Discontinuous type of splenogonadal fusion syndrome with limb defects

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ABSTRACT

Splenogonadal fusion is a rare congenital anomaly. We report a case of discontinuous type of splenogonadal fusion with congenital malformation. We focused here to avoid unnecessary orchidectomy by cementing the base of preoperative scrotal ultrasonography, for paratesticular masses.

Key words: Orchidectomy, paratesticular mass, splenogonadal fusion, syndactyly, ultrasonography

INTRODUCTION

Splenogonadal fusion is a rare polytopic condition of abnormal fusion between the spleen and the gonad or remnant of the mesonephros. The fusion may be continuous or discontinuous type. The diagnosis is usually made at operation or at autopsy related to paratesticular scrotal mass assuming a malignant nature. Preoperative diagnosis may help to prevent unnecessary invasive surgery. Here we focused the scrotal ultrasonographic finding of splenogonadal fusion.

CASE REPORT

An 18-years-old male patient presented for evaluation of a large left scrotal swelling since birth which was progressively increasing in size. He was mentally retarded and syndactyly detected over the right middle and ring finger with right lower limb shortening. Local examination of left scrotum has testicular mass measuring 8.0 × 5.0 cm, pyriform in shape with smooth in outline. Left testis was normal. Scrotal ultrasound revealed a well-defined hypoechoic, homogenous mass of size -8.0 × 5.0 cm with ample of vascularity noted at upper part of left testis [Figures 1 and 2]. On abdominal ultrasonography pancreas, spleen, kidneys, and other organs were normal. No evidence of communicating fibrous cord was noted in intraperitoneal or extraperitoneal region.

With clinical correlation and sonological findings provisional diagnosis of splenogonadal fusion limb defect syndrome was made.

On intraoperative findings pyriform shaped mass lesion of size 7.0 × 3.0 cm seen attached to the upper pole of left testis. Mass was found to be separate from the left testis [Figure 3]. Histopathologic examination of the mass revealed a fibrous capsule and normal splenic tissue.

DISCUSSION

Splenogonadal fusion is a rare congenital anomaly first seen...
reported in 1883 by Boestrom. In 1996, Karaman and Gonzales\(^1\) published a published work review including 137 cases of splenogonadal fusion. Putschal and Manion\(^2\) classified splenogonadal fusion into continuous and discontinuous types. 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 tissue is attached to the gonad, but has no connection to the spleen.\(^3\) The accessory spleen is usually found within the tunica vaginalis and is closely attached to the gonad, although a distinct capsule is present. Splenogonadal fusion is assumed to occur between 5 and 8 weeks of gestation before the beginning of gonadal descent.\(^4\) Its cause remains unclear, but two theories predominate. First,\(^5\) slight inflammation of the peritoneal surfaces over the spleen and gonadal ridge can produce partial fusion of the two organs, while second\(^6\) has postulated that a retroperitoneal pathway for splenic anlage cells may allow contact with the gonadal anlage. It has therefore been proposed that the discontinuous type may represent a rare variant of an accessory spleen.\(^2,4\) Most patients (82%) reported are younger than 30 years old and mostly male.\(^3\) However, the disorder has been described in patients from birth to age 81 years.\(^7\)

Splenogonadal fusion has been associated with other congenital anomalies, especially the continuous type. The most common anomalies are limb defect and micrognathia.\(^3\) Less common anomalies are cardiac defect, cleft palate, spina bifida, facial muscle agenesis and malformation of the anus. In the discontinuous type, few anomalies have been described.\(^1,3\) Splenogonadal fusion is most commonly an incidental discovery during a routine groin exploration for cryptorchidism or hernia. Of the reported cases, 16.8% have been diagnosed at autopsy.\(^1\) Because of the rarity of this condition, it is rarely diagnosed preoperatively. Techniques of diagnostic imaging are available if there is a clinical suspicion of splenogonadal fusion. The most reliable preoperative imaging, according to published results, is the 99mTc-sulfur colloid liver spleen scan, which detects accessory spleen.\(^3\) Unaware of the nature of left scrotal masses, many surgeons have sacrificed an intact testis because they lacked a proper preoperative diagnosis.\(^3\) Surgeons ignorant of the nature of the scrotal mass and fearing malignant degeneration have performed radical orchidectomy on salvageable testes.

**CONCLUSION**

Splenogonadal fusion is a rare entity in which discontinuous type with limb defect is extremely rare (as in our case). It should be considered in the differential diagnosis of scrotal masses especially when it occurs with congenital anomalies. It is usually mistaken for a testicular tumor. Preoperative and intraoperative diagnosis may prevent unnecessary orchidectomy.

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