Case Report

Torsion of a Splenule in a Case of Splenogonadal Fusion Mimicking a Strangulated Inguinal Hernia

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INTRODUCTION

Splenogonadal fusion (SGF) is a rare congenital anomaly which occurs due to an abnormal fusion of splenic tissue with the derivatives of the perimesonephros. It usually presents as an inguinal hernia, testicular mass, or undescended testis.[1] To the best of our knowledge, no case was presented as a strangulated hernia. We report a case of torsion of a splenule in a case of SGF in a 6-year-old boy who presented as a strangulated inguinal hernia. He was successfully managed with an inguinal procedure.

CASE REPORT

A 6-year-old boy presented to the emergency room in the evening hours with a complaint of an acute painful left inguinal swelling noticed since morning. There was no history of vomiting, distension of the abdomen, or fever. There was no history of similar episodes. On examination, he had tachycardia; cardiovascular system, respiratory system, and abdominal examination were normal. Groin examination revealed a tender, irreducible swelling in the left inguinal region with normal bilateral descended testes, leading to a working diagnosis of strangulated inguinal hernia. However, a quick ultrasonography showed a soft-tissue swelling as the content of hernia with reduced vascularity adding the possibility of torsion of a supernumerary testis. With these two diagnoses in mind, the child was taken up for an emergency inguinal exploration.

Left inguinal exploration revealed an indirect left inguinal hernia with no bowel or omentum in it, but an elongated, oval, firm, purplish-blue structure connected distally to the upper pole of the left testis by a fibrous cord and proximally entering the peritoneal cavity by a fibrous cord. The fibrous cords on either side had undergone torsion by 360°, leading to early gangrenous changes [Figure 1]. The firm swelling was excised by dividing the fibrous cords from the upper pole of the testis (taking care to avoid injury to the testis) distally and as high as possible proximally. Herniotomy was done. Oral feeds resumed on the same day, and the child was discharged on the 1st postoperative day. Histopathology revealed a fibroelastic capsule covering a tissue comprised of red pulp, white pulp, and dilated sinusoids, consistent with an ectopic spleen. There was no testicular tissue noted in the excised specimen, clinching the diagnosis of SGF, the continuous type [Figure 2]. Ultrasonography abdomen showed normal orthotopic spleen. Subsequent follow-ups were uneventful.

DISCUSSION

SGF is a rare entity with <200 cases reported in the literature till date. It results from an abnormal fusion of splenic tissue with the derivatives of the perimesonephros which occurs between 5 and 8 weeks of intrauterine life.[1] The etiology is unknown, but a few theories have been explained. A few of them include

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postinflammatory adhesions between gonadal ridge and spleen, abnormal retroperitoneal pathway allowing the communication between the gonad and spleen, and gonadal tunica enveloping the developing spleen.

There are two types of SGF; continuous and discontinuous. In the continuous type, the orthotopic spleen is in connection with the gonad by a splenic cord, fibrous cord, or beaded cord containing splenic and fibrous tissue. In the discontinuous type, the orthotopic spleen is not connected to the gonad, but an ectopic splenic tissue is attached to the gonad. The continuous type is more common than the discontinuous type (58%).

It can be associated with micrognathia or limb anomalies as the occurrence of SGF coincides with the development of the mandible and limb bud. The continuous type is found to have five times higher incidence of associated anomalies than the discontinuous variety. The other associated anomalies reported include unilateral or contralateral undescended testis, hypospadias, coarctation of the aorta, cleft palate, osteogenesis imperfecta, Potter syndrome, malrotation of gut, anorectal malformation, transverse testicular ectopia, and persistent Mullerian duct syndrome.

SGF is reported more commonly in males (93%) as most cases in females get unnoticed due to the benign nature of the disease in them. In females, SGF is detected incidentally on laparotomy or at autopsy while most males present with an inguinal hernia, undescended testis, supernumerary testis, or scrotal swelling. The other uncommon presentations include inguinal swelling which enlarges during malaria and viral infections, torsion testis, or intestinal obstruction. To the best of our knowledge, this is the 1st time SGF mimicked a strangulated hernia.

SGF was diagnosed more common on the left side (97%) and 68% cases presented within 20 years of life. Most of them were diagnosed intraoperatively or after histopathological examination with only a few were diagnosed preoperatively. Ultrasound may show an extratesticular mass which is hypo- or isoechoic to the adjacent testis. In our case, a soft-tissue lesion with reduced vascularity was found in the inguinal canal, which we thought as either a strangulated omentocele or torsion of a supernumerary testis. In a suspected case, technetium-99-sulfur colloid imaging will help to recognize ectopic splenic tissue. Magnetic resonance imaging delineates soft-tissue and vascular characteristics; hence, it is valuable.

Complete excision of the splenic tissue with preservation of gonad is the treatment of choice. The splenic tissue can be easily dissected off the testis. Unfortunately, 37% of the reported cases underwent unnecessary orchidectomy as many surgeons were unfamiliar of SGF. In our patient, there was a fibrous cord connecting the splenule to the testis. The testis was saved, and SGF was excised as high as possible. Histopathology shows splenic tissue containing red and white pulp.

**CONCLUSION**

SGF, although rare, can mimic a strangulated inguinal hernia, uncomplicated inguinal hernia, undescended testis, torsion testis, or supernumerary testis. Every surgeon should be aware of this condition to avoid the dreaded morbidity of orchidectomy in this benign condition.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be...
reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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