonadotropin)

also to rule out any germ cell component [5]. Unfortunately, the diagnosis of discontinuous type of SGF is confirmed by orchiectomy [3]. Or sometimes we can rely on 99 m Tc-sulfur colloid liver spleen scan [4]. Intra-abdominal band of hypoechoic tissue in ultrasonography is a suggestive for a continuous type of SGF. While it is a controversial in diagnosis a discontinuous type [3] However, a scrotal ultrasonography (SUG) & Doppler were done for our patient. Orchiectomy is used to be the only management for most of the reported cases; "137 patients, of whom 37% had unnecessary orchiectomy." [6] And it's rare to be found associated with testicular aplasia [7].

Conclusion:

Because the SGF is mimicking a testicular tumor, the urologic surgeons must consider it in their differential diagnosis upon facing a case of testicular mass, especially with long standing unchanged or un-noticed left testicular mass [5]. Recommend inguinal incision, and possible biopsy of the mass (laparoscopic assessment) before orchiectomy to avoid excision of a non-neoplastic testis [3].

References:

1. Bal, K. et al., 2014. Splenogonadal fusion : a very rare congenital anomaly in the differential diagnosis of a testicular mass. , 40(1), pp.62–64.

2. Kumar, S. et al., 2014. Case Report A Rare Case of Continuous Type Splenogonadal Fusion in a Young Male with Primary Infertility. , 2014(Figure 3).

3. Report C. Journal of Surgery : Open Access Splenogonadal Fusion : Laparoscopic Diagnosis and Treatment. 2015;1–2.

4. Kocher, N.J. et al., 2014. Images in Clinical Urology Solid Testicular Mass. *Urology*, 83(1), pp.e1–e2. Available at: http://dx.doi.org/10.1016/j.urology.2013.09.019.

5. Choi, J., Han, S. & Kim, Y.J., 2011. Splenogonadal Fusion : A Case Report 비성선융합 : 증례보고. , 65(6), pp.589–593.

6. Chen, S. et al., 2008. Splenogonadal Fusion. *Journal of the Formosan Medical Association*, 107(11), pp.892–895. Available at: http://dx.doi.org/10.1016/S0929-6646(08)60206-5.

7. Ando, S. et al., 2006. Case Report Splenogonadal fusion : Case report and review of published works. , pp.1539–1541.