

Case Report

Splenogonadal Fusion Associated with Bilateral Cryptorchidism, Presenting as an Operative Surprise

Maryam Abolhasani^{1,2}, Mojgan Asgari^{1,2}, Hossein Keymoosi³

1. Oncopathology Research Center, Tehran University of Medical Sciences, Tehran, Iran

2. Hasheminejad Clinical Research Developing Center (HCRDC), Tehran University of Medical Sciences, Tehran, Iran

3. Department of Pathology, Tehran University of Medical Sciences, Tehran, Iran

ABSTRACT

Splenogonadal fusion is a rare entity with approximately 150 reported cases until the year 2005. The entity is a rare congenital anomaly in which there is fusion of splenic and gonadal anlagen or mesonephric derivatives. Splenogonadal fusion has two continuous and discontinuous types. About ten cases have been reported in association with bilateral cryptorchidism. Very few cases have been diagnosed preoperatively. Many cases present as a testicular swelling and undergo an unnecessary orchiectomy with the suspicion of a testicular neoplasm. Herein, we report a new case of discontinuous splenogonadal fusion in a 29 year old man with bilateral cryptorchidism who underwent surgery. Surprisingly two masses were noted adjacent to undescendent testis in left groin in operative room which were resected and proved to be a new case of splenogonadal fusion in histological exam.

Keywords: Splenogonadal Fusion, Bilateral Cryptorchidism, Iran

Introduction

Splenogonadal fusion is a rare entity with approximately 150 reported cases until year 2005 since the first description in 1883 by Bostroem (1). It occurs predominantly in males with a male to female ratio of about 16:1 (2). The age of reported cases ranges from stillborn to 81 years (3). Nearly half of the

cases presented below 10 years (2) and 82% below 30 years (4). Two forms of continuous and discontinuous splenogonadal fusion have been described (5). The continuous form occurs when the anatomic spleen is connected by a discrete cord to the gonad. The discontinuous type consists of a fused splenogonadal structure that has lost continuity with the main spleen. The

Received: 04 April 2012

Accepted: 21 May 2012

Address communications to: Dr. Maryam Abolhasani, Oncopathology Research Center, Tehran University of Medical Sciences, Tehran, Iran

E mail: m-abolhasani@sina.tums.ac.ir

latter type is a variant of accessory spleen. The continuous type seems to be the predominant (6). Splenogonadal fusion has been associated with other congenital anomalies. Most cases that had multiple anomalies were of the continuous variety. The most common defect is of the limbs (2, 7-9). The association of limb defect supports the believed gestational teratogenic period. The differentiation of limb buds and mandible occurs from the sixth to seventh week of gestation (2, 10). Other associated anomalies include micrognathia, cardiac defect, cleft palate, anal defects, spina bifida and facial muscle agenesis (2, 7, 10). Furthermore, most cases have been reported with ipsilateral cryptorchidism (2, 8-9, 11-13).

About ten cases have been reported in association with bilateral cryptorchidism (2, 14-16). During surgery for orchiopexy of undescended testes, the surgeon sees a mass that may be clinically mistaken for neoplasm and an unnecessary orchiectomy may be performed. Knowledge of this anomaly may spare patients an unnecessary orchiectomy (1, 17-18).

Case Report

A 29 year old male with empty scrotum was admitted to our center which is a referral center for urology. Physical examination showed bilateral undescended testes but penis had normal anatomy. The patient had azospermia in semen analysis. Other physical exams and laboratory data were normal. He underwent laparoscopic surgery. Right undescended testis had normal size but left testis was atrophic and separate from epididymis. Surprisingly two ovaloid shaped masses were noted in left groin during surgery. The clinical impression was that patient had polyorchidism. Right testicular orchiopexy and left orchiectomy along with resection of two masses were performed and the specimens were sent to pathology ward.

Gross examination showed small size of left testis and epididymis. The two masses had reddish

brown cut surface and macroscopic appearance of spleen (Fig. 1).

