REVIEW ARTICLE

SPLENOGONADAL FUSION – A REVIEW OF RARE ANOMALY

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ABSTRACT

Splenogonadal fusion is a rare congenital malformation that involves an abnormal connection between the spleen and the gonad or mesonephric derivatives. It manifests itself as a mass consisting of splenic and testicular or ovarian tissue. Splenogonadal fusion has been classified into two types; continuous, where there is a direct connection between spleen and gonad; and discontinuous, where ectopic splenic tissue is attached to the gonad, but there is no connection to the spleen. Many cases had an associated other anomalies either genital or systemic. Knowledge about the existence of such an uncommon entity is essential even to be suspected preoperatively as the appearance at exploration can be misleading as a malignant mass and often results in unnecessary gonadal removal.

KEY-WORDS: Splenogonadal Fusion; Congenital; Anomaly

Introduction

Splenogonadal fusion is a rare benign congenital malformation that is characterized by an abnormal connection between the spleen and the gonad or mesonephric derivatives.[1-4] It is also called as supernumerary intrascrotal ectopic spleen.[5]

The anomaly was first mentioned by pathologist Eugen Bostroem in 1883, but it was not until 1888 when pommer described the malformation in details.[1] In 1988, Petrik recruited and studied the largest series, consisting of 87 cases. Approximately 160 cases of splenogonadal fusion have been reported to date.[5] The most recent comprehensive review of splenogonadal fusion was in 1990 by Carrgher et al, describing 123 cases.[1-4]

Incidence

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). Cases have been reported in intersexual also. It should be noted, that in 4 cases reported in females, splenic tissue was adjacent to ovary and mesovarium.[5] However, the embryological origin as described below does not explain the different incidence between both sexes, although cases in the ovary may be underdiagnosed because of the inaccessibility to the female gonads for examination and will probably never be diagnosed if they cause no symptoms and so have been found at autopsy or as incidental findings at laparotomy.[6,7]

Splenogonadal fusion may be diagnosed at any age (1-81 years), but is most often diagnosed in the first two decades of life (72% to 82%), with 50% of the cases diagnosed prior to 10 years of age.[8]

Most cases have occurred on the left side, but a single case of splenogonadal fusion occurring on the right side has been reported.[5]

Etiology

Splenogonadal fusion is thought to occur between the 5th and 8th gestational weeks. The etiology of such anomaly is uncertain, but may be secondary to injury of the fetus during this time. The spleen appears as a thickening of the layers of splenic mesothelium of the dorsal mesentery of the stomach. The gonad, arising as an indifferent sex gland from the medial surface of the wolffian body, comes to lie in front of the primitive kidney

on the posterior wall of the abdomen. Complex gastrointestinal rotation processes occur in the 5th and 8th week of development, resulting in a retroperitoneal position of duodenum and pancreas. This involves the fusion of adjacent celomic surfaces, during which the two organs are brought into close apposition, and fusion of the two is not inconceivable. Under such circumstances the gonad, in its descent during the latter weeks of intrauterine life, might readily drag along a tail of developing juxtagonadal splenic tissue.[1,5]

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.[5]

Pathology

In splenogonadal fusion, splenic remnants are bound to the surface of the testis/ovary or epididymis where it is separated from the gonadal tissue by a capsule. However, it can be even in an intragonadal location.[5]

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 hemopoetic potencies in gonadal mesenchyma.[5]

Putschar and Manion classified splenogonadal fusion into two types which occur with the same frequency[9];

(1) Continuous type – the spleen and splenic remnants adhered to the testis/ovary, epididymis, or deferent duct are attached through a cord like structure which may be fibrous, totally splenic or beaded with multiple splenic nodule which is usually in a retroperitoneal location, but may sometimes be intraperitoneal. All the cases described in female patients are the continuous type.[10,11]

(2) Discontinuous type – no connection is found between both structures. It may be due to breakdown of the attachment cord or represent a rare variant of an accessory spleen.

