CASE REPORT

Discontinuous-Type Splenogonadal Fusion in Abdominoscrotal Hydrocele: First Reported Case

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Abstract

Splenogonadal fusion is a rare congenital malformation, which is an abnormal connection between the spleen and gonads or mesonephric remnants. It usually presents with left cryptorchidism, scrotal mass or left inguinal hernia. Here we present the first case report of splenogonadal fusion as large left scrotal mass leading to abdominoscrotal hydrocele in a 2-year-old boy.

Keywords

Splenogonadal fusion; Abdominoscrotal hydrocele; Scrotal mass; Child

Introduction

Splenogonadal fusion (SGF) was first reported by Bostroem in 1883[1]. It is a rare congential anomaly where fewer than 200 cases have been reported until 2018[2]. Seventy percent of patients are less than 20 years old when diagnosed and fifty percent are less than 10 years old. Male to female ratio is 16.6:1[3], and almost always on the left side. The etiology is still unknown, but fusion occurs during embryogenesis, between the 5th and 8th weeks of gestation, as during the development on 5th week of gestation from the dorsal mesogastrium, and the gonadal ridge is formed at approximately the same time between the mesonephros and dorsal mesentery. During that time of primitive gut rotation, the spleen and the gonads (testis or ovary) get closer which might result in fusion before gonadal descend[4-6]. Similarly abdominoscrotal hydrocele is a rare condition where the tense hydrocele expanding up to external inguinal ring acquires intra-abdominal extension[7,8]. Here we present the first case report of discontinuous splenogonadal fusion as large left inguinoscrotal mass leading to abdominoscrotal hydrocele in two-year-old boy with further explanation of the mechanism of development of the intra-abdominal extension of hydrocele by the effect of the large splenogonadal mass.

Case Report

A two-year-old healthy boy referred to pediatric surgery clinic with left inguinal scrotal mass as case of testicular neoplasm (Fig. 1) which was discovered incidentally by the parents. They deny any history of trauma with no family history of testicular tumors. On physical examination there was a palpable large left testicular mass extending to spermatic cord which was thickened. It was firm, non-tender with no signs of inflammation. The right scrotum was empty, right testis was small and palpable in the superficial inguinal pouch. There were no palpable lymph nodes in the inguinal region. Laboratory investigations were unremarkable including the tumor markers. Abdominal ultrasound showed the abdominal spleen

is of normal size, shape and position. Other abdominal organs were unremarkable. Scrotal ultrasound showed right testis at medial part of inguinal canal. Diffuse left testicular mass (5x3x1.5 cm) and marked increase in vascularity on Doppler exam (Fig. 2). The left spermatic cord was thickened as well with increase in vascularity associated with left abdominoscrotal hydrocele (Fig. 3 and 4).

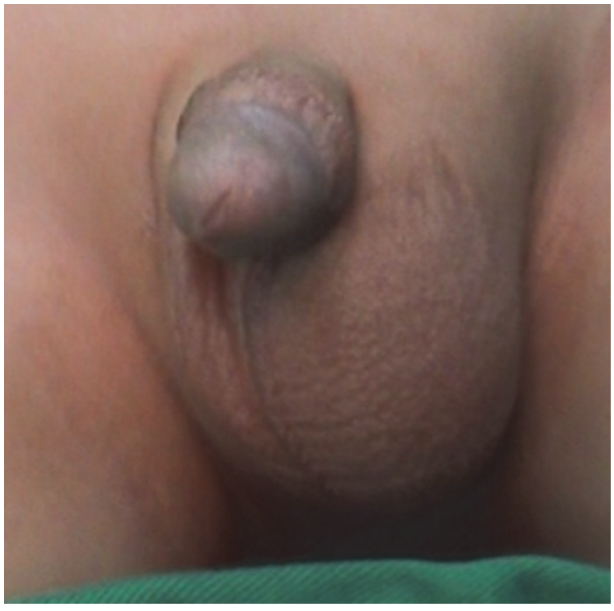

Figure 1. Left inguinoscrotal mass with empty right hemiscrotum (right undescended testis).