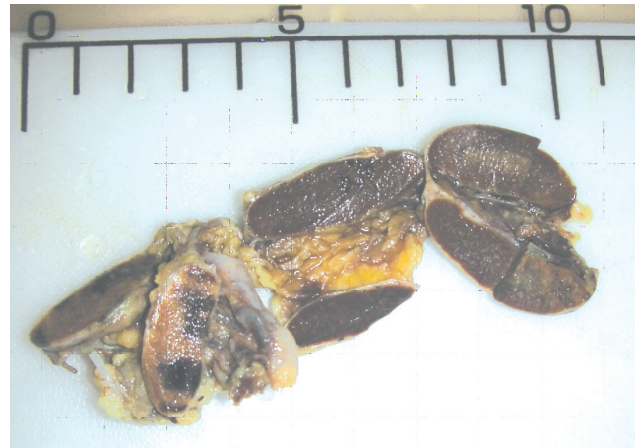


Fig. 1- Gross examination of left testis and epididymis along with the two masses having reddish brown cut surfaces.

Microscopic examination of masses revealed typical normal splenic tissue with white and red pulps. Histological exams of H&E stained slides of left testis and right testicular biopsy displayed seminiferous tubules with thick hyalinized basement membrane, filled only by Sertoli cells and showing germ cell aplasia (Fig. 2). Leydig cells were increased in interstitium. There was no evidence of intratubular germ cell neoplasia or malignancy. Microscopic evaluation of left epididymis showed primitive tubules. These microscopic findings are usually noted in histological examination of undescended testis and epididymis.

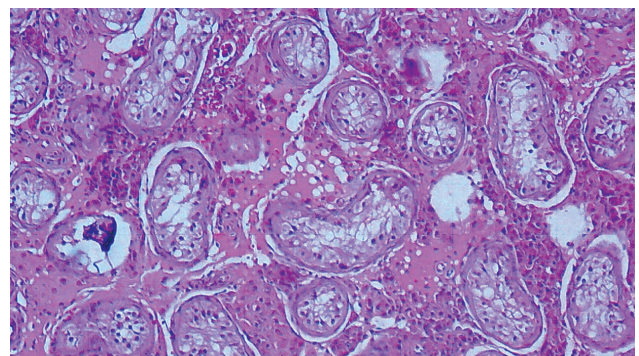


Fig. 2- Histological examination of testes with seminiferous tubules, filled only by sertoli cells and showing germ cell aplasia (H&E stain $\times 20$)

Discussion

Our case was a new case of splenogonadal fusion in a 29 year old man presenting with bilateral cryptorchidism and azospermia. During operation, he seemed to have polyorchidism in left groin but it proved to be splenogonadal fusion in histological exam.

In most instances, the anomaly is recognized as an incidental finding at autopsy or at surgical exploration of the abdomen (10). Our case presented with infertility and was diagnosed in work up for azospermia, which is an unusual presentation.

Most cases of splenogonadal fusion have been reported with ipsilateral (2, 8-9, 11-13) and rarely with contralateral cryptorchidism (18). About ten cases were associated with bilateral cryptorchidism (2, 14-16). The rarity of this case was its presentation as azospermia and association with bilateral cryptorchidism.

Many of the reported cases had other congenital anomalies including the limb abnormalities which are the most common reported anomalies in literature (2, 7-9). Our patient did not have other congenital anomaly, so preoperatively there was no clinical suspicion for presence of splenogonadal fusion.

Techniques of diagnostic imaging are available if there is a clinical suspicion of splenogonadal fusion. The most reliable preoperative imaging, according to published articles, is technetium isotope scanning, which detects accessory splenic tissue (10). This study was not performed for our patient because there was no clinical suspicion of this diagnosis.

It must be known that intraoperative frozen sections may be mistaken for malignancy and orchiectomy may be performed unnecessarily (2, 19). In our case there was no suspicion for neoplasm, so no frozen sample was sent to pathology room.

In literature, there was another case of splenogonadal fusion reported in Iran, who was a boy with left cryptorchidism operated at the age

of 1 year. A reddish-brown structure was noted during surgery which was traced to the lower pole of the spleen and was normal splenic tissue in histopathological examination (13). That case had unilateral cryptorchidism and was younger than our patient and had a continuous form of splenogonadal fusion.

