

Case report

Splenogonadal fusion

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Splenogonadal fusion is a rare congenital anomaly, which is described as abnormal fusion between the spleen and gonad or derivatives of the mesonephros. Surgical exploration is generally performed to rule out malignancy. The appearance at exploration can be misleading and often results in unnecessary orchiectomy.

CASE REPORT

A 17-year-old male patient have had a left scrotal mass for fourteen years and the mass had enlarged quickly in the last six months. A normal right testicle was palpated. On the left side, the testis was easily separable from a 3 cm × 2 cm scrotal mass located superior to the testis. The mass was soft and smooth. No other physical anomalies were found. Scrotal sonogram showed a homogeneous, well encapsulated left extratesticular mass, which was less echogenic and more vascular than the adjacent normal testis. There were no positive findings in the B ultrasound examination for liver, spleen or pancreas. Testicular tumour markers were normal. The blood, urine and stool routine tests were all within the normal ranges. The mass was thought to have originated from epididymis.

Surgical exploration was carried out via scrotal incision. Attached to the upper pole of the left testis, there was a purplish red mass, measuring about 3 cm × 2 cm × 2 cm and a cord like structure upwards through the inguinal canal. Inside the tunica albuginea at the upper pole of the left testis, another purplish red, well encapsulated, round nodule sized 1.5 cm × 1.5 cm was found. The extratesticular mass with the cord like structure was completely excised. The intratesticular was enucleated from the upper pole of the left testis leaving the left testis. Frozen and paraffin embedded sections showed the mass to be ectopic spleen tissue; final diagnosis was splenogonadal fusion (Figure).

DISCUSSION

Splenogonadal fusion was first reported in 1883 by Boestrom and described in 1889 by Pommer.¹ Although at least 150 cases have been reported in English medical literature, our case was the second report of splenogonadal fusion in China. More than 70% of reported cases were younger than 20 years and approximately 50% were younger than 10 years. Splenogonadal fusion can occur in male and female patients; the ratio of male to female

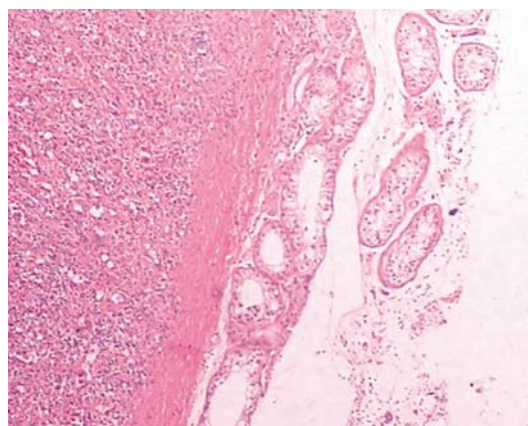


Figure. Histology of splenogonadal fusion showing splenic tissue with adjacent immature testicular tubules (HE staining, original magnification×40).

is approximately 15:1.² However, the ratio may be inaccurate because of the inaccessibility to the female gonad for examination.

Splenogonadal fusions were categorised into continuous and discontinuous types by Putschar et al.³ In the continuous form, the orthotopic spleen connects to the gonad with a cord of fibrous or splenic tissue. In the discontinuous form, there is no connection between the orthotopic spleen and gonad. The two forms appear to occur equally often. Almost all lesions occur on the left side. Splenogonadal fusion may be associated with other congenital anomalies such as cryptorchidism, limb defects, micrognathia and hypospadias. Of these, the association with cryptorchidism is the most common (31%), especially intraabdominal cryptorchism. Splenogonadal fusion is associated with other congenital anomalies in about one third of the cases and occurs most frequently with continuous form. This case is continuous type: there were no other congenital anomalies.

Most patients present with a scrotal mass, which generally attaches to the testis. Some of the growths were within the tunica albuginea of the testis. In this case, there

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were two masses, one attached to the left testis, another within the tunica albuginea. Some cases were discovered incidentally during orchiopexy or inguinal hernia repair. Approximately a fifth of the cases were discovered at autopsy. However, some unusual symptoms were presented such as painful scrotal swelling secondary to malaria and mumps or traumatic rupture of the ectopic spleen tissue. Bowel obstruction may be caused by the intraperitoneal cord. Up to date, only three cases of splenogonadal fusion were reported with a testicular neoplasm.⁴ It is difficult to make out a correct diagnosis preoperatively: most patients were thought to have a tumour, epididymitis, testicular duplication or torsion. Sonographic findings generally demonstrate a well encapsulated, homogeneous left extratesticular mass, which is less or equally echogenic to that of the adjacent normal testis.⁵ Radiocolloid spleen scintigraphy, preferably by single positron emission computerized tomography proved to be the best procedure to establish the correct diagnosis of splenogonadal fusion, especially in patients with congenital limb defects or orofacial anomalies.⁶

When the diagnosis is confirmed and no symptoms are present, it is controvertible whether further surgical intervention is needed. Surgical exploration is generally needed to rule out malignancy especially in patients with intratesticular mass. If the surgical intervention is performed, orchiectomy is generally not indicated. Splenic tissue can be dissected out of the gonadal structure and the testis can be saved. There is a growing importance of laparoscopy in the diagnosis and treatment of splenogonadal fusion especially in the cases of intraabdominal cryptorchism. In 37% of reported cases, orchiectomy was carried out unnecessarily because of suspected neoplasm. When doubt exists, a frozen section will clarify the diagnosis. Given the fact that the majority (82%) of patients were younger than 30 years at the time of diagnosis, orchiectomy should be avoided as often as possible. Familiarity with splenogonadal fusion allows one to recognize the lesion intraoperatively and preserve the testis.

The aetiology of splenogonadal fusion is obscure. The most widely accepted aetiology of splenogonadal fusion is assumed to occur between 5 and 8 weeks of gestation before the beginning of gonadal descent. When the splenic anlage is formed in the dorsal mesogastrum by several groups of cells derived from the coelomic epithelium and the mesenchyme of the mesogastrum, the

gonadal ridge is formed at almost the same time between the dorsal mesogastrum and the mesonephros on either side. During rotation of the dorsal mesogastrum, the splenic anlage moves to be adjacent to the left gonadal ridge. Fusion of these two primitive organs may occur during caudal migration.⁷ The exact cause of this malformation remains unknown but two hypotheses exist. Sneath suggested that slight inflammation of the peritoneal surfaces over the spleen and gonadal ridge could produce partial fusion of the two organs, while vonHochstetter postulated that a retroperitoneal pathway for splenic anlage cells might allow contact with the gonadal anlage.²

The reason for the combined congenital defects at a relatively higher frequency, in patients with splenogonadal fusion, is even more complex. As the limb buds, mandible and splenic bud in the dorsal mesogastrum develop at approximately the same time and injury at this critical stage of development would explain the associated anomalies.^{4,8}

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