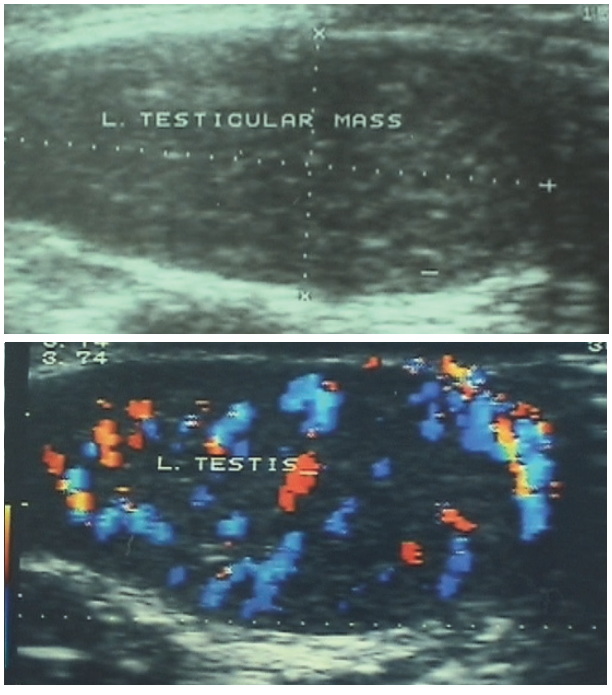

Figure 2. Ultrasound with Doppler of the scrotum showing very high vascularity all over the left inguinoscrotal mass in left hemiscrotum.

Intraoperative finding: the right testis was small and spermatic cord was short. Right orchidopexy was done. Left inguinal exploration showed a highly vascularized splenic mass attached to the upper pole of left testis enclosed in a hydrocele sac with intra-abdominal extension and moving upward in an opposite direction of an inguinal hernia (Fig. 4) this large splenic mass was receiving strongly large pulsating independent blood supply and the left testis supplied by the left testicular artery so the splenic tissue was remove with preserving the left testis which was fixed in the left hemiscrotum comfortably (Fig. 5), also small ectopic adrenal tissue was seen in the cord and

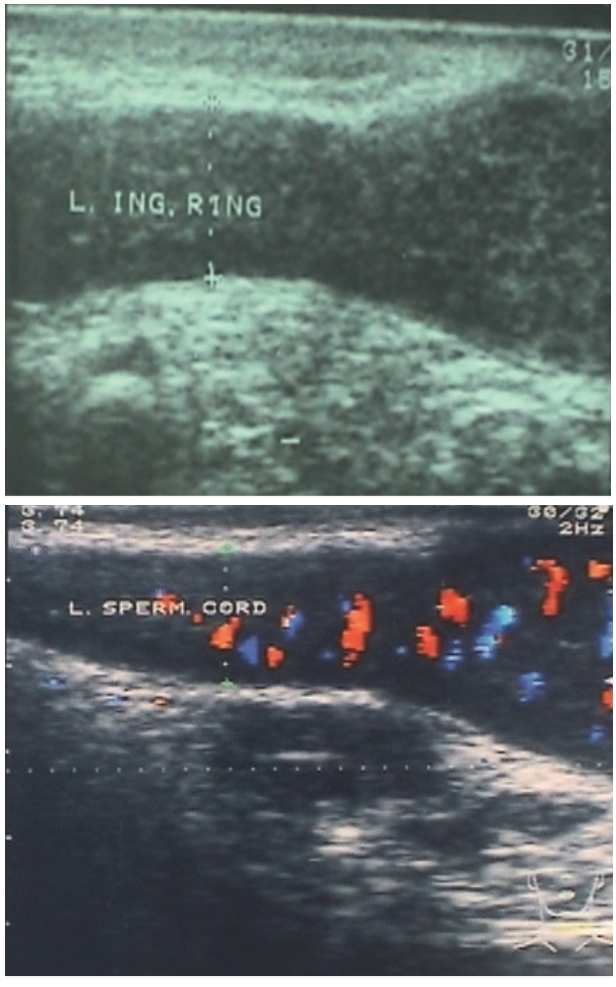

Figure 3. Ultrasound with Doppler showing the inguinal extension of the highly vascular mass.

