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Case Report

Splenogonadal Fusion Presenting as Acute Torsion of Testis

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Abstract: Splenogonadal fusion (SGF) is a rare congenital anomaly noticed mainly in boys. The usual presentation is of testicular swelling and is managed most of the time with avoidable orchidectomy. Here we report a case of SGF in a one year old boy presenting with tender swelling on the left inguinal region and empty left scrotum. With the suspicion of torsion left inguinal testis, left inguinal exploration with laparoscopy was done as emergency procedure. It revealed left inguinal testis adherent to a torted segment of continuous type of spleenunculi which extended up to the spleen intra abdominally. Child underwent excision of this splenunculi with left orchidopexy. Pathology of the specimen was reported as torsion of splenunculi. Most of the SGF are incidental finding that are noticed during elective herniotomy or orchidopexy. But there are few cases reported in literature where SGF can present as an acute condition mimicking torsion of testis. A testis preserving excision of splenunculi should be done in these patients as there is usually a clear demarcating plane between them.

Keywords: Splenunculi, Torsion, Orchidectomy, Intra Abdominal

1. Introduction

Splenogonadal fusion (SGF) is a rare congenital anomaly noticed mainly in boys. It results due to abnormal connection between the spleen and the testis during embryonic stage. The usual presentation is of testicular scrotal swelling or an undescended testis. Most of the SGF are incidental finding that are noticed during elective herniotomy or during orchidopexy. But there are very few cases reported in literature where SGF can present as an acute condition mimicking torsion of testis. A testis preserving excision of splenunculi should be done in this scenario, as there is usually a demarcating plane between them. The procedure should be combined with laparascopy in the continuous type of SGF to excise the intraabdominal part of splenunculi.

2. Case Study

One year old boy presented with three days history of tender left inguinal swelling. There was no history of fever or trauma. On examination, a tender swelling of size 2x3cm was noted in left inguinal region with empty hypoplastic left scrotum. Right testis was also undescended and palpable high in the inguinal region. No other anomaly was noted elsewhere. Urgent ultrasound revealed left testis in continuity with a heterogeneous mass extending intra abdominally.

With the suspicion of Left inguinal torsion testis the child was posted for emergency surgery. Left inguinal exploration revealed a distal torted splenunculic part closely adherent to the normal left testis and a proximal splenunculic part extending intra abdominally. There was a short fibrous cord like tissue separating both these parts. Laparoscopy done

revealed the proximal part continuous with the spleen (classified as continuous type of SGF). The intraabdominal part of the splenunculi was disconnected from the spleen by ligasure through laparoscopy. By inguinal approach the distal part of splenunculi was completely dissected off the normal left testis and the whole specimen excised. The whole procedure was then completed by left orchidopexy.

Pathology of the excised tissue was reported as haemorrhagic necrosis consistent with torsion. Grossly there was a demarcating plane and capsule covering of the splenunculi and there was no evidence of malignancy.

The patient is being followed up post operatively and the left testis is showing adequate size for his age.

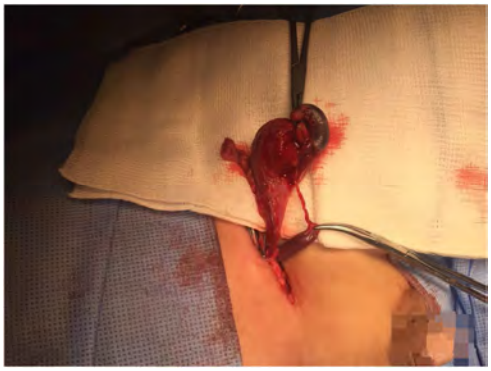

Figure 1. *Intrabdominal and inguinal part of continuous SGF separated by a short fibrous cord.*

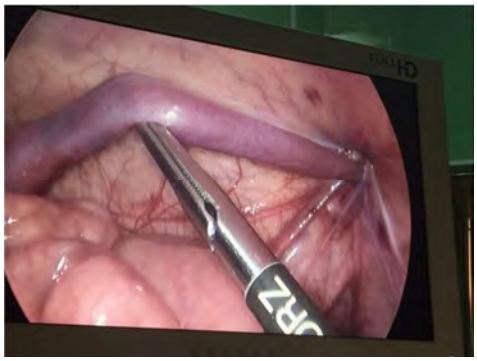

Figure 2. *Laparascopy showing the continuous type of splenunculi exiting the left internal ring.*

