Rare Case of Continuous and Discontinuous Splenogonadal Fusion in A single Testicle

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ABSTRACT

Splenogonadal fusion is a rare congenital malformation that involves an abnormal connection between the spleen and the gonad. It manifests itself as a mass consisting of splenic and testicular or ovarian tissue. Splenogonadal fusion has been classified into two types; continuous, where there is a direct connection between spleen and gonad; and discontinuous, where ectopic splenic tissue is attached to the gonad, but there is no connection to the spleen. Many cases had an associated other anomalies either genital or systemic.

Index Word: Splenogonadal, accessory spleen, and testicle.

INTRODUCTION

Splenogonadal fusion is a rare benign congenital anomaly thought to occur between the 5th and 8th gestational weeks. The etiology of such anomaly is uncertain, but may be secondary to injury of the fetus during this time. The abnormality is virtually always on the left side where immature splenic tissue adheres to the developing gonad, epididymis or vas deferens. The splenic tissue is subsequently pulled in a caudal direction with descent of the gonad.  

The anomaly was first mentioned by pathologist Eugen Bostroem in 1883, but it wasn't until 1888 when Pommer described the malformation in details.  

Putschar and Manion classified splenogonadal fusion into two types: continuous (direct connection between the spleen and gonad) and discontinuous (no anatomic connection between ectopic splenic tissue and the principal spleen), each form occurring with equal frequency.  

One-third of cases are associated with other congenital abnormalities including limb and orofacial malformations which are more likely to associate the continuous form.  

In this presented case there is combination of both continuous and discontinuous splenogonadal fusion in a precious single left testicle, which make the option of orchiectomy impossible and make the decision of excision of the ectopic splenic tissue and preserving this testicle rational.

CASE PRESENTATION

A 9 months-old boy was referred with a history of a right undescended testicle. On physical examination, his abdomen was normal without evidence of any organomegaly. His genitalia revealed a normal phallus with an empty well formed right hemiscrotum, but in left side the testicle felt in normal position, but it is bulky and at its upper pole there is a small mass not attached to it, not tender, no hernia
can be detected in this side. There were no other abnormalities. Urinalysis and serum beta-hCG, AFP and LDH were normal.

An ultrasound revealed that there was no shadow of the right testicle in the scrotum, inguinal region or the retroperitoneal space. There was a small amount of fluid around the left testis, its epididymis was enlarged, the whole testicle was 20X 5X 9mm.

There was a mass above the upper pole of the testicle about 7.9 X5.7 mm with smooth outline, regular echogenicity with no calcifications. There was prominent flow involving the entire mass noted with power Doppler. Spectral Doppler showed two components of vascular supply, one with a lower peak systolic amplitude and one with higher peak systolic amplitude. There was a significant venous flow detected within the mass which had the echotexture and similar flow characteristics of the testis. Technetium study was not done.

The diagnosis of left crossed testicular ectopia was made according these data. The child underwent a left inguinal exploration with delivery of his testis and the cord, the testis was completely extra peritoneal, with unusual long vas and a patent processes vaginalis enclosing a thin film of peritoneal fluid, and a small mass at its end near the external inguinal ring. The testicle itself looked fleshy, highly vascular and the epididymis was slightly separated with wide sinus. (Fig 1)

When the processes vaginalis was opened, the small mass was completely intra peritoneal, connected to the peritoneal cavity with a vascular pedicle. (Fig 2). At this stage, a decision was made to take frozen sections from both testicle and the mass, the result came for both to be a splenic tissue. (Fig 3)

The small mass(accessory spleniole) was removed as much high as possible with ligation of its pedicle. Exploration of the right side revealed no testicle or vas.

Fig 1: shows the tumor which is attached to the posterior rectus sheath. Note the stomach and liver appear grossly normal

Fig2:- The accessory spleniole coming from inside the peritoneum with its vascular stalk

Fig3:- Histopathological picture showing splenic and adjacent testicular tissue
The process vaginalis was transfixed after dissection of the vas and testicular vessels from it, and as this the testicle was single, so its tunica was opened and most of the splenic tissues was shaved from its surface and the tunica then was closed with fine 6/0 vicryl stitches.

The baby went through an uneventful postoperative course and he will be followed up with $^{99m}$Tc-sulfur colloid scanning to detect any splenic tissue in the single lift testicle.

