Dear Editor:

Splenogonadal fusion, sometimes called supernumerary intrascrotal ectopic spleen, is a relatively uncommon condition, to judge by the few cases reported, where splenic remnants are bound to the surface of the testis or epididymis and even in an intragonadal location.

The exact incidence of splenogonadal fusion is unknown, and may be greater than reported. According to Montes and Prada, approximately 160 cases have been reported to date. Clinically, splenogonadal fusion appears as a testicular tumor or paratesticular mass, but may also be diagnosed during surgery for cryptorchism (as occurred in our case) or inguinal hernia. The interest in this condition stems from the fact that it occurs as a palpable tumor and therefore requires use of a whole series of laboratory and imaging tests, a process usually ending in the operating room. While awareness of its existence does not prevent surgery in most cases, it may avoid an excessively aggressive approach in some patients.

Our patient was a black male who attended our practice reporting absence of both testes from the scrotum. Physical examination revealed poorly developed secondary sexual characteristics and a small scrotum. Testes could not be palpated either in scrotum or in inguinal canals. Laboratory test values include: FSH, 50.53 mU/mL; LH, 27.39 mU/mL; testosterone, 3.54 ng/mL; and free testosterone, 5.96 pg/mL (hypergonadotrophic hypogonadism). Testes were not seen in scrotum in MRI, which showed images consistent with testes of an ovoid morphology. A subsequently requested spermiogram was reported as “no spermatozoa in fresh sample or in sediment after centrifugation of the whole sample; cryopreservation was therefore not performed”.

Based on these findings, patient was proposed testicular conservation to preserve hormone production, but he finally elected laparoscopic bilateral orchiectomy (fig. 1) + placement of bilateral testicular prosthesis + subsequent hormone replacement therapy. According to the pathology report (fig. 2), the left testis consisted of tubuli formed by Sertoli cells only, with no evidence of intratubular neoplasm and, continuous with parenchyma, an accessory spleen with no changes (“splenogonadal fusion”).

Splenogonadal fusion is a congenital anomaly characterized by an abnormal fusion of spleen and gonad and mesonephric derivatives. Although splenogonadal fusion may occur in both sexes, it is much more common in males (in a 9:1 or 15:1 ratio according to various authors). It should be noted, however, that in 4 cases reported in females, splenic tissue was adjacent to ovary and mesovarium. Cases have been reported in intersexuals. This condition was first described in 1889 by Pommer, but it appears that it had already been

Figure 1 – Gross image of the left testis (orchiectomy specimen 18.4 g in weight in which a 3.5 cm testis showing no relevant changes is identified). An accessory spleen continuous with the parenchyma and with no relevant changes is identified.
In the ovary may be underdiagnosed because they cannot be explored as easily as those in the testis, and will probably never be diagnosed if they cause no symptoms. Only the few cases with intragonadal spleen would hardly be explained by the previous theory. Such cases may be hypothesized to occur as a consequence of induction of hemopoietic potencies in gonadal mesenchyma.

There is currently another theory stating that an abnormal development of the cranial suspensory ligament of testis causes an abnormal involution resulting in cryptorchism and colonization of the abnormal ligament by splenic cells. Associated congenital anomalies such as cryptorchism, peromelia, micrognathia, hypoglossia, palatine defects, polymicrogyria, craniosynostosis, spina bifida, cardiac defects, diaphragmatic hernia, hypoplastic lung, and anorectal abnormalities have been reported in 30% of continuous forms. When Putschar y Manion described the two forms, they associated continuous splenogonadal fusion to a high proportion of cryptorchism and inguinoscrotal hernia due to the inguinal closure defect produced by the fibrous cord.

Clinically, splenogonadal fusion occurs in 40% of cases as a paratesticular mass, preferentially located close to the upper testicular pole. Occasionally, when the mass is intimately attached, differential diagnosis with paratesticular solid masses (rhabdomyosarcoma, lymphoma, metastasis, etc.) becomes very difficult. Although the presence of splenic remnants at scrotal level does not usually cause any problem, it should be reminded that the spleen, even when ectopic, may experience the same disorders as its normally located counterpart. This explains why in patients with hypersplenism and diseases where the spleen is involved, such as malaria, mumps, mononucleosis, or leukemia, splenic remnants adjacent to the testis may be affected similarly to the spleen, causing an enlargement of the mass associated to pain. Associated rupture has also been reported in some cases.

Continuous fusion sometimes causes abdominal symptoms, and intestinal obstruction may eventually occur when the attachment cord is in an intraperitoneal location or as part of the clinical picture of cryptorchism.

Although exceptional, germ tumors associated to splenogonadal fusion have been reported in three patients, all of them with a history of associated cryptorchism. Diagnosis is usually made during surgery for a paratesticular mass, inguinoscrotal hernia, or cryptorchism, but palpable nodules in scrotum with no other symptoms are found in most patients. If a reasonable suspicion of splenogonadal fusion exists (coexistence of bone changes with an asymptomatic left scrotal mass), some authors advocate use of radionuclides with Tc99m to confirm or rule out the condition. Antenatal ultrasound diagnosis of two cases has been reported (gastrointestinal malrotation plus splenogonadal fusion syndrome).

No ultrasonographic differences exist between the accessory spleen and normal testis. A clearly outlined 2-3 cm mass in close paratesticular relationship is seen. Interestingly, Patel advises that when a mass with these characteristics is manipulated, it should be watched whether testicular movements translate into spleen movements, which would only occur when continuous fusion exists.
On gross inspection during surgical examination, splenogonadal fusion appears as a 2-3 cm solid red mass consisting of a parenchyma similar to that of spleen. Histologically, the mass consists of completely normal splenic tissue, including red pulp and sinusoids.

If diagnosis is made at examination, it is advised to take a tissue sample for confirmation by the pathologist, so that partial mass excision sparing the testis, particularly in young males, may be performed.

It should be noted that some authors, such as Patel, advise that ectopic splenic tissue is searched for when splenectomy is required, assuming a high incidence of asymptomatic splenogonadal fusion, because as this is a normal tissue, though ectopic, remnants would be functionally normal and symptoms derived from hypersplenism could recur.

The clinical interest of splenogonadal fusion lies, apart from its low frequency, in the fact that it is most often diagnosed as an intrascrotal tumor commonly requiring surgery and orchiectomy, procedures which could be avoided if there is a prior suspicion, because the presence of normal splenic tissue in the scrotum does not represent in itself any problem for the patient. It should also be reminded that accessory and ectopic splenic tissue may be involved in conditions causing splenism such as mumps, leukemia, mononucleosis, and even malaria. Splenic remnants in the scrotal area would therefore increase in size.

REFERENCES


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