Splenogonadal Fusion
Sung-Lang Chen,1* Yu-Lin Kao,1 Hung-Shun Sun,1 Wei-Lung Lin2

Splenogonadal fusion (SGF) is a rare congenital non-malignant anomaly characterized by fusion of splenic tissue to the gonad, and can be continuous or discontinuous. Very few cases have been diagnosed preoperatively, and many patients who present with testicular swelling undergo unnecessary orchiectomy under the suspicion of testicular neoplasm. A 16-year-old boy presented with a left scrotal mass and underwent total excision of a 1.6-cm tumor without damaging the testis, epididymis or its accompanying vessels. Pathologic examination revealed SGF (discontinuous type). If clinically suspected before surgery, the diagnosis may be confirmed by Tc-99m sulfur colloid imaging, which shows uptake in both the spleen and accessory splenic tissue within the scrotum. Frozen section should be considered if there remains any doubt regarding the diagnosis during operation. [J Formos Med Assoc 2008;107(11):892–895]

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Splenogonadal fusion (SGF) is a rare congenital anomaly where the spleen is abnormally attached to a gonad. The condition was first described by Bostroem in 1883.1 Putschar and Manion reported two types of SGF in 1956.2 The continuous type involves a direct anatomic connection between the ectopic and orthotopic spleen, by a cord that may be totally splenic, beaded with multiple splenic nodules, or composed of fibrous tissue. The cord runs via either a retroperitoneal or transperitoneal course, in which it has been reported to cause small-bowel obstruction by entanglement with bowel loops.3 The discontinuous type consists of gonadal fusion with an accessory spleen or ectopic spleen tissue. Le Roux and Heddle suggested that the latter type was simply a rare variant of an accessory spleen and there was no solid evidence that it shared the same etiology or that it is associated with the same congenital anomalies as the continuous type.4

During the 5th week of embryological development, the splenic anlage develops in the left dorsal mesogastrium. The developing spleen comes into close proximity of the left urogenital fold during rotation of the embryonic gut. The urogenital fold contains the gonadal mesoderm, which develops into gonadal structures. The spleen–gonadal relationship remains until the 8th week of development when gonadal descent is initiated.4

The diagnosis is almost always made when SGF presents as a testicular mass, or as an incidental finding during orchiopexy or inguinal hernioplasty.5 Increased awareness of this condition could avoid unnecessary orchiectomy as the testis can usually be separated and preserved while the lesion is removed. We present a child...
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with discontinuous-type SGF. Testis sparing surgery was undertaken after confirmation with frozen section.

Case Report

A 16-year-old boy presented to our institute for evaluation of a left testicular mass. On physical examination, the right testis was normal in shape and size. The left testis was in the scrotum, and an elastic mass was identified at the upper pole of the left testis. Testicular tumor markers were within normal limits. Alpha-fetoprotein (AFP) was 1.85 ng/mL. Beta human chorionic gonadotropin (β-HCG) was 0.1 mIU/mL. Sonography of the scrotum revealed a well-circumscribed hypoechoic lesion that measured about 1.6 × 1.1 cm over the upper pole of the left testis (Figure 1). Magnetic resonance imaging of the scrotum confirmed that there was a mass lesion about 1.6 cm in size in the left testis, which showed low SI in T2WI and T1WI (Figure 2). Contrast study showed good enhancement. Malignant testicular tumor could not be ruled out by imaging study.

The left testicle was approached through an inguinal incision. A reddish brown mass measuring about 1.6 cm was found attached to the upper pole of the left testis, which was removed entirely without any damage to the testicle, vessels or epididymis (Figure 3). The excised mass was sent for frozen section examination. Microscopically, the section revealed lymphoid tissue with hyperplastic follicles and was well encapsulated. The biopsy tissue was consistent with splenic tissue, separated from normal testicular tubules by a dense fibrous band (Figure 4). The patient’s postoperative course

Figure 1. Sonography reveals a well-circumscribed hypoechoic lesion measuring about 1.6 × 1.1 cm over the upper pole of the left testis.

Figure 2. Magnetic resonance imaging of the scrotum confirms the presence of a mass lesion about 1.6 cm in size in the left testis with good enhancement.

Figure 3. Gross photograph of the excised splenogonadal fusion lesion.

Figure 4. Splenogonadal fusion: normal testicular tubules (upper left) and splenic tissue (lower right) separated by a fibrous band (hematoxylin & eosin, original magnification 400×).
was unremarkable. He remains well at 36 months after the operation.

**Discussion**

Approximately 150 cases of SGF have been reported since the first description of this condition by Bostroem in 1883. The most recent comprehensive review of this entity was in 1990 by Carragher. At that time, just more than 120 cases had been reported in the literature. Approximately one fifth were not discovered until autopsy. Seventy-one (56%) were continuous and 52 (44%) were discontinuous. The study reports a male: female ratio of 16:6:1. However, the incidence of females may be underestimated due to the inability to palpate the female gonads by routine physical examination. All the cases described in females are of the continuous type and found at autopsy or as an incidental finding at laparotomy.

There are several theories of causation, all based on the closed proximity of the left gonadal ridge and splenic anlage between the 5th and 8th weeks of gestation. This explains why it is almost always on the left. As the presence of SGF interferes with normal gonadal descent and closure of the processus vaginalis, this condition is commonly associated with inguinal hernia or undescended testis. The most common association with SGF is cryptorchidism. Cortes et al, in their review of 111 SGF cases, reported cryptorchidism in 31%, of which 59% were bilateral undescended testis. Associated malformation such as limb anomalies and micrognathia are seen, as the limb buds and mandible undergo active development during this period. Other associated anomalies have been described in up to 33% of patients with continuous type SGF. On the other hand, it is proposed that the discontinuous type of SGF is a rare variant of accessory spleen. This variant is rarely associated with congenital anomalies.

The clinical diagnosis is problematic, and imaging is not conclusive. Sonography is currently the imaging modality of choice for the assessment of scrotal abnormalities. The appearance of SGF on sonography has previously been described as resembling a mass within the scrotal sac of low reflectivity in comparison with normal testicular parenchyma. The accessory spleen is usually found within the tunica vaginalis and is closely attached to the gonad, although a distinct capsule is present. Even more rare are ectopic rests discovered in the epididymis or spermatic cord. The mass may not be seen as separate from the testis and, as such, can be readily confused with primary testicular tumor, as occurred in our case. Except for one reported case, there is no intermingling of testicular and splenic tissues. If clinically suspected before surgery, the diagnosis may be confirmed by Tc-99m sulfur colloid imaging, which shows uptake in both the spleen and the accessory splenic tissue within the scrotum. However, on sonography, differentiation of SGF from testicular malignancy is difficult, and the rarity of SGF makes the clinical pathway of performing a Tc-99m sulfur colloid examination an unlikely scenario. Very few cases are diagnosed preoperatively, leading to suboptimal management by orchiectomy in the mistaken belief that it is a malignant process. This was the case in the review of Karaman and Gonzales of 137 patients, of whom 37% had unnecessary orchiectomy.

Increased awareness of this entity should allow for a more prompt diagnosis and avoid unnecessary orchiectomy. In most cases, the splenic tissue may be dissected off the gonadal structures easily. If there is doubt about the diagnosis, frozen section and inguinal approach should be considered even if tumor markers are normal.

**References**