Sneath proposed that inflammation over two opposing peritoneal surfaces namely, the gonadal ridge and spleen, could cause fusion. During gonadal migration, the peritonealized adhesion would lengthen and develop as a cord continuous with the spleen or rupture during development, making it discontinuous with the spleen.[12]

Association with Other Congenital Abnormality

Splenogonadal fusion may be associated with other congenital anomalies such as testicular maldescent and cryptorchism, inguinoscortal hernia (due to the inguinal closure defect produced by the fibrous cord)[7], peromelia, micrognathia, hypoglossia, palatine defects, polymicrogyria, craniosynostosis, spina bifida, cardiac defects, diaphragmatic hernia, hypoplastic lung and anorectal abnormalities, abnormal fissures of lung and liver, hypospadias.[3] These have been reported in 30% of continuous forms.[13-19]

Association between splenogonadal fusion with limb abnormalities and micrognathia is common due to close proximity between differentiating precursor structures of shoulder bones[5], Meckel's cartilage[20] and developing spleen. It is listed as a rare disease by the office of rare diseases (ORD) of the National Institutes of Health (NIH). This condition affects less than 2, 00,000 in the US population. Most patients die shortly after birth although some do survive into childhood.[3]

Presentation

In 40% of cases, patients present with cryptorchidism, left inguinal hernia and a paratesticular mass, preferentially located close to the upper testicular pole. Occassionally, when the mass is intimately attached, differential diagnosis with paratesticular solid masses (rhabdomyosarcoma, lymphoma, metastasis etc) becomes very difficult.[5]

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. Falmann and Settle reported cases of splenogonadal fusion presenting with painful scrotal swelling secondary to malaria.[5]

However, presentation may be due to the complications of the fusion, e.g. bowel obstruction, rupture.

Other presentations are infertility, testicular pain after gastroenteritis and acute scrotal pain resembling testicular torsion.[19]

Athough exceptional, germ tumors associated to splenogonadal fusion have been reported in three patients, all of them with a history of associated cryptorchism.[21]

Diagnosis

Although preoperative diagnosis of this rare condition is difficult and rarely achievable, techniques of diagnostic imaging for confirmation are available for any suspected case of splenogonadal fusion.

Ultrasonography

The literature review revealed few cases where an ultrasound examination has been performed for splenogonadal fusion. No ultrasonographic differences exist between the accessory spleen and normal testis. A clearly outlined 2-3 cm mass in close paratesticular relationship is seen. Some authors advisees 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.[22]

However, Doppler ultrasonography may help to differentiate between splenogonadal fusion and paratesticular or testicular mass. The extensive vascularity observed by dopper ultrasonography of splenogonadal fusion is more extensive than any typical paratesticular or testicular tumor and may easily be compared to the vascularity of the patients own intraabdominal spleen.[23]

More recently, imaging using hifh frequency linear array probes with high color sensitivity showed different vascular patterns in focal testicular lesions, suggesting a “crisscross” pattern in primary malignancy which helps in further differentiation.[24]

Spleen Scintigraphy Enhanced by SPECT[20]

Only two cases out of four previously reported cases where the diagnosis was suspected preoperatively, confirmation done by radionuclide scanning, using radionuclide sulfur colloid/Tc99m imaging to demonstrate splenic uptake by the mass.

Care should be taken to obtain good scintigraphic visualization of the spleen, the whole abdomen and pelvis. The sctroum should be scanned in case of scrotal mass. As the bridge of splenic tissue connecting the spleen and the left gondad may be thin or discontinuous, it is imperative to obtain scans of impeccable quality. The appropriate use of image processing may be required to enhance the low-count density of thin or small splenic tissue fragments.

The noninvasiveness, relatively low price, sensitivity and specificity make it well suited for screening purposes.

CT Guided Biopsy[3,4]

If diagnosis is suspected preoperatively, it is advised to take a tissue sample for confirmation, so that partial mass excision sparing the testis, particularly in young males, may be performed. Histologically, the mass consists of completely normal splenic tissue, including red pulp and sinusoids.

Management

When the diagnosis is confirmed and no symptoms are present, it is controvertible whether further surgical intervention is needed. Surgical exploration is generally needed to rule out malignancy especially in patients with intratesticular mass. On surgical exploration, if a

mass is highly suspicious of splenogonadal fusion by visualizing a 2-3 cm solid red mass consisting of a parenchyma similer to that of spleen, the splenic tissue can be severed from the tunica albuginea in order to prevent orchidectomy, but in some case, orchidectomy may be required when the splenic tissue is adherent to the testes making it difficult to separate. Frozen section is crucial if splenogonadal fusion is suspected in patient with single gonad. Given the fact that the majority (82%) of patients were younger than 30 years at the time of diagnosis, orchidectomy should be avoided as often as possible. Familiarity with splenogonadal fusion allows one to recognize the lesion intraoperatively and preserve the testis.[1-4]

There is a growing importance of laparoscopy in the diagnosis and treatment of splenogonadal fusion especially in the cases of intraabdominal cryptorchism.[13]

Conclusion

Splenogonadal fusion is a rare congenital abnormality that is rarely suspected preoperatively. Careful medical history recording and high suspicion for the condition with confirmation by sophisticated investigations may prevent the patient from unnecessary orchidectomy.