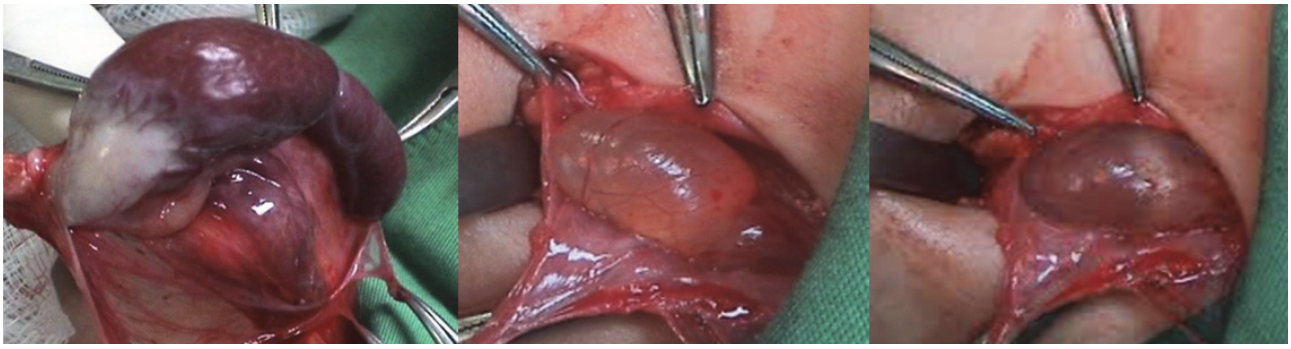

Figure 4. Splenogonadal mass enclosed in abdominoscrotal hydrocele sac no communication with the peritoneum (intra-abdominal extra-peritoneal). Photos showing (Left) fused spleen to upper pole of the testis. (Middle) opened inguinal canal with the intra-abdominal extension of the hydrocele (Right) the spleen moving to the intraabdominal part of the abdominoscrotal hydrocele.

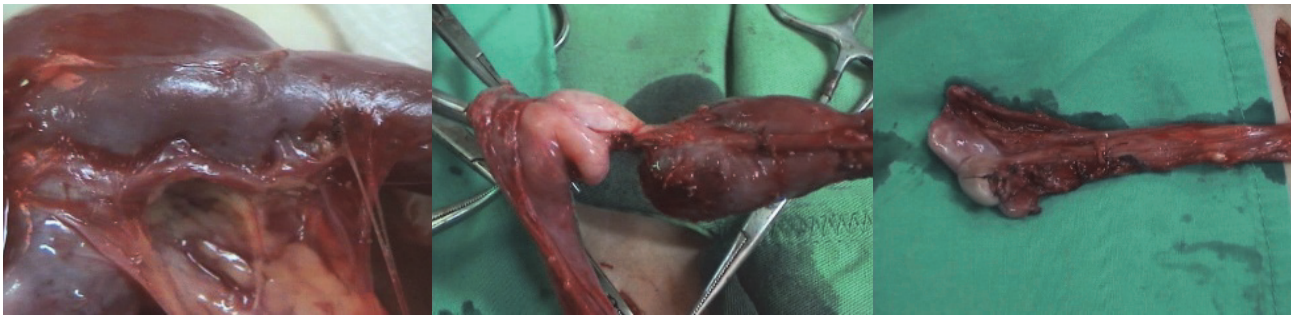

Figure 5. Intraoperative photos showing (left) splenic mass with prominent blood vessels (middle) separation of the splenic tissue from the upper pole of the testis (right) preserved left testis with its normal blood supply.

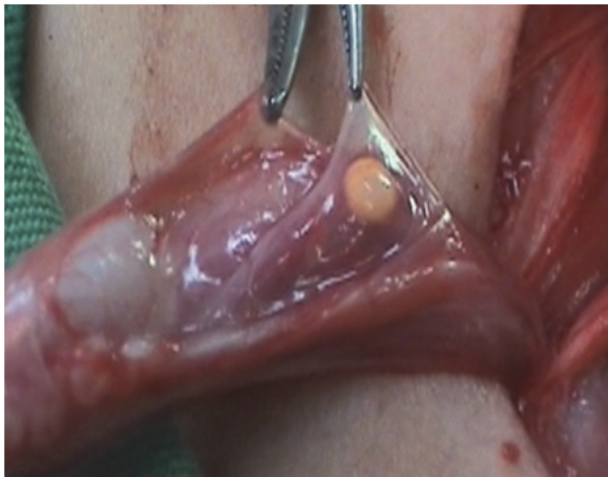

Figure 6. Adrenal tissue seen in the spermatic cord.

was excised (Fig. 6). Histopathology result confirmed that the mass composed of splenic tissue with no signs of malignancy.

