DISCONTINUOUS INTRABDOMINAL SPLENOGONADAL FUSION WITH GERM CELL TUMOR. EXCISION WITH ROBOTIC ASSISTANCE. ADULT CASE REPORT

Laura Velarde Ramos, Francisco Sepulveda, Eloy Silva and Octavio A. Castillo.

Urology Department. Clínica Indisa. Faculty of Medicine. Universidad Andres Bello. Santiago. Chile.

Summary.- OBJECTIVE: Present the case of a patient with a discontinuous intrabdominal splenogonadal fusion with an associated germ cell tumor.

MÉTODOS: A case of a man of 29 years with bilateral cryptorchidism and left intra-abdominal discontinuous splenogonadal fusion associated with seminoma as an accidental finding in the context of a robotic pyeloplasty due to ipsilateral ureteropelvic junction stenosis.

RESULTS: The total operative time was 80 minutes (atrophic gonad removal and pyeloplasty by the Anderson-Hynes technique) with an estimated blood loss less than 100 cc. The hospitalization time was less than 36 hrs. The pathology and immunohistochemical report was compatible with intratubular germ cell neoplasia (seminoma).

CONCLUSION: The splenogonadal fusion is an uncommon pathology. While a high clinical suspicion may avoid unnecessary orchietomy in young patients, its association with disorders such as cryptorchidism should make us suspect the possible presence of a concomitant germ cell neoplasia.

Keywords: Splenogonadal fusion. Testicular tumor. Cryptorchidism.

Resumen.- OBJETIVO: Presentar el caso de una fusión esplenogonadal intrabdominal discontínua asociada a neoplasia germinal.

MÉTODOS: Se presenta el caso de un varón de 29 años con criptorquidia bilateral y fusión esplenogonadal discontinua izquierda intrabdominal, asociada a Seminoma como hallazgo casual en el contexto de una pieloplastía robótica debido a una estenosis pieloureteral ipsilateral.

RESULTADOS: El tiempo operatorio total fue de 80 minutos (extirpación de gónada atrófica y pieloplastía según técnica de Anderson-Hynes) con un sangrado estimado menor a 100 cc siendo el paciente dado de alta antes de 36 hrs. El informe anatomopatológico e inmunohistoquímico fue compatible con neoplasia intratubular de células germinales [Seminoma].

CONCLUSIÓN: La fusión esplenogonadal es una patología infrecuente. Si bien una alta sospecha clínica puede evitar una orquiectomía innecesaria en pacientes jóvenes, su asociación a alteraciones como la criptorquidia debe hacernos sospechar la eventual presencia de una neoplasia de células germinales concomitante.

Palabras clave: Fusión esplenogonadal. Tumor testicular. Criptorquidia.

INTRODUCTION

The splenogonadal fusion is an uncommon condition characterized by abnormal attachments of the splenic tissue to the ipsilateral or contralateral gonad. The first description of this condition is attributed to Bostroem in 1883 and since then more than 160 cases have been reported, mostly in men younger than 30 years (1,2). The diagnosis is usually an incidental finding but it may present as cryptorchidism or as a painless scrotal mass...
(predominantly left). For this reason is more readily detectable in the male but it should not be neglected the cases described in women and intersexual states (3). When presented as a scrotal mass, this is often misdiagnosed as a malignant testicular tumor, deriving in an unnecessary orchiectomy.

In the case of patients with undescended testes is important to note that the incidence of testicular malignancy is 3-48 higher than in the normal population, so in the case of its association with splenogonadal fusion must be taken into consideration.

**CASE REPORT**

Twenty nine years old male patient, diagnosed in childhood of bilateral cryptorchidism. Bilateral inguinal exploration was performed at 4 and 12 years without demonstrable testicles. Because of the development of secondary sexual characteristics and testosterone levels within the normal levels, follow up was decided.

He came to our clinic with left flank pain. After specific studies (ultrasound, CT and renogram MAG-3) the diagnosis was stenosis of the left ureteropelvic junction. As a parallel finding, inguinoscrotal ultrasound describes the right testicle in the inguinal canal and absence of the left one. In the CT scan a structure is displayed at the lower pole of left kidney interpreted as embryonic remnants of the Wolffian duct (Figure 1). He was scheduled for surgical repair with robotic assistance (da Vinci, Intuitive, CA) of his main condition placing the patient in flank position with a left paraumbilical 12-mm trocar for the optics, three 8 mm robotic trocars in the left lower quadrant, subcostal and epigastrum for the first, second and third robotic arms and a further 10 mm trocar in the lower abdomen above the midline for the assistant (Figure 2). The robot assembly is carried from the back of the patient.

In the initial exploration an intraabdominal structure is visualized in the left Toldt line compatible with atrophic testes merged in the upper pole with a separate tab similar to the splenic tissue (Figure 3). First we performed the removal of that structure with robotic assistance, continuing with dismembered pyeloplasty by the Anderson- Hynes technique with a 5/0 Monocryl suture without incidents. The total operative time was 80 minutes and the patient was discharged after 36 hours of surgery.

