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Case Report

Splenogonadal Fusion Presenting as a Multiple Nodular Paratesticular Masses: A Case Report

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Received: October 19, 2021; **Published:** October 29, 2021

DOI: 10.31080/ecpe.2021.10.01050

Abstract

Splenogonadal fusion (SGF) is a rare congenital anomaly.

We report the case of a three and a half year- old boy with continuous type of SGF presenting as a multiple left sided nodular paratesticular masses.

This characteristic presentation can help increase awareness of this benign condition to avoid unnecessary orchiectomies in children.

Keywords: *Splenogonadal Fusion; Testis; Ectopic Spleen*

Introduction

A testicular mass in children should evoke the possibility of a malignant tumor. However most testicular tumors in children are benign [13].

SGF is one of the rarest causes of testicular masses discovered in children. It is a rare congenital anomaly defined by an abnormal connection between the primitive spleen and the gonad [8]. Preoperative diagnosis of this anomaly is exceptional.

This report describes a child presenting with two left sided palpable extra testicular masses discovered incidentally during an evaluation of vaginal hydrocele. The intraoperative findings appeared benign and the diagnosis of SGF was made histologically.

Case Report

A three and a half year- old boy was followed up for a left sided vaginal hydrocele diagnosed seven months ago. This hydrocele had spontaneously regressed.

Physical examination revealed two mobile contiguous painless nodular masses at the upper pole of the left testicle. Two normal sized intrascrotal testicles and a very small left-sided vaginal hydrocele were present. There were no signs of scrotal inflammation. No other physical abnormalities were detected.

Routine hematologic and biochemical analyses were normal.

A scrotal ultrasound revealed two extra-testicular rounded hypoechogenic and hypervascularized nodules of 4 and 6 mm respectively at the upper pole of the left testicle (Figure 1). No intra-abdominal lesions on abdominal ultrasound were found.

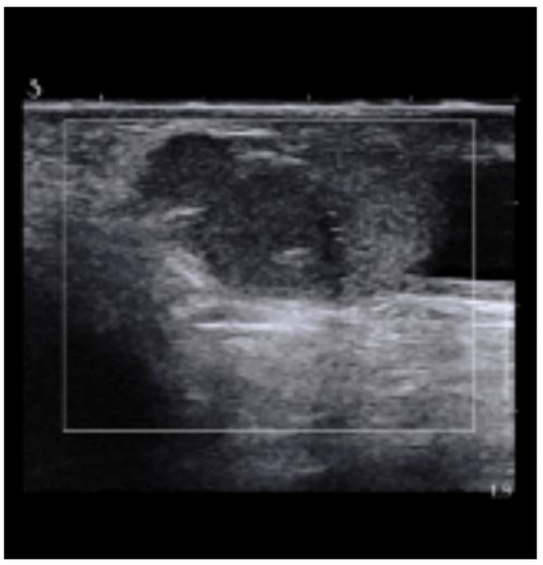

Figure 1: *Ultrasound image of the left testicle showing two well-defined hypoechogenic masses appended to the upper pole.*

Tumor markers were requested, revealing normal levels of alpha - foetoprotein, ß HCG and LDH. Chest X-ray was normal.

After six months of follow-up, the two palpable nodular masses had the same volume.

Based on these stable evolutionary clinical features and radiological characteristics of the two extra testicular small nodules, as well as the negativity of tumor markers these lesions were presumed to be benign and after validation in a multidisciplinary oncology team meeting, the decision was made to perform a surgical scrotal approach.

We opened the tunica vaginalis and exploration revealed several purple nodules. The largest one was a 6 mm nodule fused to the upper pole of the testicle and extending parallel to the spermatic cord with a fibrous cord beaded by other infracentimetric nodules (Figure 2 and 3).

