Splenogonadal fusion: a forgotten cause of testicular swelling in children

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Splenogonadal fusion is a rare congenital anomaly in which there is fusion of the spleen and the gonad. This report describes a 25-month-old male child who presented with left scrotal swelling. On exploration, a discontinuous type of splenogonadal fusion was found. It was successfully and completely separated from the testis and removed. The diagnosis of splenogonadal fusion should always be kept in mind and included in the differential diagnosis of testicular swelling in infants and children. Rarely, it is diagnosed preoperatively and physicians caring for these patients should be aware of this. Intraoperatively, it should be recognized and excised. This is to obviate unnecessary orchiectomy, mistaking it for testicular neoplasm. *Ann Pediatr Surg* **10:25–26 © 2014 Annals of Pediatric Surgery.**

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Introduction

Splenogonadal fusion is a very rare congenital malformation that is rarely diagnosed preoperatively. It is characterized by fusion of the spleen and gonad [1–6]. There are two types of splenogonadal fusion: continuous and discontinuous [4,5]. The continuous form occurs when the normally located spleen is attached to the gonad by a discrete cord. In the discontinuous type, the splenic tissue is attached to the gonad and completely separated from the normal spleen. Both types occur with equal frequency and the discontinuous type may be discovered incidentally during herniotomy or orchidopexy or present as a scrotal swelling [2,3,6,7]. This report describes a child who presented with left scrotal swelling and the diagnosis of splenogonadal fusion was made intraoperatively. The literature on the subject is also reviewed. Awareness of this is of great importance to avoid unnecessary orchidectomy if mistaken for a testicular tumor.

Case report

A 25-month-old boy was referred to our hospital with a left scrotal swelling that was noticed 12 months before presentation. The swelling was painless and did not change in size. There was no history of trauma. On examination, there was a firm swelling just above the left testis that appeared like an encysted hydrocele of the cord but it did not transilluminate. Exploration was performed through a left inguinal incision. This showed a bluish-red mass that grossly had the appearance of splenic tissue (Fig. 1). The mass was connected to the upper pole of the testis by fibrous adhesions. The testis appeared normal. The mass was completely removed from the testis using both sharp and blunt dissection without any damage to the testis, vas, vessels, or epididymis. Histology of the resected mass showed normal splenic tissue. Postoperatively, he did well and was discharged home on the second postoperative day. It has been 14 months postsurgery and he is well.

Discussion

In 1883, Bostroem described the first case of splenogonadal fusion [1–3]. Since then, more than 150 cases have been reported in the literature but because of its rarity, most publications are case reports [3]. In 1990, Carragher [1] reported the first extensive review of 123 cases of splenogonadal fusion. Karaman and Gonzales, in 1996, reviewed 137 cases of splenogonadal fusion and, surprisingly, 37% of the patients had orchiectomy because of suspicion of a testicular tumor [4]. This is of great importance and to obviate this, physicians caring for these patients should be aware of this differential diagnosis and if there is doubt intraoperatively regarding the nature of the swelling, frozen section biopsy should be performed to avoid unnecessary orchiectomy.

Putschar and Manion, in 1956, and in a collective review of 30 cases classified splenogonadal fusion into two types: continuous and discontinuous [2]. In the continuous type, there is a direct connection between the normal spleen and gonad by a cord that may be totally made up of splenic tissue, multiple connected beads of splenic tissue, or a cord made up of fibrous tissue. In the discontinuous type, there is no connection between the normal spleen and ectopic spleen that is attached to the gonad. This is considered a rare variant of an accessory spleen. Both types occur with relatively equal frequency [3].

