

Spleno-gonadal fusion

Hock Beng CHUA

Department of Surgery, RIPAS Hospital, Bandar Seri Begawan, Brunei Darussalam

ABSTRACT

Spleno-gonadal fusion is a rare malformation consisting of an abnormal connection between the spleen and the gonad. Spleno-gonadal fusion has been classified into two types: continuous and discontinuous. The majority of cases present as a scrotal mass, scrotal tenderness or as clinical suspicion of testicular pathology. Some cases present as incidental findings during herniotomy or orchidopexy procedures. About 25% of the reported cases of continuous spleno-gonadal fusion have associated anomalies. No anomalies have been reported in discontinuous spleno-gonadal fusion. An incidental case of discontinuous spleno-gonadal fusion in a three year old boy after a patent processus vaginalis ligation procedure is reported.

Keywords: Orchiectomy, spleen, spleen abnormalities, testicular neoplasm

INTRODUCTION

Spleno-gonadal fusion is a rare malformation consisting of an abnormal connection between the spleen and the gonad. Putschar and Manion classified spleno-gonadal fusion as continuous and discontinuous. In the continuous type, the principal spleen is connected to the gonad by a cord like structure that may be fibrous, totally splenic or beaded with multiple splenic nodules. In the discontinuous type there is no connection between the main spleen and the gonad. The majority of cases present as a scrotal mass, scrotal tenderness or as clinical suspicion of testicular pathology. Some cases can present as incidental findings during herniotomy or

orchidopexy procedures. About 25% of reported cases of continuous spleno-gonadal fusion have associated anomalies, however this has not been reported for the discontinuous type. An incidental case of discontinuous spleno-gonadal fusion in a three year old boy after a patent processus vaginalis ligation procedure is reported.

CASE REPORT

A three year old boy presented with a one month history of a bluish cystic swelling along the left spermatic cord which was non-tender. After thorough examination, a differential diagnosis of encysted hydrocele was considered. He was otherwise well with no other abnormalities detected. He underwent ligation of left patent processus vaginalis procedure and was found to have a 1 cm bluish lesion

Correspondence author: Hock Beng CHUA
Department of Surgery, RIPAS Hospital,
Bandar Seri Begawan BA 1710,
Tel+673 2242424 Ext 6278
E-mail: chbtln@hotmail.com

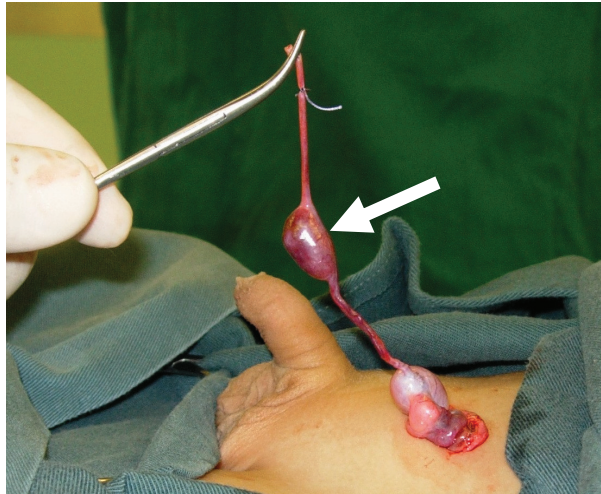


Fig. 1: Left continuous spleno-gonadal fusion anomaly (arrow).

attached to the left testis by a fibrous cord, a separate vessel along the spermatic cord and a patent processus vaginalis (Figure 1). The patent processus vaginalis was dissected and transfixed at deep ring. The lesion was excised after division of the fibrous cord and vessel pedicle. The histology of the lesion showed splenic tissues confirming the diagnosis of a discontinuous type of spleno-gonadal fusion (Figure 2). Post-operatively, he made an uneventful recovery and follow-up ultrasound scan did not reveal any other congenital anomalies.