The pathology report of the specimen was reported as: Splenogonadal Fusion.

Splenic tissue was normal. The testis evaluation revealed: a small focus of atypical cell interstitial infiltration of neoplastic appearance not associated with mass or nodule, compatible with pure seminoma of classic type in its original form; unclassified extensive testicular intratubular germ cell neoplasia; a 0.6 cm...
intratesticular benign nodule of Sertoli cells; testicular tissue with signs of atrophy and interstitial fibrosis; scattered testicular intratubular microcalcifications; reactive immunohistochemical pattern compatible with intratubular germ cell neoplasia and seminoma with negative inhibin and positive PLAP and CD 117.

DISCUSSION

Splenogonadal fusion corresponds to the abnormal connection between the gonad and spleen tissue and its mesonephric derivatives. Most reported cases were males under 30 years old with a history of left intratesticular mass, painless and long-standing in which orchiectomy were performed for suspected testicular tumor, but have also been described in female and intersexual states. Cases of ovarian splenogonadal fusion due to the difficulty in the physical examination and lack of symptoms are probably underdiagnosed. There is only a few cases of intrabdominal location and are usually diagnosed by chance in the context of the study of other pathologies.

Its classification is established according to the anatomic relationship between the gonad and the spleen (1). The continuous form is slightly more frequent, showing the continuity between the spleen and splenic tissue attached to the gonad by fibrous cord that is usually retroperitoneal. During the gonadal descent this fibrous cord may fragment causing the discontinuous form. In the Discontinuous form the ectopic splenic tissue can be found all along the way but it is usual to descend through the inguinal canal beside the gonad, separated by a capsule but covered by tunica vaginalis. Less common is the presentation of intraabdominal discontinuous form, as is the case described. The literature reports that 31% are patients with cryptorchidism and of these 91% are intraabdominal location.

During embryonic development, between 5th and 8 weeks failure occurs in the separation of cells from the urogenital ridge (gonadal mesoderm and metanephros (precursor of the epididymis and vas deferens)) and precursor cells of the spleen and extremities. This explains that during the descent gonadal spleenic tissue can be found all along the way (spermatic cord, epididymis, testicular albuginea and even intraparenchymal) and can be associated with malformations of the limbs being more often found in continuous forms (2,4,5).

Splenogonadal Fusion is a benign congenital malformation, often asymptomatic unless the patient develops a disease with splenic involvement (leukemia, mononucleosis, malaria), because the splenic ectopic tissue is also affected, leading to symptoms of volume augmentation and pain. Its association with germ cell tumors has been described in a few cases with the particularity that these patients had ipsilateral cryptorchidism (2).

The diagnosis, unfortunately, is usually performed after histological confirmation postorchiectomy because these patients are often mistakenly labeled as malignant testicular tumor.

The diagnostic algorithm is not clearly defined, for this reason it is important to have this diagnosis in mind as a differential diagnosis of the testicular tumors, cryptorchidism and in the case of its association with bone alterations so we can avoid an unnecessary orchiectomy.

In 1980, Cortes et al7 performed a review of 111 cases of splenogonadal fusion of which 31% had cryptorchidism (59% bilateral, 26% right intraabdominal and 65% intraabdominal left). In the continuous forms 44% had cryptorchidism. Associated malformations were found only in cases of cryptorchidism (agenesis of the lower limbs, spina bifida, imperforate anus, diaphragmatic hernia, hypospadias).

We found 3 indexed cases of splenogonadal fusion associated with testicular neoplasia (1980 Falkowski WS8, scrotal, anaplastic seminoma; 1996 Thomsen BM9, scrotal, mixed tumor; 1999 Imperial SL10, intraabdominal seminoma), being our case the second of intraabdominal location associated with seminoma. In our patient, despite high suspicion of splenogonadal fusion during surgical exploration, complete excision was decided because it is an adult patient with intraabdominal cryptorchidism with little chance of a descent to scrotum, a greater likelihood of testicular neoplasia and the atrophic appearance of the gonad.

CONCLUSION

The rarity of this condition and the consequences of its ignorance make it necessary to describe the cases diagnosed in our midst. A detailed description of the history, physical examination, laboratory tests and diagnostic technique will allow us to take it into
consideration in the differential diagnosis of testicular tumors, cryptorchidism, inguinal hernias and abdominal masses. When in doubt, complete excision of the mass with intra-or postoperative histological confirmation should be performed, thus preventing poorly planned surgery for misdiagnosis. In the case of undescended testis in adults we should take into account the increased incidence of associated neoplasia so we should consider perform a complete excision of the mass.

REFERENCES AND RECOMMENDED READINGS
(*of special interest, **of outstanding interest)