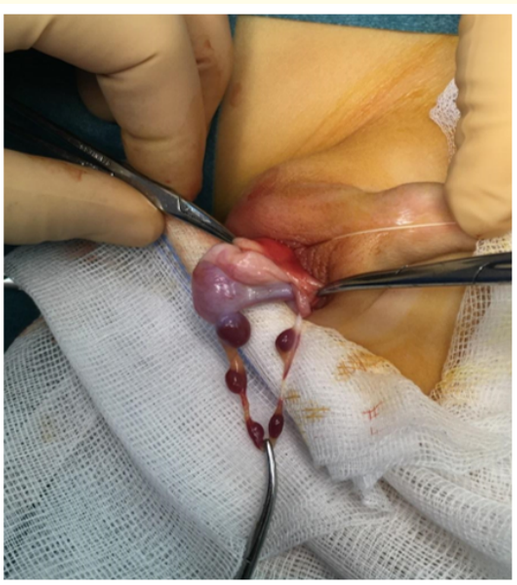

Figure 2: *Intraoperative view of SGF with the six nodules organized along a fibrous cord.*

Figure 3: *Resected specimen.*

The albuginea was incised to excise the largest lesion. This lesion was completely removed from the testis without any damage to the testis, vas deferens, vessels, or epididymis.

An inguinal incision was also performed to facilitate removing all the other lesions and to check their extension.

The fibrous cord measuring 11 cm in length containing 6 nodules was completely independent of the spermatic cord. These lesions were excised after division of the fibrous cord of the vessel pedicule (Figure 3). The cord was followed until its fusion with the peritoneum at the level of the deep inguinal orifice. We checked the absence of communication of this fibrous cord with the intraperitoneal cavity.

Histologic examination of the operative specimen revealed splenic tissue without any signs of malignancy (Figure 4). These findings were consistent with a diagnosis of continuous-type SGF.

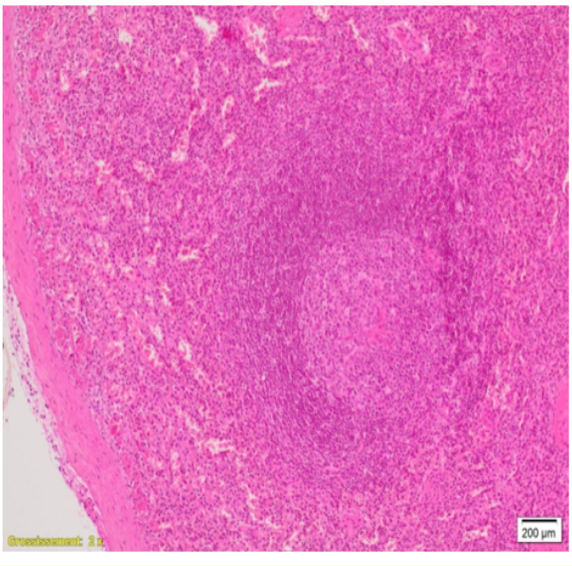

Figure 4: *Histopathological picture showing splenic tissue.*

Postoperative course was uneventful. The child was discharged home the same operative day.

Early and twenty two months postoperative follow-up revealed a left testicle well positioned in the scrotum with no palpable masses.

Discussion

SGF is a rare, benign, congenital anomaly consisting of abnormal fusion between the spleen and the gonad or the remnant of the mesonefros, almost always presenting on the left side [10].

It was first described by Bostroem in 1883, and approximately 200 cases have been reported in the literature [2,6,11].

The exact pathogenesis of SGF anomaly is unknown, however it is postulated that SGF results from the development of an abnormal connection between the gonad and spleen during approximately the 5th and 8th weeks of embryonic life, when the organs are in close proximity to each other. When gonadal descent begins, the attached splenic tissue follows the gonadal path [1].

SGF is divided into continuous and discontinuous types [12].

In the continuous type there is a direct anatomical attachment between the spleen and the 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 [12].

In the discontinuous type, there is no direct attachment between these two organs [12].

Our case is considered as a continuous type. Both types occur with equal frequency [9].

