Introduction

Splenogonadal fusion is a rare congenital anomaly that results from abnormal coalition of splenic and gonadal anlagen in utero. Typically, this entity presents as a testicular mass, and the diagnosis of this condition is seldom made preoperatively. Awareness of this entity is necessary because 37% of cases reported in the literature are associated with unnecessary orchiectomy [1]. This anomaly has been reported in the pediatric and urologic literature [1–3] but only rarely in the radiologic literature [4, 5]. To our knowledge, this is the first sonographic case of splenogonadal fusion reported in the radiology literature. We discuss the ultrasound findings of this condition and review its etiology.

Case report

A 5-month-old boy was evaluated for a left scrotal mass. On physical examination, a firm 1-cm mass was palpated above a normal left testis. The right testis was smaller than the left testis but felt normal. No scrotal changes were identified, and a normal phallus was present. Laboratory examinations were normal except for a mild decrease in the patient’s hemoglobin level and a mildly elevated alpha-feto protein level at 44.5 ng/ml (normal range 0.0–30.0 ng/ml). A scrotal sonogram was performed, which demonstrated a bilobed appearance to the left testicle (Fig. 1), thought to be most consistent with testicular duplication as opposed to extratesticular neoplasm. No other sonographic abnormality was identified.

Surgical exploration was performed through a left inguinal incision. A solid mass with a fibrous band extending into the peritoneum was completely excised from the superior pole of the left
testis. The remainder of the left testicle appeared normal, as did the epididymis. Grossly, the mass consisted of a well-encapsulated red-brown soft tissue measuring 5 × 5 × 10 mm (Fig. 2). Frozen and permanent sections showed the mass to be ectopic splenic tissue. Final diagnosis was splenogonadal fusion.

Discussion

Splenogonadal fusion is usually identified serendipitously during inguinal hernia repair or orchiopexy [1]. Preoperatively, most symptomatic patients are thought to have a tumor, epididymitis, testicular duplication, or torsion [2]. Surgical exploration is generally needed to rule out malignancy. However, orchiectomy is generally not indicated because the splenic tissue can be dissected away from the tunica albuginea in most cases [1]. The sonographic appearance in our case was of a homogeneous left extratesticular mass with echogenicity equal to that of normal testis. The mass was well encapsulated and was not associated with hyperemia or scrotal skin thickening. Differential diagnostic possibilities include extratesticular neoplasm, polyorchidism, and splenogonadal fusion. Although ultrasonography can reliably demonstrate the extratesticular location of a palpable mass, it cannot provide a definitive histologic diagnosis. Scintigraphy with a \(^{99m}\)Tc sulfur colloid liver-spleen scan can identify ectopic uptake within the scrotum or tail of functioning splenic tissue emanating from the spleen to the testis [4], thereby adding specificity to the imaging diagnosis. However, surgical investigation is typically warranted.

Splenogonadal fusion was first described by Bostroem in 1883 and then published by Pomer in 1889 [6]. Splenogonadal fusion is thought to occur between the 5th and 8th weeks of gestation, before gonadal descent [1]. The splenic anlage forms in the left dorsal mesogastrium approximately during the 5th week of gestation. At this point, the anlage consists of multiple small aggregates that later coalesce to form the spleen. At the 6th to 7th week, the stomach translocates to the left side of the abdominal cavity, causing the splenic anlage to be adjacent to the left urogenital fold, which includes the gonadal mesoderm. Fusion of these two primitive organs may occur during caudal migration. The pathogenesis is unknown, but two hypotheses exist. One postulates that mild inflammation on the peritoneal surface near the spleen and gonadal ridge results in partial fusion of the two organs before gonadal descent. The other suggests that a retroperitoneal pathway of the splenic anlage cells allows contact and fusion with the gonadal anlage [1].

Two different patterns of fusion exist: continuous and discontinuous. Continuous splenogonadal fusion consists of ectopic splenic tissue connected to the spleen by a fibrous band or cord, as was seen in our case. The cord can be composed of splenic or fibrous tissue or a mixture of both. In most cases, the cord usually attaches from the upper pole of the orthotopic spleen to the testis, although the cord can attach to the epididymis, ovary, or mesovarium. The continuous form of splenogonadal fusion is often associated with other congenital anomalies [1, 3, 6] and cryptorchidism [2]. With the discontinuous form, the ectopic splenic tissue is attached to the gonad, but no connection with the orthotopic spleen is identified. The ectopic spleen is usually found