

Case Report

Laparoscopic diagnosis and management of splenogonadal fusion: Case report and review of literature

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Splenogonadal fusion is a rare congenital anomaly in which there is fusion of the spleen and the gonad or mesonephric derivatives. Approximately 150 cases have been reported since the condition was first described by Bostroem in 1883. The diagnosis of this uncommon anomaly is rare even to be suspected preoperatively; Here a case describing laparoscopic diagnosis and management has been done and review of the literature.

Key words: Testis, spleen, splenogonadal fusion, laparoscopic.

INTRODUCTION

Splenogonadal fusion is a rare entity with approximately 150 cases reported since the first description of this entity in 1883 by Bostroem and in 1888-1889 by Pommer. Close proximity of the spleen and gonad during early embryological development allows fusion, whether continuous or discontinuous, of these seemingly unrelated organs. The continuous type of splenogonadal fusion describes the gonad attached to the anatomic spleen. The discontinuous type consists of gonadal fusion with an accessory spleen or ectopic splenic tissue. The diagnosis of this uncommon anomaly is rare even to be suspected preoperatively. Laparoscope is more diagnostic than the ultrasound, CT scan, magnetic resonance imaging (MRI), and helpful in the management. Here a case of continuous splenogonadal fusion is presented as an impalpable left testicle. This case is unique in that the laparoscopic management in such condition is revealed after negative open groin exploration.

CASE PRESENTATION

An 11 year-old boy with impalpable left testis since birth was presented. He had ultrasound, CT scan examinations and open groin exploration which revealed no left testis. Physical examination, apart from the left

groin scar and the impalpable left testis was unremarkable. Routine pre-operative laboratory investigations were within normal range. On laparoscopic exploration a reddish brown, smooth cord of tissue measuring about 20 mm in diameter was observed to be coming from above to down in a peritoneal fold ending by fusion to the superior pole of the testis intra-abdominal higher to the internal ring of the inguinal canal (Figure 1). Grossly, the tubular cord had the appearance of splenic tissue having a serosal capsule and fibrous trabeculae and a vascular pedicle running on its medial aspect (Figure 2). The splenic cord like tissue fuse with upper pole of the testis, with a line of demarcation between the different tissues. Laparoscopic assisted left orchidopexy with preservation of the spleen was then performed. The patient has uneventful postoperative follow up for one year. Histo-pathological examination confirmed that the specimen was spleno-gonadal fusion and has no evidence of malignance.

DISCUSSION

This case is an unusual presentation of splenogonadal fusion. The case is unique in that it was diagnosed and managed with laparoscope. Meta-analysis of published reports of 111 boys with splenogonadal fusion revealed

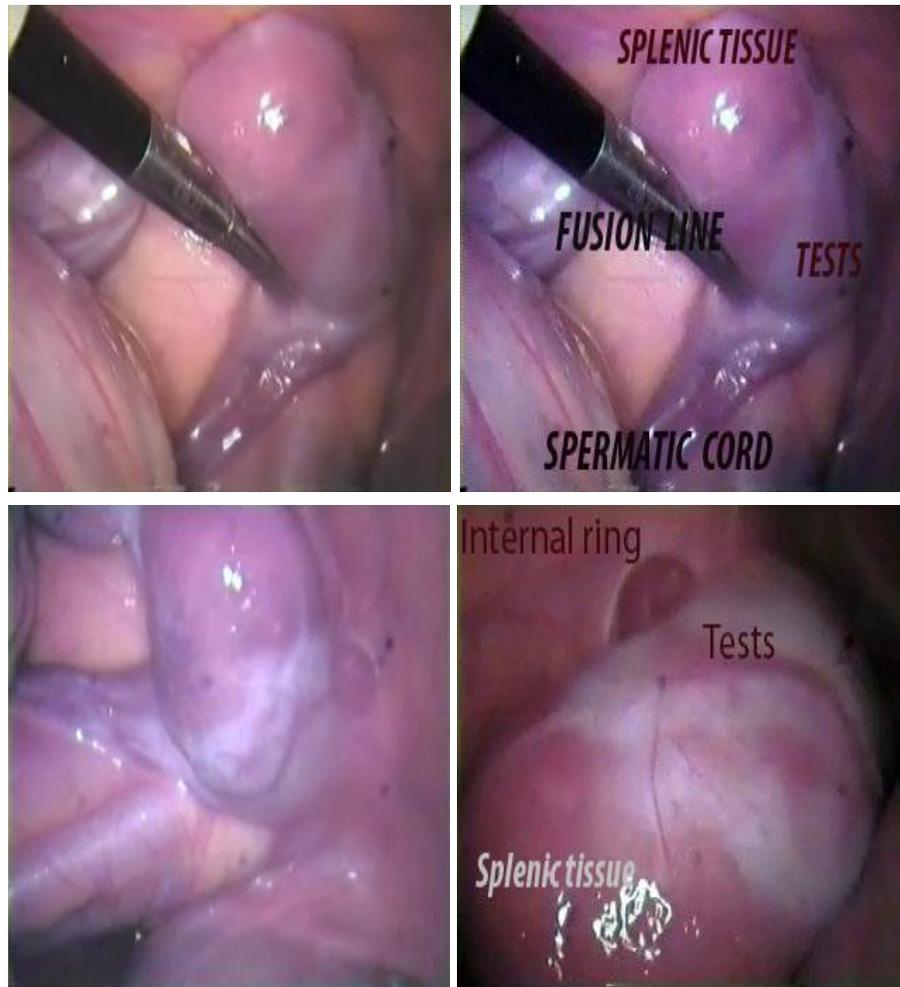


Figure 1. Multiple pictures of splenogonadal fusion.