Continuous-type SGF is more commonly associated with other congenital anomalies, including cryptorchidism, limb defects, cardiac defects, micrognathia, cleft palate, anal atresia, and spina bifida. Of these malformations, limb defects are most common and is termed splenogonadal fusion-limb defect syndrome [3].

The age of presentation is less than 10 years in half of the reported cases, and 82% of the cases occur in patients younger than 30 years [1].

Occurrence in females is very rare with a male:female ratio of 15:1 [9]. This difference is likely due to under diagnosis in females due to the intraabdominal location of the gonad [3]. SGF is rarely diagnosed or suspected preoperatively. SGF is most commonly seen incidentally during inguinal exploration for cryptorchidism, hernia, hydrocele or left scrotal mass [4,6]. The presentation may also be due to complications of the fusion, e.g. bowel obstruction, acute painful scrotal splenic enlargement or rupture [8,9].

Scrotal ultrasound is not sufficiently accurate preoperatively [8]. It typically reveals an encapsulated iso- or hypoechogenic, homogeneous extratesticular mass that is more hypervascular at Doppler US when compared to the surrounding testis [6]. Some authors report that this Doppler feature allows to differentiate between testicular cancer and SGF with a blood flow in the upper testis lesion similar to that of healthy splenic tissue [4,5]. In our case, ultrasound founded two small extra testicular hypoechogenic and hypervascularized infracentimetric lesions.

Computed tomography and magnetic resonance may detect the connecting fibrous tissue in the continuous form [4,6]. This diagnosis should be considered when the imaging show such features.

When suspected clinically, technetium-99m DTPA isotope scanning is used to detect accessory splenic tissue [11].

Often, the definitive diagnosis is made postoperatively by histology as in our case [9].

Orchidectomy is not necessary as the splenic component can be dissected and separated from the gonad or from the spermatic cord elements as in our case [9].

In a review of 137 cases of SGF, 37% of the patients with SGF underwent an unnecessary orchiectomy for a suspected primary testicular neoplasm [12].

There are about seven reported cases of SGF and testicular malignancy. In all of these cases, the malignancy developed in adults with undescended testes or following orchidopexy for undescended testes. Thus it is rather cryptorchidism that may increase the risk of cancer [2,5,7,12].

For some authors, non-operative management may be recommended if the anomaly is recognized preoperatively and is asymptomatic clinically. But others consider that excision of the ectopic splenic tissue is essential to prevent testicular atrophy, infarction or torsion and preserve fertility [7].

The presence of fibrous cord beaded with 6 nodules in our case, in addition to the clinical evolutionary characteristics and radiologic features, was very specific and suggestive of the diagnosis and allowed us, to presume the benign nature of these lesions and to favour a conservative approach.

This typical macroscopic aspect suggestive of this malformation as it was in our case enabled us to avoid an orchiectomy.

This testicle-sparing surgery should be considered in SGF especially in case of testicular masses in children in which the epidemiologic, histologic and prognostic features are different from cases in the postpubertal age.

However, malignant tumor especially rhabdosarcoma should be a diagnostic consideration in cases of para testicular masses in children. When in doubt, inguinal approach with isolation and occlusion of the spermatic cord should be recommended and frozen biopsy is indicated [11].

This conservative approach must be performed under the strict usual criteria of tumorectomy (small size under 2 cm and negative tumor markers) [13].

Conclusion

The diagnosis of SGF prior to surgery is challenging.

This rare diagnosis should be kept in mind in case of a testicular mass in children, especially if the mass is on the left side.

Through this observation, we highlight the multiplicity of lesions as an additional characteristic to the other known benign characteristics which could well lead to this diagnosis in order to avoid unnecessary orchiectomy in children. Taking into consideration the clinical, radiological, biological, evolutionary and peri- operative features of this rare anomaly, ligation of the spermatic cord should be decided on a case-by-case basis which calls into question the classic "dogma" of systematic ligation of the spermatic cord and orchiectomy for any scrotal tumor in children.

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Volume 10 Issue 11 November 2021