Pathology International 1995; **45**: 871–874

Case Report

Splenic-gonadal fusion of the continuous type in an adult female

Antonio Alujevic,[1] Simun Andelinovic,[1] Gea Forempoher,[1] Snjezana Nigojevic,[1] Mihovil Biocic,[3] Zoran Rumboldt,[2] Leonardo Kapural[4] and Dragan Primorac[1,2]

Departments of [1]Pathology and [3]Surgery, Zagreb University School of Medicine, Split Branch, Clinical Hospital Split, Republic of Croatia and Departments of [2]Pediatrics and [4]Physiology, School of Medicine, University of Connecticut Health Center, Farmington, Connecticut, USA

The first case of a continuous type splenic-gonadal fusion in an adult female is described. This extremely rare anomaly occurs more often in the male where the abnormality may be associated with skeletal abnormalities or cryptorchidism. The few cases of splenic-gonadal fusion in the female previously described were found mainly in infants.

The present case of splenic-gonadal fusion occurred as a duct, approximately 10 cm long, extending from the inferior pole of the spleen to a junction in the left ovarian suspensory ligament. The superior two-thirds of this tubular structure consisted of splenic tissue, while the caudal one-third of the duct was composed of fat and fibrous tissue. In addition, two blood vessels, an artery and a vein, were present throughout the entire length of duct, and were located extracapsullary to the spleen in the cranial portion of this anomaly. The patient had no associated malformations in contrast to the majority of patients with continuous-type splenic-gonadal fusions.

Key word: splenic-gonadal fusion

Splenic-gonadal fusion (SGF) is an extremely rare congenital anomaly that occurs during embryogenesis; it consists of an abnormal connection between splenic tissue and mesonephric-gonadal structures.[1] This anomaly almost always affects the left gonad. Only two cases have been described in which the right gonad was affected.[2,3] Since 1889, a total of 128 cases have been reported,[4] of which eight were female.[5,6] Furthermore, only two cases of this malformation have been reported in adult females. Putschar and Manion[6] found that splenic-gonadal fusion occurs more often in males than in females (ratio of 9 : 1),[1] even though some reports describe a male : female ratio of 16.6 : 1.[5] Males most often present with either a swollen scrotal mass, inguinal hernia or cryptorchidism. Half of all reported cases were found during childhood. Splenic-gonadal fusions are characterized by two types, continuous or discontinuous.[1] In the continuous type, the spleen and gonad are connected by a cord consisting of either splenic or fibrous tissue. This type is usually linked with other abnormalities such as a limb defects,[7] micrognathia or cryptorchidism. In the discontinuous type, a remnant of splenic tissue attaches to the gonads with no connection to the spleen. Congenital defects are only rarely correlated with the discontinuous type of this disease.[5,8]

This report describes a case of continuous type splenic-gonadal fusion with an anomalous duct extending from the inferior pole of the spleen and terminating in the suspensory ligament of the left ovary. Histopathological findings revealed ductal structure composed primarily of splenic tissue.

CLINICAL SUMMARY

A 74 year old woman presented to the Department of Emergency Medicine at the Split Clinical Hospital Center with an acute abdomen and shock. Despite cardiopulmonary resuscitative efforts she expired before surgery could be performed. The cause of death was determined as hemorraghic shock induced by severe bleeding from a perforating ulcer on the duodenal bulb. The patient's medical history revealed that she had neither been diagnosed nor treated for this duodenal ulcer.

PATHOLOGICAL FINDINGS

An autopsy revealed pulmonary edema, with bilateral pleural effusions, vesicular emphysema, and a perforating ulcer on

Correspondence: Dragan Primorac, MD, Department of Pediatrics, School of Medicine, University of Connecticut Health Center, Farmington, Connecticut 06030, USA.

Received 30 January 1995. Accepted for publication 5 July 1995.

Figure 1 Photographic evidence of gross pathological findings, showing the cord like structure extending from the lower lobe of the spleen to the side of the left ovary.

the anterior surface of duodenal bulb 1.5 cm in radius and 3 mm deep. The damaged surface of two blood vessels was visualized through this defect.

Examination of the abdominal cavity revealed normally situated and sized organs with both ovaries fully descended. The spleen had a wrinkled purple surface with prominent ductal structures of the same color. The splenic-gonadal fusion extended from the inferior pole of the spleen as a yellow-colored duct that terminated in the suspensory ligament of the left ovary (Fig. 1). This duct was approximately 10 cm long with a diameter of 0.5 cm. Cross-sections of the duct showed a follicular structure with a dark purple pulp of moderate consistency. Histopathologic analysis showed that the superior two-thirds of the tubular structure consisted of splenic tissue (Figs 2,3). The inferior third of this structure consisted of fatty and fibrous tissue with blood vessels. The connection between splenic and fibrous tissues is not shown as longitudinal sections were not performed. However, two histologically distinct parts of the cord can be easily recognized in Figs 2, 3 and 4.

