Discontinuous splenogonadal fusion mimicking testicular tumor

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ABSTRACT
The fusion between splenic tissue and the gonads or derivatives of the mesonephros is a rare congenital anomaly. This was first described by Pommer in 1887. The diagnosis is usually made at operation or autopsy. Here we present a case of left splenotesticular fusion where the diagnosis was made intraoperatively and the testis was saved. The case is discussed with a review of the literature.

Keywords: Splenogonadal fusion, tumor, testicular.

Testicular fusion with splenic tests is a very rare anomaly. Approximately 124 cases have been reported in the latest review in 1993.1 Putschar and Marion in 1956 first defined its pathology. The diagnosis in most cases was made during surgery.

Case Report. An otherwise healthy four-year-old Saudi Arab boy was referred for evaluation of a right-sided cryptorchism. Physical examination revealed a right inguinal undescended testis. The left testis, which was situated in the scrotum, contained a discrete, firm mass, about 2 cm in diameter, related to the upper pole. All laboratory tests, including B-HCG and alpha-feto proteins, were normal. Ultrasonography confirmed the solid nature of the mass as well as its attachment to the upper pole of the left testis (Fig. 1).

The child underwent bilateral groin exploration. Right orchidopexy was performed. The left spermatic cord was isolated and cross clamped with a non-crushing clamp. The left testis was then mobilized upwards into the wound. The mass within the left testis was found to be smooth walled, dark brown and well encapsulated within the tunica vaginalis but adherent only to the tunica albuginea with fibrous strands (Fig. 2). The mass, 3 x 1.5 cm, was dissected free from the testis and was sent for frozen section examination. The preliminary examination reported the mass as a benign lymphohistiocytic nodule with no malignancy. The mass was totally excised and the testis preserved. The final histopathologic examinations showed splenic tissue composed of white and red pulps (Fig. 3). The red pulp was made up of cords and sinuses. The white pulp was composed of lymphocytes with follicle formation and active germinal centers. Post-operative ultrasonography of the left upper quadrant revealed a normal spleen.

Discussion. Splenogonadal fusion is a rare malformation. The possible origin of such a union has been outlined in detail by Putschar and Marion.2 The most likely theory is based on the embryological development of the spleen and the gonadal structures.3 Some undefined insult seems to allow fusion between some primitive splenic cellular clumps and the nearby most cephalad extension of the mesonephric (Wolfian) body, which occurs between the fourth and tenth weeks, when the genital glands start to migrate caudally.4,5

There are two pathological types of this anomaly, the continuous and the discontinuous, depending on the presence or absence of a structural connection that may contain splenic tissue between the original spleen and the ectopic splenic tissue.2

In their detailed report (1988), Walter and colleagues pointed out that splenogonadal fusion occurs 13 times more frequently in males and almost always with the left testis. About one-third of cases are associated with other congenital defects, which are five times more common in the continuous type. Peromelia (deformed limbs), micrognathia (small

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jaws), inguinal hernias and cryptorchism are among the commonly associated anomalies.

The anomaly is usually recognized as an incidental finding at autopsy or during exploration of a testicular mass. Review of the literature shows few cases with unusual presentations as epididymitis, torsion or enlargement of splenic rest during an active attack of malaria, bowel obstruction secondary to a cord of splenic tissue compressing the colon or a case of intra-abdominal testis with seminoma and splenic fusion.

It has been reported that orchietomy was performed in about 50% of patients with splenogonadal fusion undergoing surgery due to clinical unawareness of this rare benign condition. Part of the difficulty is when the splenic tissue is fused with the testicular tissue within the tunica albuginea. It was emphasized that in almost all cases of splenogonadal fusion, testicular development is normal and, therefore, orchietomy should be avoided.

Some authors claim, therefore, that surgery may be avoided for splenogonadal fusion if the diagnosis could be made by Tc99m sulfur colloid splenic scanning. This is often not the case, however, as surgery is still necessary in most instances for correction of associated conditions. Many surgeons believe that intra-operative frozen section examination of suspicious benign testicular neoplasms may be the best way to diagnose the splenogonadal fusion and hence preserve the testicles. Caution should be taken by the pathologist, however, since teratoma or lymphoma have a similar pathological appearance.

Our case adds to the number reported in the literature of this anomaly and demonstrates the value of intra-operative frozen section in the diagnosis and eventually leads to the preservation of the testis.

References