Because of rarity of this condition, it is infrequently diagnosed preoperatively (1, 17) and is often diagnosed as a histological surprise. Knowledge of this anomaly may prevent surgeons from an unnecessary orchiectomy, on the basis of a suspected primary neoplasm of the gonad (1, 17-18). This diagnosis should be kept in mind during inguinal surgery. The correct recognition of the lesion can preserve normal function of the testicle at least from the endocrinal standpoint (17).

Acknowledgments

We acknowledge Pathology technicians of "Hasheminejad kidney Center" (Mrs Elham Goldar and her colleagues) for embedding Paraffin blocks and staining.

References

1. Khairat AB, Ismail AM. Splenogonadal fusion: case presentation and literature review. *Pediatr Surg* 2005; 40(8):1357-60.
2. Carragher AM. One hundred years of splenogonadal fusion. *Urology* 1990; 35(6):471-5.
3. Diebold J, Le Blaye O, Le Tourneau A, Marichez P. Intra-scrotal supernumerary spleen. A long silent case of discontinuous spleno-gonadal fusion. *Ann Pathol* 1990; 10(3):174-6.
4. Karaman MI, Gonzales ET Jr. Splenogonadal fusion: report of 2 cases and review of the literature. *J Urol* 1996; 155(1):309-11.
5. Keyik B, Yanik B, Conkbayir I, Tuygun C, Kizilgoz V, Hekimoğlu B. Continuous-type splenogonadal fusion associated with an ipsilateral testicular atrophy: sonographic findings. *J Clin Ultrasound* 2010;

- 38(3):161-3.
6. Bostwick DG, Cheng L. Urologic Surgical Pathology. 2nd ed. China:Elsevier Inc;2008; 866-867.
 7. Cortes D, Thorup JM, Visfeldt J. The pathogenesis of cryptorchidism and splenogonadal fusion: a new hypothesis. *Br J Urol* 1996; 77(2):285-90.
 8. Hizli F, Uygur MC, Irkkan C. Splenogonadal fusion: report of a case. *Int J Urol* 2005; 12(6):591-2.
 9. Basnyat PS, Jones DA, Morgan RJ, Davies CJ, Foster ME. Splenogonadal fusion: report of a rare variety. *J R Coll Surg Edinb* 2001; 46(2):108-9.
 10. Guarin U, Dimitrieva Z, Ashley SJ. Splenogonadal fusion-a rare congenital anomaly demonstrated by 99Tc-sulfur colloid imaging: case report. *J Nucl Med* 1975; 16(10):922-4.
 11. Braga LH, Braga MM, Dias MA. Laparoscopic diagnosis and treatment of splenogonadal fusion associated with intra-abdominal cryptorchidism in a child. *Pediatr Surg Int* 1999; 15(7):465-6.
 12. Khelif K, Maassarani F, De Keuleneer R, Segers V. Splenogonadal fusion: a case report. *Acta Chir Belg* 2010;110(6):607-8.
 13. Molaeian M, Shojaei H. Splenogonadal fusion presented with cryptorchidism. *Urol J* 2009; 6(2):130-1.
 14. Duncan WL Jr, Barraza MA. Splenogonadal fusion: a case report and review of literature. *J Pediatr Surg* 2005; 40(12):e5-7.
 15. Knorr PA, Borden TA. Splenogonadal fusion. *Urology* 1994; 44(1):136-8.
 16. Al-Marhoon M, Matthew J, Nirmala V, Kehinde EO. Splenogonadal fusion associated with primary male infertility. *BJU Int* 2000; 86(4):560-1.
 17. Breda G, Zattoni F, Artibani W, Brunetti A, Vancini P. Splenogonadal fusion. *Chir Ital* 1976; 28(6):862-8.
 18. 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):173-5.
 19. Bearss RW. Splenic-gonadal fusion. *Urology* 1980; 16(3):277-9.