At the level of the splenic-cord junction, histologically normal splenic tissue was present, while peripheral to the splenic capsule were two blood vessels surrounded by fat and fibrous tissue (Fig. 2). Specifically, the lumina of the two blood vessels, an artery and accompanying vein, were found throughout the whole duct length, and peripheral to the splenic capsule on multiple cross-sections. Unfortunately,

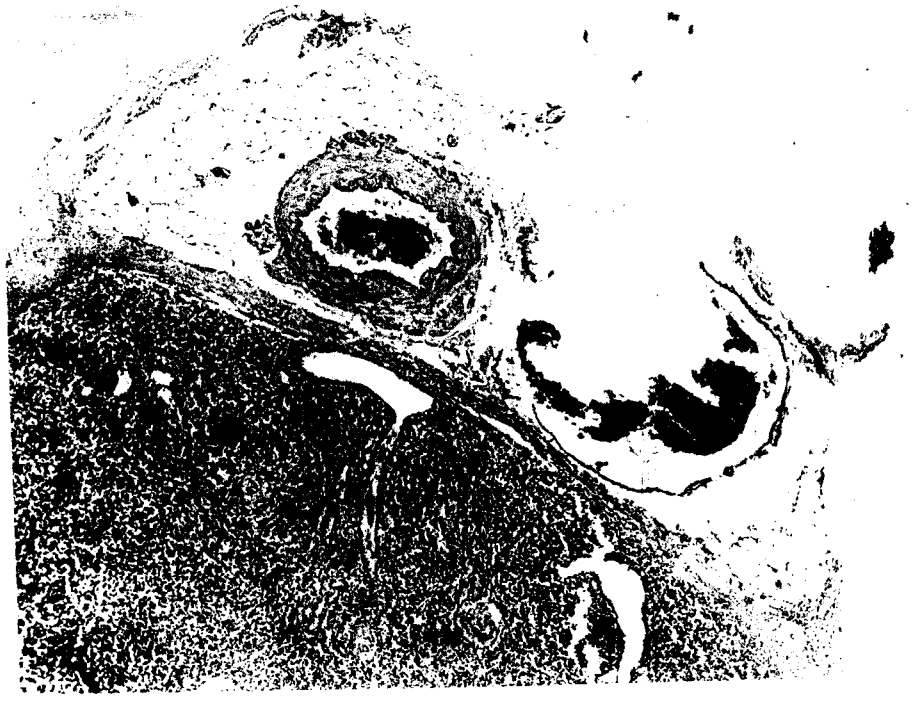

Figure 2 Cross-section of the cord showing the upper two-thirds of the structure which shows lymphoid tissue consistent with spleen (HE).

Figure 3 High power view of the upper two-thirds of cord (HE).

Figure 5 Histological findings of the left ovary. Normal ovarial tissue is present. As cord terminates in the suspensory ligament there is no direct involvement of the ovarian tissue in anomaly.

Figure 4 Lower one-third of cord consists of 2 vessels, embedded in the fatty tissue (HE).

radiographic contrast studies were not performed at that time to demonstrate possible anastomosis between branches of the splenic and ovarian arteries.

Figure 4 shows a representative caudal transverse cut at the level of the left ovarian suspensory ligament. The anomalous duct terminated within the suspensory ligament and did not have direct connection with ovarian tissue.

The histologic findings of the ovarian tissue are consistent with the age of the patient. These are thickened walls of blood vessels and white bodies, which suggest previous ovulatory activity (Fig. 5). Figure 5 also shows the ovarian artery and vein.

DISCUSSION

This report details a case of splenic-gonadal fusion, the third such observation in an adult female. To our knowledge it is

the first case describing a continuous type of splenic-gonadal fusion in an adult woman and the oldest patient described with this abnormality. Only eight cases of splenic-gonadal fusion have been described thus far with only two cases in adult women. One of these was a 44 year old nulliparous woman with a double uterus, menorrhagia and anemia.[9] However, questions still exist about the true nature of the anomaly in that case, as it was never classified as either the continuous or discontinuous type.[9] The second case was a 19 year old woman without symptoms,[6] in whom it was discovered as an incidental finding, and was the first splenic-gonadal fusion of the discontinuous type described in the female. Splenic-gonadal fusion may demonstrate diverse clinical pictures ranging from profuse vaginal bleeding and anemia to an absence of other anomalies or symptoms. Although the etiology of this disease has yet to be determined, some authors suggest that splenic-gonadal fusion may be linked with stomach rotation during the fifth gestational week, where the dorsal mesogastrium moves to the left side so that the splenic angle nears the left gonad.[6] The majority of cases discovered on the left side gives partial support to this theory. Putschar and Manion believe that since limb development occurs at approximately the same time as spleen differentiation, and because of the frequent combination of splenogonadal fusion with peromelus and micrognathia, these defects share a causative factors.[1] Pauli and Greenlaw have also suggested that the drug phenothiazine is a possible cause of splenogonadal fusion.[10]

