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Case Report

An Uncommon Cause of Testicular Lump: Discontinuous Type Spleno-Gonadal Fusion

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Abstract

Splenogonadal fusion is a rare congenital abnormality. Preoperative diagnosis is difficult but can be based on scintigraphy using technetium 99m. It's a benign lesion that remains to be differentiated from a testicular tumor.

In most cases, the splenic tissue can be dissected of the gonadal structures easily, and if there are any doubts concerning the nature of the swelling, an intraoperative frozen section can be performed to avoid an unnecessary orchiectomy.

We report the case of a 4-year-old child in whom a scrotal mass indicated many investigations before a surgical exploration, and in whom the surgical specimen was diagnosed histologically as a splenogonadal fusion.

Objective of our work

Do not ignore and evoke splenogonadal fusion despite the exceptional location of splenic ectopia before a vascularized para-testicular mass and make an extemporaneous anatomopathological study of the lesion avoiding an unnecessary orchiectomy.

Introduction

A recent increase in testicular volume in children should suggest the possibility of a testicular tumor which is benign in 48% according to a French multicenter study [1]. The majority of them prove to be a hydrocele, a varicocele, a spermatocele, an inguinal hernia, an epididymitis.

More rarely it may be a macro-orchidism as seen in fragile X syndrome; voluminal compensation in the absence of a controlateral testicle testis; or spleno-gonadal fusion.

Described for the first time in 1883 by BOSTROEM [2], spleno-gonadal fusion is a rare etiology of testicular tumor, but must be evoked to avoid an abusive orchiectomy with harmful endocrine impact, exocrine and psychological consequences. In our present work, we report the case of a splenon-gonadal fusion in a 4-year-old child and discuss its embryology, associated abnormalities and the importance of diagnosing preoperatively (à reformuler la phrase) to avoid orchiectomy.

Case report

H.M., a 4-year-old child with unremarkable medical history, presented with a left scrotal mass of 3 x 2 cm, hard and not painful, accidentally discovered by the mother. This motivated the family to consult elsewhere, where an ultrasound was performed and showed a large left testis with heterogeneous echostructure and disseminated hypo-echogenic nodules. The epididymal tail was enlarged with hydrocele. The diagnosis of tuberculosis was suspected.

A Rhabdomyosarcoma not being ruled out. The patient was referred to our hospital afterwards where a search for tuberculosis was carried out with a negative results.

Ultrasonography identified an intrascrotal homogenous multi-nodular mass crushing the testicle downwards, measuring 12 x 6 mm, with heterogeneous echongenecity (Figure 1). On Color Doppler, the mass showed a hypervascularized heterogeneous tissular echostructure, this process being multi-nodular with a hydrocele reaction (Figure 2).

A computed tomodensitometry was performed and revealed a left, well circumscribed multi-nodular intra-scrotal mass measuring 20 x 21 mm x 28 mm(height), with tissular density that enhanced intensely and homogeneously after injection and which crushes the testicle downwards (Figure 3).

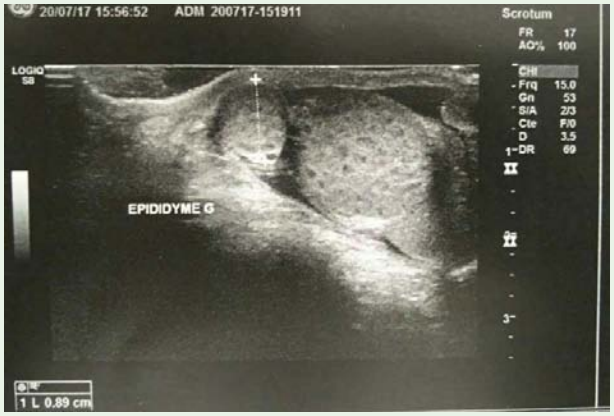

Figure 1: An Ultrasonography showed an intrascrotal homogenous multi-nodular mass crushing the testicle downwards, measuring 12 x 6 mm, with heterogeneous echongenecity.

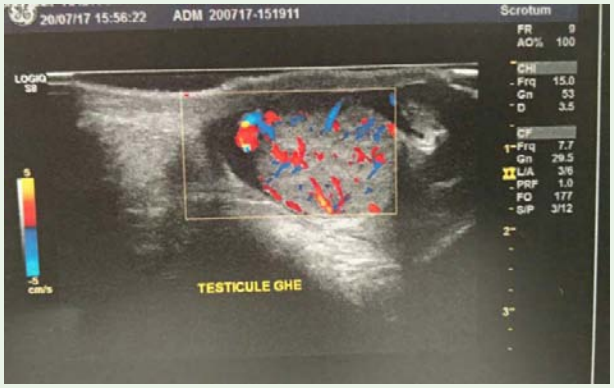

Figure 2: A Color Doppler showed a hypervascularized heterogeneous tissular echostructure mass.

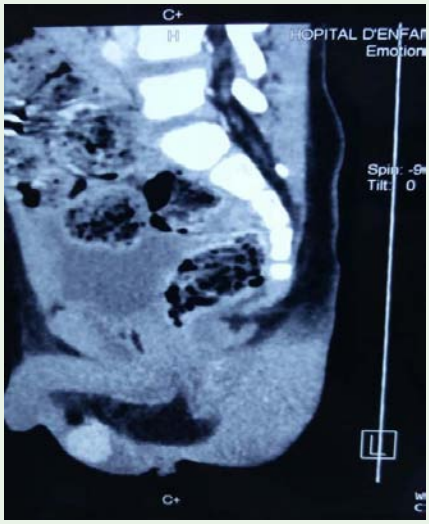

Figure 3: A computed tomodensitometry showing a left, limited, multi-nodular, intra-scrotal lesional process of tissular density, enhanced intensely and homogeneously after injection, measuring 20 x 21 mm that evokes a Rhabdomyosarcoma.