Figure 2. This is splenic tissue with vascular pedicle on its surface coming out from the upper pole of the spleen and going down to fuse with the upper pole of the left intra-abdominal testes.

that 31% had cryptorchidism. Of these, 59% were bilateral, 26% had right intra-abdominal testes, and 65% had left intra-abdominal testes. Of those with continuous splenogonadal fusion, 44% had cryptorchidism. Solely cryptorchid cases with spleno-gonadal fusion reportedly

had bilateral absence of legs, imperforate anus, spina bifida, diaphragmatic hernia, and hypospadias (Cartes, 1995).

About 120-to-150 case of splenogonadal has been reported in the literature. Splenogonadal fusion is most

commonly an incidental discovery during a routine groin exploration for an undescended testis or hernia. In our case the surgeon who had done the open groin exploration could not find the testes because the testes and the cord were totally highly intra-abdominal. Nearly 17% of the splenogonadal fusions were diagnosed at autopsy (Karaman and Gonzales, 1996). Testicular or inguino-scrotal swelling was the most common presentation [5]. In this case there was no scrotal or inguinal swelling; there was also a scare of negative open groin exploration. According to Karaman and Gonzales (1996), 37% of 137 cases underwent an unnecessary orchiectomy because of suspicion of a primary testicular neoplasm. Only four were reported with a malignant testicular neoplasm and a coexistent splenogonadal fusion. Other presentations include acute painful scrotal swelling secondary to affection of the ectopic splenic tissue by various processes. Talmann (1926) and Settle (1940) reported cases presented with acute scrotal pain and swelling secondary to malaria involvement of the ectopic splenic tissue. These patients' symptoms subsided as the malaria resolved. Acute torsion of the splenic tissue (Karaman and Gonzales, 1996), mumps, leukemia, and mononucleosis (Andrews, 1985) and traumatic rupture of the ectopic spleen (Halvorsen and Stray, 1978) also presented as painful scrotal swellings. Mechanical bowel obstruction by the intra-peritoneal cord of the continuous splenogonadal fusion was described by Hines and Eggum (1961). Sripathi (1999) reported one case of macro-orchidism. Few cases were diagnosed preoperatively; one of such was reported by Kadlic in 1943. Three cases were diagnosed by 99mTc-sulphur colloid liver-spleen scan; one of them during workup of a patient with an undescended left testicle and associated limb malformations (Guarin, 1975), and two cases during evaluation of intra-abdominal mass Falkowski and Carter, 1980; Steinmetz et al., 1997). Patel (1995) diagnosed one case by ultrasonography when he followed a tubular process arising from the upper pole of the spleen down to the upper pole of a left undescended testis. Our case has similar anomalies with Patel's case but cannot suspect or diagnose by ultrasound or CT-Scan prior to the previous surgery what had been done. He also noted movement of the upper splenic pole when applying traction to the testis. The left side is far commonly involved than the right side. Only three cases (2%) had a discontinuous right sided splenogonadal fusion and were all male (Gordeef and Cuenant, 1951; Tiberio et al., 1965). Half of the cases presented below 10 years (Carragher, 1990) and 82% below 30 years (Karaman and Gonzales, 1996). It is predominant in male; however, male-to-female ratio is about 1:16 (Carragher, 1990).

Two forms of splenogonadal fusion have been described as continuous and discontinuous. The continuous form occurs when the anatomic spleen is connected by a discrete cord to the gonad. The discontinuous form consists of a fused splenogonadal structure that has lost continuity with the main spleen.

This is a variant of an accessory spleen. The continuous type seems to be predominant (Putschar and Manion, 1956). Our case is a continuous type of splenogonadal fusion. A column of splenic tissue come out from the upper pole of the spleen, passes through downward anterior to the anterior splenic boarder, swing to the left over the splenic flexure of the colon, then passes through left paracolicgatture to fuse with the left test in the abdominal cavity.

Two theories have been proposed to describe splenogonadal fusion. Von Hochstetter attempted to explain this entity by a retroperitoneal pathway for the splenic angle to come into contact with the developing gonad. In this theory, the splenic cells could potentially be found along the pathway of gonadal descent (May and Bourne, 1974; Von Hochstetter, 1953). Sneath (1912-1913) proposed that inflammation over two opposing peritoneal surfaces, namely, the gonadal ridge and spleen, could cause fusion. During gonadal migration, the peritonealized adhesion would lengthen and develop as a cord continuous with the spleen or rupture during development, making it discontinuous with the spleen (May and Bourne, 1974). Because of the rarity of this condition it is infrequent to be diagnosed preoperatively (William and Mark, 2005). Techniques of diagnostic imaging are available if there is a clinical suspicion of splenogonadal fusion. The most reliable preoperative imaging, according to published results, is technetium isotope scanning, which detects accessory splenic tissue (William and Mark, 2005). Laparoscopic diagnosis of impalpable testes is superior to all investigation including ultrasound, CT scan, or even MRI (Gregory et al., 2011). Laparoscope was valuable and highly effective not only in the diagnosis but also in the management of this case. It should be pointed out that orchiectomy has been performed needlessly (Pommer, 1888-1889). The uniqueness in this case is the use of laparoscope in the diagnosis and management. The search of the database shows there is laparoscopic use in splenogonadal fusion.

CONCLUSION

Splenogonadal fusion is a rare condition, seldomly malignant. Diagnostic imaging has a limited role in the evaluation of boys with undescended testes and it its related condition. It is recommended that efforts be made to increase routine use of laparoscope in the evaluation of patients with cryptorchidism. Laparoscope is essential for diagnosis and management of simple, complex and rare anomalies associated with undescended testes such as splenogonadal fusion.

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