Discontinuous splenogonadal fusion diagnosed on computed tomography

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Abstract

Splenogonadal fusion is a very rare congenital anomaly which often manifests as a scrotal mass and rarely as cryptorchidism. It can be of continuous and discontinuous type based on the presence of a band of connecting splenic tissue. We report a rare case of discontinuous type of splenogonadal fusion in an adolescent male presenting as cryptorchidism. We emphasize the computed tomographic findings, which helped us in preoperative diagnosis and aided in appropriate management.

Key words: Computed tomography; cryptorchidism; splenogonadal fusion

Introduction

Splenogonadal fusion is a rare congenital anomaly due to fusion between of spleen and the gonad or remnant of mesonephros, often seen on the left side in a majority of cases (98%) and in males (95%).[1,2] Relatively low reported incidence in females is attributable to the internal location of gonads and inaccessibility of ovary to clinical examination.[3] Till date, approximately 150 cases of this entity have been described, however, most of the cases were discovered intraoperatively.[4]

Case Report

A 16-year-old male patient was referred to the urology department for evaluation of bilateral cryptorchidism. Physical examination demonstrated normal secondary sexual characters. Local scrotal examination showed empty scrotal sacs with oval soft tissue structure felt in the right groin, which was possibly inguinal testis. Ultrasound examination was initially done on GE Voluson E8 using 3.5 and 7 mHz convex and linear probes revealed an oval-shaped testis-like structure in the right groin with normal testicular echotexture and vascularity and non-visualized left testis. Ultrasound abdomen showed few splenunculi in the left hypochondrium and lumbar region and another oval-shaped mass with internal vascularity in left lumbar region. A diagnosis of intraabdominal testis was offered according to the sonography findings. Contrast-enhanced computed tomography (CECT) abdomen was advised for further assessment.

CT abdomen was done on GE discovery CT 750 HD 64 Slice that showed an oval hypodense lesion in the right medial inguinal region, possibly an inguinal testis in correlation with clinical examination and ultrasound findings. A well-defined oval hypodense soft lesion with two components of differential enhancement was seen in the left lumbar and iliac fossa region. Inferior soft tissue component was relatively small.

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and showed less enhancement, with its enhancement pattern similar to the right inguinal testis with draining left testicular vein, which was seen to be emptying into the left retroaortic renal vein [Figure 1]. The superior soft tissue component demonstrated homogenous enhancement, matching with the enhancement pattern of splenic parenchyma. Further analysis showed another vascular pedicle extending superiorly from this component and reaching up to the splenic hilum with multiple splenunculi seen along the course of these vessels [Figure 2]. Imaging findings suggested fusion of testis or a gonadal remnant, with splenunculus forming a conglomerate mass which resulted in cryptoorchidism on the left side. Features were suggestive of splenogondal fusion with no continuous band of connecting splenic parenchyma. The patient was managed surgically. Pre-anesthetic evaluation revealed significantly enlarged adenoids and tonsils with compromised airway; and hence, additional adenotonsillectomy was contemplated with the help of an ENT surgeon. Intraoperative finding showed a large pinkish mass with vascular pedicle in the left lumbar and iliac region which was resected [Figure 3]. Multiple splenunculi were also found. Right inguinal testis was managed by laparoscopic Stephen-Fowler stage procedure by high ligation of testicular artery. No evidence of fibrous cord or band of splenic parenchyma was seen between the left lumbar mass and spleen intraoperatively. Histopathological examination of the resected mass showed testicular tissue with seminiferous tubules with sertoli cells and occasional germ cells, and adjacent to testicular tissue splenic parenchyma tissue with congestion was found. Small adrenal rest was also found. No evidence of malignancy was seen.