DISCUSSION

It is estimated that only 150 cases of spleno-gonadal fusion have been documented. Although the anomaly was first mentioned by the prominent pathologist Eugen Bostroem in 1883, it was not until 1889 when Pommer described the malformation in detail. Spleno-gonadal fusion limb defects micrognathia is listed as a rare disease by the Office of Rare Diseases (ORD) of the National Institutes of Health (NIH). This condition affects less than 200,000 in the US population. This is a rare genetic condition characterised mainly by a

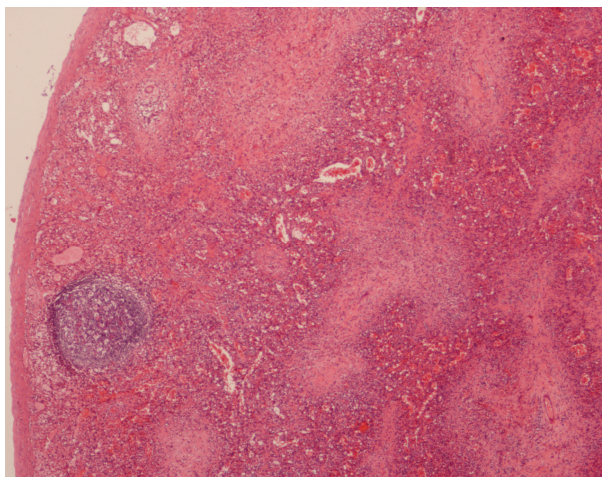


Fig. 2: Histology showing splenic tissue (H&E stain, x10).

small jaw, limb defects and fusion of the spleen to the gonads. Most patients die shortly after birth although some do survive into childhood.

Although spleno-gonadal malformation affects both sexes, it is more common in males. Cryptorchidism is the most common associated anomaly. About 25% of the reported cases of continuous spleno-gonadal fusion have associated anomalies such as micrognathia, anal atresia, asymmetry of the skull, or abnormal fissures of lung and liver. Other associated anomalies include bilateral absent legs, imperforate anus, spina bifida, diaphragmatic hernia and hypospadias.¹⁻⁵ Preoperative diagnosis of this rare condition is difficult and rarely achievable. Ultrasound⁶ and technetium-99m-sulfur colloid scan² have been suggested as preoperative tests but the results may not be reliable.

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 orchidectomy due to suspicion of a testicular neoplasm. In order to prevent an orchidectomy, the splenic tissue can be severed from the tunica albuginea.

In conclusion, this case demonstrated the discontinuous type of spleno-gonadal fusion. Pre-operative diagnosis was difficult and the patient had the splenic tissue excised separately preserving the testis. The patient recovered with a satisfactory outcome and post-operative imaging did not reveal other associated anomalies.

REFERENCES

- 1:** Cortes D, Thorup JM and Visfeld J. The pathogenesis of cryptorchidism and splenogonadal fusion: a new hypothesis. *Brit J Urol* 1996; 77: 285-90.
- 2:** Guarin U, Dimitrieva Z, Ashley SJ. Splenogonadal fusion - a rare congenital anomaly demonstrated by 99mTc-sulfur colloid imaging: case report. *J Nucl Med* 1975; 16: 922-4.
- 3:** Carragher AM. One hundred years of splenogonadal fusion. *Urology* 1990; 35: 471-5.
- 4:** Lipson AH. Amelia of the arms and femur/fibula deficiency with splenogonadal fusion in a child born to a consanguineous couple. *Am J Med Genet* 1995; 55: 265-8.
- 5:** Gouw ASH, Elema JD, Bink-Boelkens MTE, de Jongh HJ, ten Kate LP. The spectrum of splenogonadal fusion: case report and review of 84 reported cases. *Eur J Pediatr* 1985;144:316-23.
- 6:** Patel RV. Splenogonadal fusion. *J Pediatr Surg* 1995; 30: 873-4.
- 7:** Putschar WGJ, Manion WC. Splenic-gonadal fusion. *Am J Pathol* 1956; 32: 15-35.