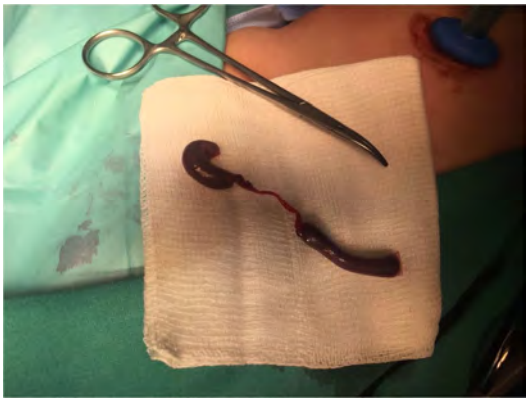

Figure 3. *Excised specimen of intra abdominal and inguinal part of splenunculi.*

3. Discussion

3.1. Classification

Splenogonadal fusion is an abnormal connection between the spleen and the gonad or derivatives of the metanephrons. It is a rare congenital anomaly first described by bostroem in 1883 and later in detail by pommer in 1889. In 1956 splenogonadal fusion was classified into continuous and discontinuous types by putschar and manian [1]. The continuous type is characterized by connection of the spleen and the gonad by a cord of splenic or fibrous tissue. Rarely, beads of splenic tissue are interspersed throughout the fibrous cord. In the discontinuous type, ectopic splenic tissue is attached to the gonad but has no connection to the normally located spleen. Accessory spleen is usually found within the tunica vaginalis and is closely attached to the gonad, although a distinct capsule is present [12].

3.2. Etiology

The spleen has an interesting embryological development process. Unlike most of the intra abdominal organs, it is not a derivative of the digestive tract. Clusters of mesenchymal cells on the dorsal mesogastrium are rapidly fused and vascularised to form the future spleen. The rotation of the stomach and the growth of the dorsal mesogastrium 6-7 weeks cause the translocation of the spleen from the midline to the left side of the abdominal cavity where a significant proximity to the primordial gonad is obtained. Therefore, splenogonadal fusion anomaly is considered to happen during 5-8 weeks of gestation. Three main mechanisms such as simple adhesion, inflammatory processes and teratogens are attributed [2] Sneath suggested that slight inflammation of the peritoneal surfaces over the spleen and gonadal ridge could produce partial fusion of the two organs. [3] Von Hochsteller postulated that a retroperitoneal path way for splenic analogue cells may allow contact with gonadal analogue. [4] McPherson et al argued that SGFLD is probably a developmental field defect with a hit occurring during blastogenesis and may be a polytopic field defect. They also stated that the earlier the abnormal event happens, the greater the number of anomalies that are produced as other developmental defects [2]. Loomis et al observed 351 embryos and found that the development of extremities is most intense in Carnegie phases 15-16 and that the developing spleen has a proximity to left mesonephros in phases 17-18. They suppose that SGFLD is therefore an early embryological defect. An association with the group of oromandibular-limb-hypogenesis syndromes or Hanhart complex also has been proposed because of the high frequency of orofacial abnormalities [2]. Another hypothesis is colonization of the phrenic ligaments by splenic cells in the first two months of gestation which is believed to create the splenic processus in continuous type cases. Therefore, this splenic process is the tip of a contiguous elongation of the parent organ extending downward in the continuous type. The discontinuous type, without any attachment to the spleen

proper, is thought to be just another manifestation of much more prevalent accessory spleens [11].

3.3. Clinical Presentation

This condition is considered benign and interestingly, more than 70% of reported SGF cases are in patients younger than 20 yrs. And half of these occur at less than 10 yrs of age. It occurs on the left side in 98% of the cases. [5] SGF has been also reported in female patients occasionally. [6] Approximately 50% of patients with continuous SGF have other congenital abnormalities like cryptorchidism, limb defects and microganthia. Less common associations are cardiac defects, cleft palate, imperforate anus and spina bifida. Discontinuous splenunculi are rarely associated with other congenital abnormalities.

