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Pediatrics Splenogonadal fusion: A rare finding during routine orchiopexy

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ABSTRACT

Splenogonadal fusion remains a rare congenital anomaly, with limited reports in the literature. It is important to be aware of this benign condition as orchiectomy can result from confusion with malignant processes. We present a case of splenogonadal fusion in a 12-month-old healthy male with history of cryptorchidism found at the time of surgery.

Introduction

Splenogonadal fusion (SGF) is a rare congenital anomaly, resulting in the fusion between spleen and gonad. It was first reported in literature in 1883 by Bostroem with approximately 150 cases reported in literature since then.^{1,2} SGF can present as an isolated finding or in association with limb defects, micrognathia, anal atresia and other congenital anomalies.^{1,3} Awareness of this anomality is important as almost 37% of cases reported in literature is associated with unnecessary orchiectomy.^{1,4}

Case presentation

A 12-month-old healthy male patient was referred for evaluation of left cryptorchidism. On physical exam, he was noted to have normal descended right testicle and a palpable mass in the left groin presumed to be his left testicle. He was subsequently taken to the operating room for a left orchiopexy.

The orchiopexy began through a scrotal approach. The testis was delivered through the scrotal incision, at which time an adherent splenule was also delivered (Fig. 1). The splenic tissue, which was approximately $2 \text{ cm} \times 3 \text{ cm}$ and contained a tail extending into the retroperitoneal space, was carefully dissected free from the testicle and cord structures and released into the retroperitoneum through a subsequent inguinal incision (Fig. 2). A left hernia sac was also identified, divided and ligated. Once freed from the splenic tissue, the testicle easily reached the dependent scrotum and was secured with monocryl suture. 6 weeks post-operatively follow-up revealed the left testicle well positioned in the scrotum with no hernias or masses in the groin.

Discussion

Although SGF is more commonly reported males with a male:female ratio of 15:1, this discrepancy is likely due to under diagnosis in females due to the intraabdominal location of the gonad.¹ While the exact pathophysiology of the fusion is unclear, it is thought that the fusion between the spleen and gonad occurs between the 5th and 8th week of gestation, prior testicular descent.³

There are two forms of SGF, continuous and discontinuous. In the continuous form, there is a structural attachment between the spleen and gonad. This structural attachment can range from a cord-like fibrous structure, to completely splenic or beaded with multiple splenic nodules.⁴ In the discontinuous form, accessory splenic tissue is fused to the gonad and there is no attachment to the orthotopic spleen. Slightly more than half of the cases (55%) reported in literature were cases of continuous SGF.⁴ In both forms, the fused splenic mass is most commonly fused to the testicle, though they can also be fused to the epididymis or spermatic cord.¹ The splenic tissue is usually encapsulated by its own capsule, allowing for it to be separated from the testicle; however, there has been 1 reported case of intermingling of splenic and testicular tissue.^{1,4} Histological evaluation of the fused splenic tissue is usually normal.³

The continuous form of SGF is more commonly associated with other congenital anomalies, including cryptorchidism, limb defects, cardiac defects, micrognathia, cleft palate, anal atresia, and spina bi-fida.¹⁻³ Of these malformations, limb defects are most common and is termed splenogonadal fusion-limb defect syndrome.

Conclusion

Splenogonadal fusion is a rare benign entity that is important to

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Fig. 1. Accessory splenule attached to the spermatic cord.



Fig. 2. Accessory splenule freed from the spermatic cord.

recognize as it can lead to unnecessary orchiectomy. The accessory splenic tissue can be easily separated from the testicular tissue and cord structures, thus allowing the undescended testicle to be brought down into the scrotum.

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