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Case Report

Radiology Section

A Rare Case of Atretic Uterus Causing Compression Over the Sigmoid Colon

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ABSTRACT

In pseudo-hermaphroditism a person is born with primary sex characteristics of one sex but develops the secondary sexual characteristics of different sex from what would actually be expected on the basis of the primary sex (testis or ovaries).

Sometimes, there is partial appearance of the either of the external sex organs together that is a one between a typical penis and clitoris. In rest of the cases, the expected external sex organs are seen. Thus, pseudo-hermaphroditism can be difficult to identify until puberty. The condition may also remain hidden until adulthood.

Male pseudo-hermaphroditism is an individual with XY karyotype and testes is present with a partial or complete female phenotype. This condition is attributed to hypoandrogenism in XY individuals. There is a lack in the action or presence of testosterone and dihydrotestosterone.

This is a case report of a 60-year-old male who presented to the surgery out-patient services with complain of lower abdominal pain since 6 months. After proper clinical history and consent, the patient was subjected to endoscopy and contrast enhanced CT of abdomen. On endoscopy, there was restriction at passing the probe beyond the distal end of sigmoid colon and the probe could not be passed beyond it. A stricture of unknown etiology was reported. CT revealed an ill-defined elongated enhancing soft tissue lesion noted in right side of pelvis superolateral to the urinary bladder causing compression over the sigmoid colon with no obvious bowel connection. Exploratory laparotomy was then performed which revealed an elongated soft tissue lesion adherent to the sigmoid colon without obvious communication to the bowel lumen. The organ of origin could not be confirmed. The lesion was excised and sent for histopathology which revealed atretic uterine tissue.

Keywords: Compression, Male pseudo-hermaphrodite, Phenotype

CASE REPORT

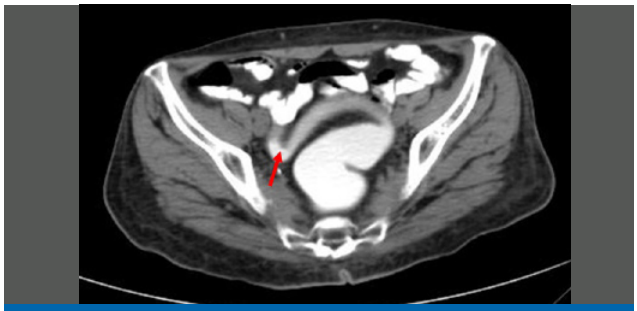
A-60-year old married male presented to the Surgery Out-patient services with the complaint of lower abdominal pain since 6 months and localized to the right iliac region with no history of any similar pain in the past. Clinical examination revealed mild tenderness in the right iliac quadrant. The patient had a history of a testicular surgery 2 years back which was uneventful with no documentation of the surgery available. The patient was a father of 2 children.

After taking consent from the patient, endoscopy and CT abdomen study was performed. On endoscopy, there was restriction at passing the probe beyond the distal end of sigmoid colon and the probe could not be passed beyond it. A stricture of unknown etiology was reported.

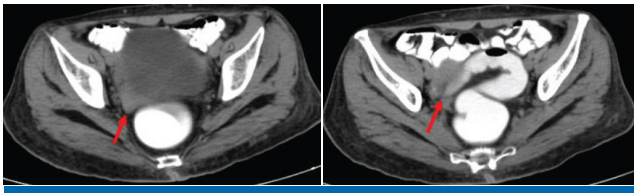
On contrast enhanced CT of the abdomen and pelvis, the sigmoid colon appeared normally opacified with rectally administered contrast material. There was no obvious focal mass or significant wall thickening [Table/Fig-1].

In lower sections, there was evidence of an ill-defined elongated enhancing soft tissue lesion noted in right side of pelvis measuring approximately 9x5.2x4 cm superolateral to the urinary bladder [Table/Fig-2] extending inferiorly upto the recto-vesical pouch [Table/Fig-3]. The lesion was abutting the mid sigmoid colon, causing mild narrowing which was responsible for the right lower quadrant pain in right iliac region the patient was complaining of. A possibility of a neoplastic mass in mesentry, exophytic lesion from small bowel wall or sigmoid, gastrointestinal stromal tumour was kept. The right scrotal sac was empty [Table/Fig-4] which led us to believe a rare possibility of undescended intraabdominal testis with associated neoplastic mass. There was also evidence of calcified prostate gland in lower abdominal sections as depicted on the axial CT images [Table/Fig-5].

