

CASE REPORT

Splenogonadal fusion associated with primary male infertility

M. AL-MARHOON, J. MATTHEW, V. NIRMALA* and E.O. KEHINDE†

Division of Urology, Department of Surgery, *Department of Pathology, Sultan Qaboos University, Muscat, Sultanate of Oman and †Division of Urology, Department of Surgery, Kuwait University, Kuwait

Case report

A 25-year-old man, married for 4 years, presented with infertility. A physical examination showed normal secondary sexual characteristics, an undescended right testis and a 4 × 4 cm left testicular mass, with a palpable single left vas deferens. He had no other obvious congenital anomalies. The patient was investigated to exclude left testicular tumour and to locate the right testis. Semen analysis revealed azoospermia, which together with elevated FSH and LH levels, suggested primary testicular failure. Ultrasonography of the left testis showed two fused homogenous solid masses, raising the clinical suspicion of a crossed ectopic testis (Fig. 1). A complete blood count, electrolytes, creatinine, liver function tests, urine culture and tumour markers (AFP and β hCG, 6 kIU/L and 2 IU/L, respectively) were all normal. CT of the abdomen showed a normal-size spleen in the left hypochondrium, with other normal abdominal viscera and a normal-size right testis lying in the inguinal canal. At exploratory surgery, there was an encapsulated mass arising from the upper pole of the left testis within the tunica albuginea. Frozen-section analysis of the mass was reported as splenic tissue, consistent with splenogonadal fusion. The splenic mass was excised, preserving the left testicular tissue from which a biopsy

was taken. Histological examination of the mass confirmed normal splenic tissue, composed of red and white pulp elements and fibrous capsule (Fig. 2). The left testicular biopsy showed no evidence of spermatogenesis. The right testis was located laparoscopically as a second-stage procedure and this confirmed an intra-abdominal testis of sufficient volume at the internal inguinal ring. A right orchidopexy was performed; a right testicular biopsy showed features consistent with postpubertal cryptorchidism and no evidence of malignancy.

Comments

This patient represents a rare case of left splenogonadal fusion with a right undescended testis and associated primary male infertility. To our knowledge, no other case has been reported of an association of splenogonadal fusion with primary male infertility. This congenital



Fig. 1. Ultrasonography of the testis showing two fused homogenous solid masses in the left scrotal sac.

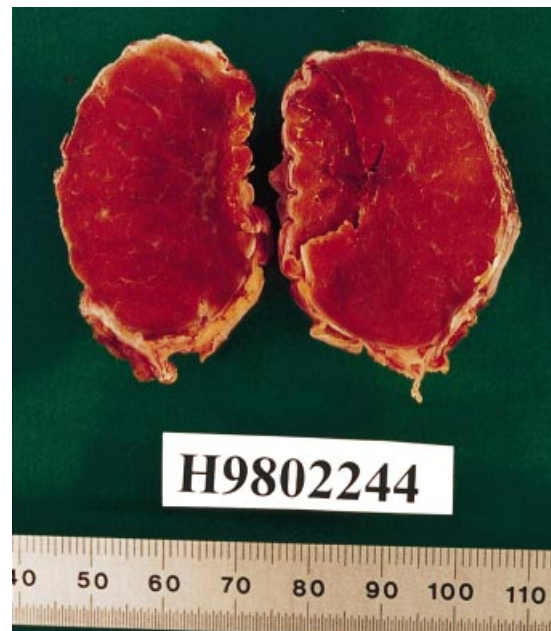


Fig. 2. Gross appearance of the excised and bisected left testicular mass. The mass has a well defined capsule. Note the deep red-brown normal splenic tissue.

anomaly contributed significantly to this patient's infertility, as the ectopic splenic tissue within the unyielding tunica albuginea must have compressed the developing and normally descended left testis, resulting in pressure atrophy of the left testis. This process resulted in primary testicular failure, as shown by high LH and FSH levels. The patient was unfortunate in that although he had a normal-size right testis, it was undescended and could therefore not compensate for the loss of function of the left testis. Splenogonadal fusion is a rare congenital abnormality of unknown aetiology, usually present on the left side (98%) and mostly in men (95%) [1]. About 125 cases of splenogonadal fusion have been reported [2]. Spleen scintigraphy is the best method of diagnosis [1,2]. There are two known types of splenogonadal fusion, i.e. continuous and discontinuous. In the former the spleen and splenogonad are connected by a fibrous cord, and it is usually associated with other congenital anomalies. In the latter there is no connection between the spleen and gonad, and other congenital anomalies are often absent [3]. The present case was of the discontinuous type. Removal of the left testicular mass helped to confirm the diagnosis and to exclude the remote possibility of a neoplasm, as testicular cancers are common in patients with undescended testis. Removal of the ectopic splenic tissue is justified considering its known tendency to enlarge under conditions causing splenomegaly. Whether removal of the ectopic splenic tissue will relieve the pressure on the left testis enough to improve the patient's fertility is doubtful. Right orchidopexy rather than orchidectomy was carried out in view of

the abnormal histology of the contralateral testis. The need to regularly self-examine the high-risk right testis (for the early detection of a malignant mass) was emphasized to this patient. Urologists and pathologists should be aware of this entity in patients with a left testicular mass, and unusual features on ultrasonography and histology of the testis.

References

- 1 Bostwick DG. Spermatic cord and testicular adnexa. In Bostwick DG, Eble JN, eds, *Urologic Surgical Pathology* 1st edn. Chapter 12. St. Louis: Mosby, 1997: 647–73
- 2 Steinmetz AP, Rappaport A, Nikolov G, Priel IE, Chamovitz DL, Dolev E. Splenogonadal fusion diagnosed by spleen scintigraphy. *J Nucl Med* 1997; **38**: 1153–5
- 3 Moore PJ, Hawkins EP, Galliant CA, Guerry-Force ML. Splenogonadal fusion with limb deficiency and micrognathia. *South Med J* 1997; **90**: 1152–5

Authors

M. Al-Marhoon, MD, Senior House Officer in Urology.
 J. Matthew, MBBS, MS, MCh(Urol), Registrar in Urology.
 V. Nirmala, MBBS, PhD, Consultant Pathologist.
 E.O. Kehinde, MBBS, FRCS(Eng), MD, Assistant Professor of Urological Surgery/Consultant Urological Surgeon.
 Correspondence: Dr M. Al-Marhoon, Department of Surgery, College of Medicine, Sultan Qaboos University, PO Box 35, Al-Khouth, Muscat 123, Sultanate of Oman.
 e-mail: mmarhoon@squ.edu.om