Figure 4: Gross description of the mass showing 5 well-limited brownish nodules with a whitish pecked appearance and which are appended to the head of the epididyme.

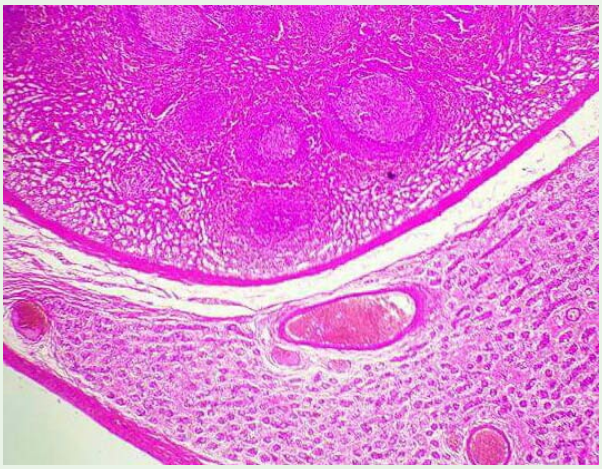

Figure 5: Microscopic picture confirming splenic tissue with scattered lymphoid follicles with germinal centers and red pulp in mass, separated from normal testis by a fibrous capsule.

Chest x-ray, abdominal Computed Tomography (CT) did not show any abnormality. Beta- Human Chorionic Gonadotropin (bHCG) and alpha foeto-protein were negative. A testicular neoplasm was suspected and the patient underwent a left radical inguinal orchiectomy. Gross examination showed several brownish nodules well demarcated from the testis (Figure 4).

The pathology report concluded to a discontinuous spleno-gonadal fusion. The microscopic examination showed normal splenic tissue circumscribed by a fibrous capsule with white pulp and red pulp .There was no evidence of spermatogenesis in the testicular tissue (Figure 5).

Discussion

Spleno gonadal fusion is defined by the intra scrotal presence of ectopic splenic tissue generated by an abnormal connection between the spleen and the gonad or the mesonephrotic derivatives during the embryonic period [2,3]. It can be discovered in the neonatal period. More than 150 cases have been published mostly in boys, although some cases of spleen fusion to the left ovary are published [4,5]. The sex ratio is 15/1 [6,7], and these are sporadic cases [8].

Two main types have been described [2]: the continuous form, which represents 56% of the cases, where there is a fibrous tract (cord-like) or a splenic parenchyma stretched between the principal spleen and the gonadal fused parenchyma and the discontinuous form, where this cord-like does not exist. This latter form represents 44% of spleno-gonadal fusions, the spleen being separated from the gonadal tissue by a capsule. In one third of the cases, there are associated congenital malformations and particularly in the continuous forms of spleno-gonadal fusion (44 to 50% of the cases). The most frequent is cryptorchidism [9] found in 31% of cases. Abnormalities of the limbs are found in 19% of cases [3] distributed among ectromelia, phocomelia and amelia. These splenoid-gonadal fusion-related limb deformities define the Spleno Gonadal Fusion Limb Defect Syndrome (SGFLD) of which 30 cases have been published; it is marked by a perinatal mortality rate ranging from 40 to 50% [2].

In 8% of cases we find a micrognaty [6]. Other associated abnormalities are: microgyria, craniofacial asymmetry, cleft palate [7], cardio pulmonary abnormalities [3], persistence of Ductus Venosus, microgastria, ano rectal malformations, inguinal hernia , hypospadia [9], sexual ambiguities, varicocele, spina bifida [6], and osteogenesis imperfecta.

The anomaly is formed during the first months of embryonic life [9] at the moment when the two structures are separated only by two layers of coelomic epithelium after gastric rotation and dorsal mesogastric growth; which explains the left localization of spleno-gonadal fusion, although a case of splenic tissue fusion with the right testis has been described [10]. For some authors [6], a peritoneal inflammation between spleen and gonad would allow their fusion; for others [2] it would be a migration of spleen cells by retroperitoneal route. The recent theories [3] resume the idea of fusion by cell migration under the effect of a teratogenic agent unknown to this day. The possibility of hereditary participation in an autosomal recessive form is also mentioned to explain the association of certain continuous forms with abnormalities of the limbs and the face [3].

Preoperative diagnosis is difficult and the left localization of the mass should attract attention. The ultrasound cannot contribute significantly in the diagnosis and shows an isoechogenic mass [3]. The scanner would show the tract of the continuous forms [5] and the arteriography, which can locate the ectopic spleen, remains an examination too aggressive for the child. The best examination is Technetium 99m scintigraphy [2,6,8]. This examination could avoid an unnecessary orchidectomy in 37% of cases.

When the abnormality is recognized by surgery, selective excision of the splenic tissue is possible and is the rule for the discontinuous forms [10,11], except in cases of intra-testicular splenic extension. It is necessary, for continuous forms, to cut the tract after haemostasis of its internal pedicle and to peritonize it to avoid the formation of intraperitoneal flanges [8]. The laparoscopy is very interesting especially for cases associated with cryptorchidism [9].

For some authors [4], abstention can be discussed if the abnormality is recognized in preoperative and if it does not manifest clinically.

Conclusion

Surgeons and clinicians should be aware of this important but rare diagnosis in front of a testicular mass of the child, especially if this mass is on the left and is isoechogenic on ultrasound. Technetium 99 m scintigraphy is then of great diagnostic value. The macroscopic aspect remains characteristic and must be recognized in order to avoid unnecessary orchidectomy.

Ethical Approval

Written informed consent to publication was obtained from the next of kin.

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