Continuous-type splenogonadal fusion: report of a rare case

Oktav Bosnalı, İnanç Cici, Serdar Moralıoğlu, Ayşenur Cerrah-Celayir

Department of Pediatric Surgery, Zeynep Kamil Maternity and Children's Training and Research Hospital, İstanbul, Turkey. E-mail: droktav@gmail.com

Received: 24 March 2014, Revised: 14 April 2014, Accepted: 7 May 2014

SUMMARY: Bosnalı O, Cici İ, Moralıoğlu S, Cerrah-Celayir A. Continuous-type splenogonadal fusion: report of a rare case. Turk J Pediatr 2014; 56: 680-683.

Splenogonadal fusion is a rare congenital anomaly. It has two subtypes, discontinuous and continuous. Up to one-third of continuous-type fusion is associated with other congenital anomalies. We present a continuous-type splenogonadal fusion case that was found incidentally during indirect hernia repair; the testicle was preserved during excision. Laparoscopic exploration was helpful in identifying the isolated polysplenia as the origin of continuous-type splenogonadal fusion, and in excising the cord-like attachment proximally. The patient had no other associated anomaly.

Key words: splenogonadal fusion, spleen, testicle, laparoscopy.

Splenogonadal fusion is a very rare congenital anomaly, which may be continuous or discontinuous, depending on the presence or absence of a structural connection between the spleen itself and the ectopic splenic tissue attached to the gonad. The continuous type involves a direct anatomic connection between the spleen and gonad by means of a cord made of splenic and/or fibrous tissue. The cord may be attached to any area of the splenic hilum or to the inferior pole of the spleen. The discontinuous type is characterized simply by gonadal fusion with an accessory spleen or ectopic splenic tissue[1]. In approximately 30-33% of cases, other congenital anomalies, such as limb defects, micrognathia, cardiac defects and cleft palate, may be associated with the continuous type[2]. The condition is usually found incidentally during inguinal hernia repair or during exploration for an undescended testis or testicular mass[3]. We present a continuous-type splenogonadal fusion case found incidentally during indirect hernia repair in a 7-year-old boy, in which the pathology was managed with a laparoscopic approach, and the testicle was preserved.

Case Report

A 7-year-old boy was admitted to our clinic, with bulging in the left groin that had existed for 3 years. Upon physical examination, there was visible bulging in the left inguinal region, and an indirect inguinal hernia was palpable. There was no cord thickening on either side; both testicles were in the scrotum and palpation of the left testis was completely normal. There were no other pathological physical examination findings except for a soft swelling in the inguinal region. The diagnosis was left indirect inguinal hernia, and he was scheduled for elective inguinal hernia repair.

During inguinal hernia repair, a fibrous cord-like structure was discovered in the hernia sac, running from the intra-abdominal cavity through the inguinal channel. The cord was lying intraperitoneally within the sac, without any adhesions. To explore further, we delivered the testis with its tunica vaginalis from the inguinal incision and opened the tunica. Exploration revealed a round mass at the end of the cord-like structure, and this round mass was fused to the upper pole of the testes, which looked different from it (Fig. 1).The presumed diagnosis according to the macroscopic appearance was splenogonadal fusion; it was decided to excise the mass, since there was a clear margin between it and the testicle. After excision, we decided to proceed with diagnostic laparoscopy as in a contralateral hernia sac investigation, since there was a cord-like structure originating from the mass and entering the abdomen through the hernia sac.

A 5 mm laparoscopy port for video use was inserted via the left hernia sac into the abdominal cavity, and laparoscopic exploration revealed polysplenia as the origin of continuous splenogonadal fusion, with splenic tissue remnants along the cord. An additional 5 mm working port was inserted suprapubically, and extensive splenic tissue and the cord-like structure were excised with bipolar cautery (Figs. 2 and 3). The patient was fed two hours after the operation and discharged home on the same day. Detailed postoperative examination showed no additional congenital anomalies. Histopathological examination reported splenic tissue in round, bead-like pieces and fibrotic tissue in the cord structure.

Discussion

Splenogonadal fusion is a rare anomaly; its subtypes, associated anomalies and treatment have been well-described to date, beginning with Boestroem's first description in 1883, as cited by Sieber in 1969[4]. Since then, approximately 160 cases have been reported, according to a recent (2010) literature review[5].