The patient described here, however, had no congenital anomalies, which was atypical as it is known that a large number of continuous-type spleen–gonadal fusions have associated malformations. She had fully descended ovaries, had no history of gynaecological disease, was nulliparous and nulligravida, as suggested by circular involution of the cervix uteri. Although splenic-gonadal fusion can be a factor for infertility in the male, this anomaly has a low probability of affecting fertility in the female. Understandably, this anomaly had not been diagnosed in our patient as she was symptom-free. Possible complications may include torsion, stasis, cystic degeneration, infarcts, volvulus and strangulation of the bowel. It has been noted that during infections, particularly with malaria and mumps, ectopic tissue may enlarge and behave as a spleen.[11]

In the adult male this anomaly may manifest as either asymptomatic or symptomatic scrotum masses or, in some cases, classified as a third testicle.[12] Unfortunately, aberrant splenic tissue may be mistaken for a tumor and orchidectomy may be performed. A differential diagnosis of splenic-gonadal fusion by ^{99m}Tc sulfur colloid imaging,[13] arteriography[14] or by ultrasound[15,16] is possible. One must be aware of this unusual disease as careful examination and prescribed isotope scanning can eliminate an unnecessary orchidectomy in a situation where malignancy is suspected.

This report presents a case of continous splenic-gonadal fusion in an adult female where the anomaly was characterized by a cord-like structure extending from the spleen to the suspensory ligament of the ovary. Included in the cord were extracapsular blood vessels also extending from the spleen to suspensory ligament. Although we were unable to directly demonstrate the presence of an anastomosis between the splenic artery and ovarian artery, we speculate that such a connection exists. It is reasonable to postulate that during fetal ovarian descent, blood vessels, together with splenic tissue, could be pulled by the descending ovary.

REFERENCES

1 Putschar GJW, Manion DW. Splenic-gonadal fusion. *Am. J. Pathol.* 1956; **32**: 15–35.
2 Pendse KA, Mathur PN, Sharma MM, Gupta PO. Splenic-gonadal fusion. *Br. J. Surg.* 1975; **62**: 624–628.
3 Rosenthal JT, Bedetti CD, Labayen RF, Christy CW, Yakulis R. Right splenogonadal fusion with associated hypersplenism. *J. Urol.* 1981; **126**: 812–814.
4 Kameoka H, Yamada R, Sonoda T, Okuyama A. Splenic gonadal fusion with persistent mullerian duct syndrome. *Urol. Int.* 1993; **50**: 170–173.
5 Carragher, MA. One hundred years of splenogonadal fusion. *Urology* 1990; **35**: 471–475.
6 Meneses FM, Ostrowski LM. Female splenic-gonadal fusion of the discontinuous type. *Hum. Pathol.* 1989; **20**: 486–488.
7 Halvorsen FJ, Stray O. Splenogonadal fusion. *Acta Paediatr. Scand.* 1978; **67**: 379–381.
8 Gouw ASH, Elema DJ, Bink-Boelkens MThE, de Jongh JH, ten Kate PL. The spectrum of splenogonadal fusion. *Eur. J. Pediatr.* 1985; **144**: 316–323.
9 Almenoff AI. Splenic-gonadal fusion. *N.Y. State J. Med.* 1966; **66**: 1679–1681.
10 Pauli MR, Greenlaw A. Limb deficiency and splenogonadal fusion. *Am. J. Med. Genet.* 1982; **13**: 81–90.
11 Watson JR. Splenogonadal fusion. *Surgery* 1968; **63**: 853–858.
12 Daniel DS. An unusual case of ectopic splenic tissue resembling a third testicle. *Ann. Surg.* 1957; **145**: 960–962.
13 Guarin U, Dimitrieva Z, Ashley SJ. Splenogonadal fusion: A rare congenital anomaly demonstrated by ^{99m}Tc sulfur colloid imaging. *J. Nucl. Med.* 1975; **16**: 922–924.
14 Heloury Y, Valayer J, Leborgne J, Rogez JM, Robert R, Barbin JY. Splenogonadal fusion: Anatomic and angiographic study of a case. *Surg. Radiol. Anat.* 1986; **8**: 147–151.
15 Henderson GR, Henderson CD, Reid NI, Atkinson MP. Splenic-gonadal fusion: The ultrasound appearances. *Clin. Radiol.* 1991; **44**: 117–118.
16 Cochlin LLD. Splenic-gonadal fusion: The ultrasound appearances. *Clin. Radiol.* 1992; **45**: 290–291.