Discussion

Splenogonadal fusion is a rare congenital anomaly often seen on the left side in a majority of cases (98%) in males (95%).[1,2] Relatively low reported incidence in females is attributable to the internal location of gonads and inaccessibility of ovary to clinical examination.[3] Till date, approximately 150 cases of this entity have been described, however, most of the cases were discovered intraoperatively.[4] Splenogonadal fusion can be of the continuous or discontinuous form based on the presence or absence of continuous band of splenic tissue of fibrous cord connecting the gonad. In the discontinuous form, ectopic splenic tissue is seen fused with the gonad and appears separate from the native spleen. The continuous variety is slightly more common and is more often associated with congenital anomalies such as limb defects, micrognathia, facial, cardiac anomalies, and persistent Mullerian duct syndrome.[5,6]

Exact mechanism of this anomaly still remains incompletely determined. The possible explanation expressed in various previous studies is that the splenic anlage develops from the dorsal mesogastrium in the 4th to 5th week of gestation, and during the gut rotation it comes in close proximity to the developing mesonephros and gonad between the 5th and 8th week when anomalous fusion can occur.[7,8] Adrenal
Splenogonadal fusion is a rare congenital anomaly and most cases are incidentally seen during exploration of groin or abdomen for hernia or cryptorchidism.[1,4] Lack of a definitive preoperative diagnosis often leads to inadvertent sacrifice of the viable testis as the fused splenic remnant can give spurious and alarming appearance of malignant degeneration. However, cases of remote risk of malignancy in retained gonadal remnant has been reported;[10] hence, surgical excision of the splenogonadal remnant was contemplated in our case as the surgeons were able to salvage the right testis.

Ultrasound in splenogonadal fusion shows hypoechoic paratesticular mass which is difficult to differentiate from neoplastic mass, and the rarity of this clinical entity results in further difficulty in accurate preoperative diagnosis based on ultrasound.[11] Doppler signal in the aberrant splenic tissue demonstrates central vascularity with branching pattern, which is different from criss-cross vascular pattern in neoplastic mass; however, this differentiation is still subjective.[10] CT findings in splenogonadal fusion have been very rarely described in the literature.[12‑14] Splenic tissue is seen as homogeneously enhancing non-calciﬁed soft tissue mass on CT, demonstrating homogeneous enhancement characteristics similar to the splenic parenchyma. This aberrant splenic tissue is seen in scrotal sac or in the intra-abdominal location on the left side and is fused with the testis. In the continuous type, there is a contiguous bridging cord-like splenic tissue connected to the spleen, and in the discontinuous type, a discrete splenunculus separate from the spleen is seen fused with the testis. The principle testicle demonstrates relatively less enhancement than splenic tissue with a differential enhancement pattern. CT can accurately depict the dual vascular supply of splenogonadal remnant which is derived from the gonadal vessels and splenic hilar vessels.[11,13] We had been able to accurately depict the vascular supply of the splenic and gonadal components and described these findings in detail in present case. Based on the characteristic CT imaging findings which are concurring with previous studies,[12‑14] we suggested the possibility of discontinuous type of splenogonadal fusion, which was conﬁrmed by intraoperative and histopathological ﬁndings.

Although ultrasound is the initial investigative modality of choice in cryptorchidism, in cases where undescended testis cannot be detected, cross-sectional imaging modalities such as CT are helpful in accurate localization and in detecting other rare anomalies such as splenogonadal fusion, as in our case, thus aiding in surgical management. There are isolated reports of splenogonadal fusion diagnosed on nuclear scintigraphy with Te99m-sulfur colloid scan by identifying ectopic spleinic tissue adjacent to gonad.[15]

**Conclusion**

Splenogonadal fusion is a rare congenital anomaly presenting with cryptorchidism or scrotal mass and is very rarely diagnosed preoperatively. Computed tomography can accurately diagnose this anomaly, thus guiding the surgeon in appropriate management. Knowledge of this clinical entity and its imaging finding helps a radiologist in correct diagnosis.

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**Conflicts of interest**

There are no conflicts of interest.

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