The exact cause of splenogonadal fusion is not clear; two theories have been proposed to explain how those two embryologically close yet different structures fuse with each other. Since the splenic anlage in the dorsal mesogastrium is opposite the genital anlage in the 6-week-old embryo, fusion can occur between those two neighboring structures during this time, either by adhesion between the peritoneal surface of the spleen and the gonadal ridge, or by a slight inflammation of the peritoneal surface over the spleen and the gonadal ridge[2,4,6].

Two types of splenogonadal fusion have been described: continuous and discontinuous. The continuous type involves a direct anatomic connection between the spleen and gonad by a cord that is made of splenic and/or fibrous tissue. The cord may be attached to any area of the splenic hilum or to the inferior pole of the spleen. The discontinuous type is characterized simply by gonadal fusion with an accessory spleen or ectopic splenic tissue[1,6]. The cord in continuous-type splenogonadal fusion may be beaded, partially fibrotic or smooth and uniform in diameter; its course may be either intraperitoneal or retroperitoneal. It has been reported that when it is located intraperitoneally, the cord can cause small bowel obstruction by entanglement with the bowel loops, or colonic obstruction at the splenic flexura by compressing the colon[4,5,7]. Thus, excision of the cord as close to the spleen as possible would seem of benefit in preventing such complications. In our case, we found a continuous-type splenogonadal fusion with a beaded cord attached to a supernumerary spleen instead of to an accessory spleen or to the spleen itself, which differed from the usual presentations reported. Excision of the cord was completed with laparoscopy.

Approximately 50% of the continuous-type splenogonadal fusion cases may be associated with severe congenital anomalies. Reported anomalies include peromelia, micrognathia,

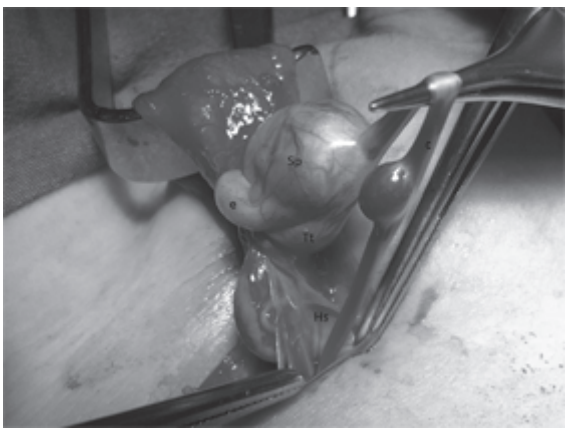

Fig. 1. Splenogonadal fusion to the upper pole of the testis. Sp: fused splenic tissue, c: fibrous cord, e: epididymis, Tt: testicular tissue, Hs: Hernia sac, grasped by two atraumatic pinsets.

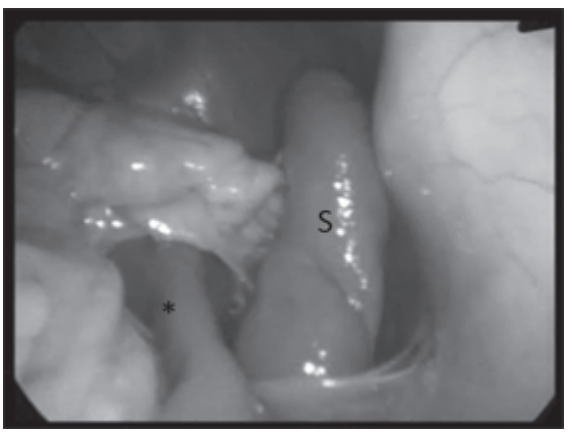

Fig. 2. Laparoscopic appearance of polysplenia as the origin of continuous-type splenogonadal fusion. S: Spleen, *: polysplenia

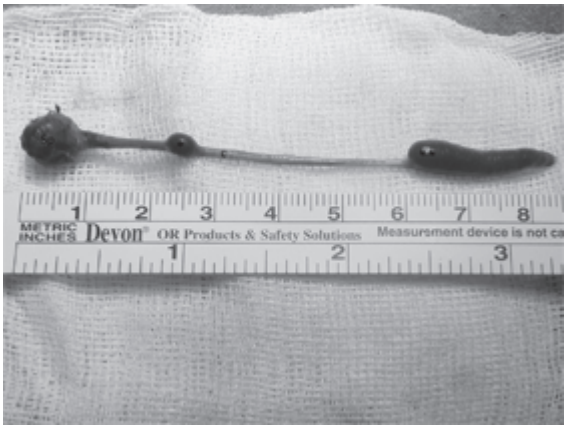

Fig. 3. Macroscopic appearance of cord structure (c), with ectopic splenic tissues (* and **) and fused splenic tissue (Sp).

cardiac defects, cleft palate, anal anomalies, microgastria, spina bifida, craniosynostosis, diaphragmatic hernia and hypoplastic lung[2,6,8,9]. In our case, a detailed postoperative physical and radiological examination was conducted, in addition to a pediatric cardiology examination. There were no other associated anomalies.