**DISCUSSION**

The spleen appears as a thickening of the layers of splenic mesothelium of the dorsal mesentery of the stomach. The testis, arising as an indifferent sex gland from the medial surface of the Wolffian body, comes to lie in front of the primitive kidney on the posterior wall of the abdomen. The two organs are thus brought into close apposition, and fusion of the two is not inconceivable. Under such circumstances the testis, in its descent during the latter weeks of intrauterine life, might readily drag along a tail of developing splenic tissue.

Splenogonadal fusion is a rare congenital anomaly whereby the left gonad is typically fused to a segment of splenic tissue. A single case of splenogonadal fusion occurring on the right side has been reported. The first report of splenogonadal fusion was by Boestroen in 1831 and detailed by Pommer in 1889. In 1956, Putschar and Manion proposed a classification in which they grouped splenogonadal fusion into 2 types. In the continuous type, there is a cord-like structure connecting the orthotopic intraperitoneal spleen to the gonadal mesonephric structure. In the discontinuous type, the accessory and orthotopic spleens are not connected.

There have been at least 150 cases of splenogonadal fusion reported in the literature. The largest review was made by Carragher et al. in 1990, describing 123 cases.

Patients most commonly present with cryptorchidism, left inguinal hernia or left scrotal mass. Presentation may, however, be due to the complications of the fusion, e.g. bowel obstruction, acute painful scrotal splenic enlargement or rupture. Often, the definitive diagnosis is made postoperatively by histology; and orchiectomy is not necessary as the splenic tissue can be dissected off the testicle. A male to female ratio of approximately 16:1 was noted. The continuous type comprised 55%. The diagnosis is most often made in the first two decades of life (72% to 82%), with 50 percent of the cases diagnosed prior to 10 years of age. Karaman and Gonzales noted that an unsuspected diagnosis during operative exploration may have lead to an unnecessary orchiectomy in one third of the reported cases. In only 4 previously reported cases was the diagnosis suspected preoperatively, including 2 cases confirmed by radionuclide scanning, using radionuclide sulfur colloid imaging to demonstrate splenic uptake by the mass.

Given that the most common clinical presentation of splenogonadal fusion is a scrotal mass, ultrasonography will likely be the first imaging study to evaluate this condition. The literature review revealed few cases where an ultrasound examination has been performed for splenogonadal fusion. However, it has been stated that a vascularized cord-like structure connecting the spleen to the splenogonadal mass should be investigated for splenogonadal fusion.

The differential diagnosis of the intra-scrotal paratesticular mass in children is broad, but includes malignant tumor, adrenal rest tissue, epididymitis, orchitis, hemangioma, and hematooma. Testicular tumors are typically irregular in echotexture and vascularity and the majority are intratesticular. The extent of vascularity observed by Doppler ultrasonography of splenogonadal fusion is more extensive than any typical paratesticular or testicular tumor and may easily be compared to the vascularity of the patients own intraabdominal spleen.

A careful examination of a left scrotal mass with Doppler ultrasonography is likely to differentiate splenogonadal fusion from a tumor. A Doppler ultrasound finding of an echogenic hypervascular lesion situated on the superior aspect of the testis may lead to the clinical impression of a splenogonadal fusion.
Although this malformation is rarely diagnosed preoperatively, surgery is usually necessary to determine if it is malignant. Historically, there have been several cases where a testicular swelling has resulted in an unnecessary orchiectomy due to suspicion of a testicular neoplasm. Anyway, the splenic tissue can be severed from the tunica albuginea in order to prevent orchiectomy, but in some case, orchiectomy was necessary because the splenic tissue was adherent to the testis making it difficult to separate. Scrotal exploration can be avoided in a few cases if preoperative scanning using 99mTc-sulfur colloid is done.9

The presented case was diagnosed preoperatively by clinical examination, ultrasonography and colored Doppler to be a case of crossed testicular ectopia, but on operative exploration the splenic tissue was found to be embedded in the left single testicle under its tunica which make it a case of continuous splenogonadal fusion, but the presence of another concomitant splenic tissue inside the processes vaginalis considered as a accessory spleniole or a combined discontinuous splenogonadal fusion.

**CONCLUSION**

Splenogonadal fusion should be considered in any baby presented with left sided testicular swelling and ultrasound alone is not enough to diagnose such cases. Frozen section is crucial if splenogonadal fusion is suspected in patient with single testicle, which should be preserved with strict follow up, where the ultimate aim should be preservation of healthy testicles.

**REFERENCES**