References

1. Fahmy MAB. Rare case of Continuous and Discontinuous Splenogonadal Fusion in A single Testicle. Annals of Pediatric Surgery 2009;5(4):282-285.
2. Xiao-cao S, Chuan-jun DU, Ji-min C, Zhe-wei Z, Yi-qing QIU. Splenogonadal fusion- case report. Chinese Medical Journal 2008;12(4):383-384.
3. Chua HB. Spleno-gonadal fusion. Brunei Int Med Journal 2011;7(2):101-103.
4. Farzaneh MR, Abbasi MZ, Amini B. Splenogonadal fusion operated as a malignant tumor. Indian Journal of Medical Sciences 2010;35(2):157-159.
5. Maestro MA, Tello L, Franjo PD, Gonzalez ER, Pineiro LM. Splenogonadal fusion. Actas Urol Esp. 2010;34(3):293-295.
6. Stewart VR, Sellars ME, Somers S, Muir GH, Sidhu PS. Splenogonadal fusion. J Ultrasound Med 2004;23:1087-1090.
7. Putschar WGJ, Manion WC. Splenic-gonadal fusion. Am J Pathol 1955;32:15-33.
8. Duncan WL. Splenogonadal fusion: a case report and review of literature. J Pediatr Surg 2005;40:5-7.
9. Putschar WGJ, Manion WC. Splenic-gonadal fusion. Amer J Path 1956;32:15-33.
10. Andrews RW, Copeland DD, Fried FA. Splenogonadal fusion. J Urol 1985;133:1052-3.
11. Chen SL, Kao YL, Sun HS, Lin WL. Splenogonadal fusion. J Formos Med Assoc 2008;107:892-5.
12. Garragher AM. One hundred years of splenogonadal fusion. Urology 1990;35-471-475.
13. Braga LH, Braga MM, Dias MA. Laparoscopic diagnosis and treatment of splenogonadal fusion associated with intraabdominal cryptorchidism in a child. Pediatr Surg Int 1999;15:465-6.
14. Hizli F,Uygur MC, Irkkan C. Splenogonadal fusion: report of a case. Int J Urol 2005;12:591-2.
15. Cortes D, Thorup JM and Visfeld J. The pathogenesis of cryptorchidism and splenogonadal fusion: a new hypothesis. Brit J Urol 1996;77:285-90.
16. Guarin U, Dimitrieva Z, Ashley SJ. Splenogonadal fusion – a rare congenital anomaly demonstrated by 99mTc-sulfur colloid imaging : case report. J Nuci Med 1975;16:922-4.
17. Lipson AH. Amelia of the arms and femur/fibula deficiency with splenogonadal fusion in a child born to a consanguineous couple. Am J Med Genet 1995;55:265-8.
18. Gouw ASH, Elema JD, Bink-Boelkens MTE, de Jongh HJ, ten Kate LP. The spectrum of splenogonadal fusion: case report and review of 84 reported cases. Eur J Pediatr 1985;144:316-23.
19. Karaman MI, Gonzales ET Jr. Splenogonadal fusion:report of two cases. J Urol 1996;155:309-11.
20. Steinmetz AP, Rappaport A, Nikolov G, Priel IE, Chamovitz DL, Dolev E. Splenogonadal fusion diagnosed by spleen scintigraphy. The journal of nuclear medicine 1997;38:1153-55.
21. Imperial SL, Sidhu JS. Nonseminomatous germ cell tumor arising in splenogonadal fusion : a case report. Arch Pathol Lab Med 2002;126:1222-25.
22. Patel RV. Splenogonadal fusion. J Pediatr Surg 1995;30:873-4.
23. Balaji KC, Caldamone AA, Rabinowitz R. Splenogonadal fusion. J Urol 1996;156:854-856.
24. Bushby LH, Sriprasad S, Hopster D, Muir G, Clarke JL, Sidhu PS. Hich frequency color Doppler US of focal testicular lesions; crossing vessels (criss-cross pattern) identifies primary malignant tumors. Radiology 2001;221:449.

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