Splenogonadal fusion is most often an incidental finding during groin exploration for cryptorchidism or hernia repair, but it may also present as an intrascrotal mass. In the literature, the anomaly has rarely been recognized and the testicle preserved at operation. Should this condition be suspected during surgery, the testicle should be preserved, as the splenic tissue can easily be separated, and the condition is completely benign[3,9]. Even if the condition is suspected preoperatively, the use of ultrasonography cannot be recommended since it does not provide sufficient information or images that are specific to splenogonadal fusion. Contrast-enhanced computerized tomography may aid in diagnosis, but exposure to radiation during CT limits its use in the clinical setting as a diagnostic tool in such cases. In cases where there is a reasonable suspicion of splenogonadal fusion, some authors advocate the use of technetium-99m sulfur colloid scintigraphy to identify ectopic splenic areas. However, scintigraphy may not be readily available at all centers[10].

Papparella et al.[8] reported laparoscopic diagnosis and treatment of a splenogonadal fusion case in a child. In their case, the laparoscopic approach allowed the vascularization of the ectopic splenic tissue to be managed by complete evaluation of the site. The advantages of using laparoscopy in children, such as its role in definitive diagnosis and treatment, low postoperative discomfort, better cosmesis and early discharge to home are well known. Laparoscopy also gives an opportunity for, in the same session, diagnosis of the splenogonadal fusion and associated intraabdominal anomalies, especially in cases in which the pathology is discovered during inguinal hernia surgery. In our case, proceeding with laparoscopy made possible the diagnosis of the accompanying polysplenia and proximal excision of the excess cord-like tissue, which might have caused future intra-abdominal complications.

In conclusion, splenogonadal fusion is an uncommon congenital anomaly that is rarely diagnosed or suspected preoperatively. Thus, when an unexpected spleen-like mass is found attached to the testicle, the testicle should be preserved, as in most cases the splenic tissue is easily dissected off the gonadal structures. Proceeding with laparoscopy helps to identify the origin of continuous-type splenogonadal fusion and to confirm the diagnosis, and also allows for the cord-like attachments to be excised proximally in order to prevent possible future complications.

REFERENCES

1. Duncan WL Jr, Barraza MA. Splenogonadal fusion: a case report and review of literature. J Pediatr Surg 2005; 40: E5-E7.
2. Khairat AB, Ismail AM. Splenogonadal fusion: case presentation and literature review. J Pediatr Surg 2005; 40: 1357-1360.
3. Alivizatos G, Skolarikos A, Sopilidis O, Ferakis N, Chorti M. Splenogonadal fusion: report of a case and review of the literature. Int J Urol 2005; 12: 90-92.
4. Sieber WK. Splenotesticular cord (splenogonadal fusion) associated with inguinal hernia. J Pediatr Surg 1969; 4: 208-210.
5. Alvarez Maestro M, López-Tello J, Domínguez Franjo P, Ríos González E, Martínez-Piñeiro L. [Splenogonadal fusion. Report of a case and review of the literature]. [Article in Spanish] Actas Urol Esp 2010; 34: 293-295.
6. 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: 161-163.

7. Chen SL, Kao YL, Sun HS, Lin WL. Splenogonadal fusion. J Formos Med Assoc 2008; 107: 892-895.
8. Papparella A, Nino F, Coppola S, Donniacono D, Parmeggiani P. Laparoscopy in the diagnosis and management of splenogonadal fusion: case report. Eur J Pediatr Surg 2011; 21: 203-204.
9. Lin CS, Lazarowicz JL, Allan RW, Maclennan GT. Splenogonadal fusion. J Urol 2010; 184: 332-333.
10. Patafio FM, Dufton JA, Walker DR. Case of the Month #173: splenogonadal fusion. Can Assoc Radiol J 2011; 62: 302-304.