The postoperative period was uneventful, and the patient discharged from the hospital on the second postoperative day. During the follow-up period, no complications occurred.

Discussion

Splenogonadal fusion was classified in 1956 by Putschar and Manion[9] into two types: Continuous and discontinuous. The continuous type characterized by direct connection between the spleen proper and the gonads with a fibrous band that contains splenic tissue and the discontinuous type an ectopic splenic tissue attached to the gonad and not to the native proper spleen as in our presented case and some other reports[10]. In a collective study of Carragher[11], of 123 cases analyzed, continuous SGF was found in 56% of the cases and the discontinuous variant in 44% of the cases. Approximately 50% of patients with continuous splenogonadal fusion have other congenital abnormalities which is five times more than the discontinuous variant, most commonly limb defects and micrognathia. Less common associations include cardiac defects, diaphragmatic hernia, cleft palate, imperforate anus and spina bifida[12]. Splenogonadal fusion is most often an incidental finding during groin exploration for cryptorchidism or inguinal hernia repair but may present as a scrotal mass as in our case. Occasionally, it presents as intestinal obstruction by intraperitoneal splenic fibrous

cord[13], or as acute scrotum[14] due to splenic rupture, spermatic cord torsion. According to a previous study, approximately 37% of cases reporting scrotal swelling are misdiagnosed as testicular tumors and result in unnecessary orchiectomy[15]. On reviewing the related international literature, there were no reported cases of SGF associated with abdominoscrotal hydrocele, clinical presentation of ASH is variable, it may present as a simple abdominoscrotal mass, or it may be discovered during management of an associated problem as in this present case study, or as a result of the pressure effect of a mass on the adjacent structure[8]. We believe that the splenic mass was the cause of increased pressure inside the hydrocele sac leading to the push up of its upper border to acquire the intra-abdominal extraperitoneal compartment. Reporting of association of splenogonadal large mass with abdominoscrotal hydrocele substantiate our previous explanation of the mechanism of development of the intraabdominal extension of the inguinal or inguinoscrotal hydrocele due to increase of the pressure in the hydrocele sac by external pressure of obstetrical cause (*e.g.,* fetal position, strong or prolong uterine contraction) or by internal mass, a mechanism similar to the one of the development of indirect inguinal hernia but in the opposite direction (*i.e.,* from scrotal compartment to abdominal compartment)[7,8].

Conclusion

Splenogonadal fusion is an embryological malformation with low incidence. Clinical diagnosis is rare but must be considered while evaluating inguinoscrotal pathology. Pre- or intra-operative diagnosis is mandatory to avoid unnecessary orchiectomy.

Isolated splenogonadal fusion has a good prognosis, we presented a case of discontinuous splenogonadal fusion presenting as large left inguinoscrotal mass leading to abdominoscrotal hydrocele.

Conflict of Interest

The authors have no conflict of interest.

Disclosure

The authors did not receive any type of commercial support either in forms of compensation or financial for this study. The authors have no financial interest in any of the products or devices, or drugs mentioned in this article.

Ethical Approval

Obtained.

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اندماج الطحال بالغدد التناسلية المحتواه داخل قيلة مائية صفنية بطنية: اول تقرير حالة

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المستخلص. الملخص اندماج الطحال بالغدد التناسلية من العيوب الخلقية النادرة الحدوث وتظهر بتصاحبها مع خصية معلقة او كتلة بكيس الصفن او فتق اربي في الجهه اليسرى .نستعرض هنا اول تقرير لحالة اندماج طحال مع الخصية اليسرى محتواة في قيله مائية صفنية بطنية في طفل يبلغ من العمر سنتين.