SGF can clinically be very difficult to distinguish from a undescended testicle or an inguinal hernia. [7] The most common forms of presentation is of testicular swelling. Less frequently SGF may present as acute scrotum mimicking torsion testis. Unusual presentations like painful scrotal swelling secondary to malaria, mumps, exercise, leukemia have been documented. Bowel obstruction due to intraperitoneal cords, traumatic rupture of the ectopic splenic tissue and association with an intra-abdominal seminoma have been also reported. [8]

Grossly the accessory spleen is closely attached to the gonad and confined in the tunica vaginalis. The splenic tissue has a capsule like the native spleen and the splenic and gonadal tissue are distinct and seperate. Although one report has stated that these tissues were intermixed. The accessory spleen resembles normal spleen. [4] Regressive changes such as fibrosis, calcification and hemosiderin deposits may occur in these splenic tissue.

3.4. Investigation

Ultrasound is helpful in diagnosis of SGF. Doppler ultrasound of the scrotum for patients with a palpable testicular mass, has been used to diagnose preoperatively some cases of SGF. If the separation of the splenic tissue from the testis by the splenic capsule can be seen with sonography then clinical suspicion of SGF should arise. Some radiologists believe that it is possible to differentiate SGF from testicular cancer if they can identify a hypervascular mass on the upper testicular pole and then comparing its sonographic blood flow with the blood flow in the patient's own intra-abdominal spleen [13]. In continuous-type SGF it is possible to adequately ultrasound the surrounding splenic tissue and the vasculature originating from the splenic hilum. Colour Doppler may be used to provide additional information on the vasculature surrounding and originating from testes. Disorganized vascular branching from the testes may suggest testicular malignancy. However, all current sonographic case reports of SGF have concluded that such imaging is often nonspecific, because the results can be consistent with the more common testicular tumour. CT imaging with contrast can be used to aid the diagnosis of SGF by enhancing the visualization of the surrounding tissue and morphology. However, the radiation exposure associated with CT imaging of the groin and systemic contrast makes CT imaging less acceptable in clinical practice. The use of technetium-99m sulfur colloid scintigraphy has been suggested as the criterion to identify ectopic splenic areas. Such imaging appears to be particularly helpful to identify the discontinuous-type SGF; however, such confirmatory imaging may not available at all centres [12].

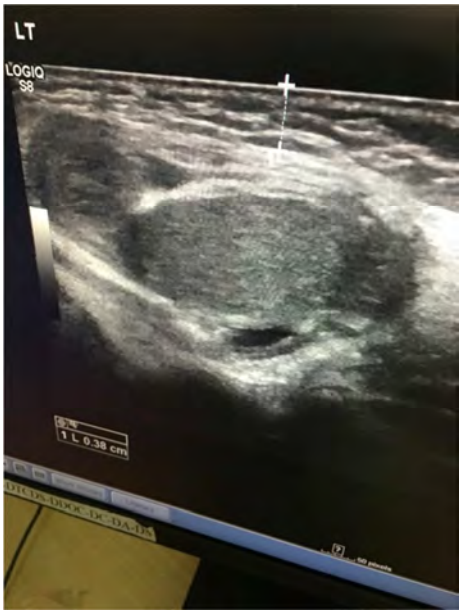

Figure 4. *USG –showing heterogeneous continuous swelling in inguinal region.*

3.5. Management

Complete excision of the torted splenunculi with preservation of testis and orchidopexy if testis undescended should be the ideal approach. In continuous type of SGF simultaneous laparoscopic exploration of the abdomen should be done to disconnect the proximal connection with the spleen.

4. SGF and Malignancy

Rarely there were cases of splenunculi associated with testicular cancer which needed orchidectomy. But till now no study has demonstrated a direct relationship between splenogonadal fusion and development of testicular cancer. [9]. In some instances patients were managed with radical orchidectomy when SGF was misdiagnosed as malignancy by radiology or during surgery. Splenogonadal fusion alone may not increase the risk of malignancy per se, but the frequent association with cryptorchidism explains the increased risk of testicular cancer. Cryptorchidism without SGF as such is associated with testicular cancer with a incidence of 1.7%, mainly when this is not treated by orchidopexy at early stages of life. But as of now in literature very few cases of testicular cancer are reported in association with SGF [14].

5. Conclusion

Thus from the case report we conclude that SGF is a benign condition and can present rarely with acute symptoms mimicking torsion of the testis. In this scenario, complete excision of the torted splenunculi with preservation of testis should be the ideal approach. In continuous type of SGF simultaneous laparoscopic exploration of the abdomen should be done to disconnect the proximal connection with the spleen.

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