The exact etiology of splenogonadal fusion is not known. Embryologically, the testis starts to descend from its initial embryological position between the dorsal mesogastrium and the mesonephros at around the eighth week of intrauterine life. This occurs at the time of splenic development. Splenogonadal fusion is considered to result from partial fusion of splenic and gonadal tissues in the 4–8 weeks of intrauterine life. Subsequent descent of the gonad during the 8–10 weeks of gestation results in descent of a part of the developing spleen along with it. In the discontinuous type, there is complete detachment from the normal spleen, whereas in the continuous type there is

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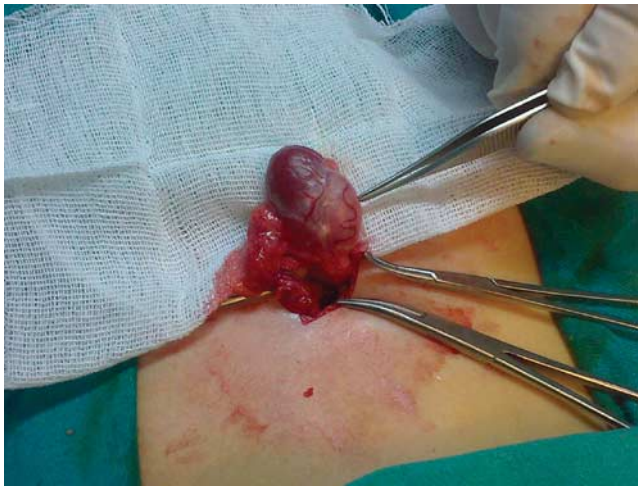
Fig. 1



Intraoperative photograph showing splenogonadal fusion.

attachment to the normal spleen by a cord-like structure. This cord can be made up of splenic tissue or may be totally fibrotic. Occasionally, there are multiple nodules along this cord that represent foci of splenic tissue that became detached and developed separately [2,3]. This, however, does not fully explain the occasional occurrence of right-sided splenogonadal fusion.

Splenogonadal fusion is commonly asymptomatic, discovered incidentally during routine herniotomy or orchidopexy [2,3,6,7]. Many of these cases, however, go unnoticed or discovered at autopsy. In the pediatric age group, these patients commonly present with a scrotal swelling and may rarely present with acute scrotal pain as a result of torsion or involvement of splenic tissue with other pathological conditions such as mumps, malaria, leukemia, trauma, and infectious mononucleosis [3]. The left side is far more commonly affected than the right side in 98% of the cases [3]. It is more common in males. The ratio of M : F is 16 : 1 [3]. This, however, may not be true as the ovary is not easily accessible and as the majority of these cases are asymptomatic, the incidence of splenogonadal fusion in females may be underestimated. Not uncommonly, splenogonadal fusion is discovered during evaluation of associated anomalies [2,3,8]. This is more so with the continuous type, which is known to be associated with other anomalies in as much as 33% of the cases [3]. These include peromelia, which is categorized as a separate syndrome (splenogonadal fusion limb syndrome), micrognathia, congenital heart disease, microgastria, cleft palate, craniosynostosis, osteogenesis imperfecta, spina bifida, congenital diaphragmatic hernia, and anorectal anomalies [2,3,6,8]. Our patient did not have any other associated anomalies.

There is also an association between splenogonadal fusion and testicular malignancy. In the literature, there are about seven reported cases of splenogonadal fusion and testicular malignancy, but in all of these cases, the malignancy developed in adults with undescended testes or following orchidopexy for undescended testes [2,3,5,9]. This may represent an association rather than an increased risk in this subset group of patients as patients with undescended testes have a well-known increased risk of malignancy.

Conclusion

Splenogonadal fusion is a very rare congenital anomaly that must be kept in mind and considered especially if a child presents with an unusual scrotal swelling. Awareness of this is of great importance to avoid unnecessary orchidectomy if mistaken for a testicular tumor. If suspected preoperatively, the diagnosis can be confirmed by a ^{99m}Tc-sulfur colloid scan [10]. Treatment is surgical excision and every attempt should be made to preserve the gonad at the time of dissection and excision, which should not be difficult as true fusion with the gonad is rare. Although there have been a few reports of an association of testicular neoplasms with splenogonadal fusion, it appears that this is a coincidental finding rather than an association as these tumors occurred in adults with undescended testes.

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Conflicts of interest

There are no conflicts of interest.

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