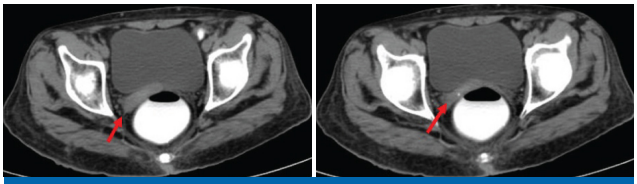
Exploratory laparotomy was planned corresponding to the



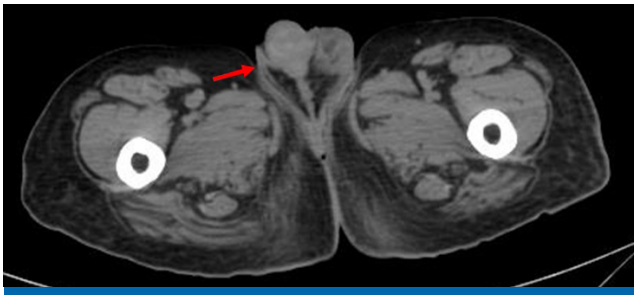
[Table/Fig-1]: CECT showing a normally opacified sigmoid colon with rectally administered contrast material.



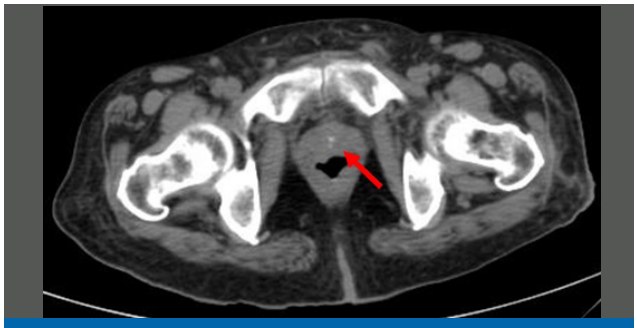
[Table/Fig-2]: CECT showing an ill-defined elongated soft tissue lesion in right side of pelvis superolateral to the urinary bladder.



[Table/Fig-3]: CECT reveals the lesion extending inferiorly upto the rectovesical pouch.



[Table/Fig-4]: CECT showing an empty right scrotal sac



[Table/Fig-5]: CECT showing the calcified prostate gland.

reports on CT and endoscopy, done in the operation theatre which revealed an elongated soft tissue lesion adherent to the sigmoid colon without obvious communication to the bowel lumen. The organ of origin could not be confirmed. The lesion was excised and sent for histopathology.

Histopathology report revealed uterine tissue with atretic endometrium implying atretic uterine tissue. The patient was called for follow-up after 2 weeks and then 4 weeks. There was relief of his complaints with no post procedural complications.

DISCUSSION

Pseudo-hermaphroditism is so called when a person is born with primary sex characteristics of one sex but develops the secondary sexual characteristics of a different sex from what would actually be expected on the basis of the primary sex (testis or ovaries) . Sometimes, there is partial appearance of the either of the external sex organs together that is a one between a typical penis and clitoris. In rest of the cases, the expected external sex organs are seen. Thus, pseudo-hermaphroditism can be difficult to identify until puberty. The condition may also remain hidden until adulthood [1].

Male pseudo-hermaphroditism is an individual with XY karyotype and testes is present with a partial or complete female phenotype. This condition is attributed to hypoandrogenism in XY individuals. There is a lack in the action or presence of testosterone and dihydrotestosterone [2].

Sexual development disorders are congenital in origin and occur due to atypical development of gonadal, chromosomal or anatomical sex. They are classified into four categories on the basis of gonadal histologic features:

Type I is female pseudo-hermaphroditism (46,XX with two ovaries);

Type II is male pseudo-hermaphroditism (46,XY with two testes);

Type III is true hermaphroditism (ovotesticular) (both ovarian and testicular tissues) and,

Type IV is gonadal dysgenesis, either mixed (a testis and a streak gonad) or pure (bilateral streak gonads).

Conception is the time where the chromosomal basis for sex is occurs. The genital structures, both external and internal remain undifferentiated till upto 6 weeks gestation. The 3 components of genital system which act as precursors are the germ cells, genital ridge and two sets of internal sex ducts, which are, the müllerian-paramesonephric ducts and the wolffian-mesonephric ducts. The genital ridge becomes either an ovary or a testis at around 6 weeks of gestation. These undifferentiated gonads are then populated by the germ cells. Testis determining factor, which is encoded by the SRY gene located on the short arm of the Y-chromosome guides the testicular development. The germ cells then, differentiate into sertoli cells and leydig cells in the genital ridge. Sertoli

cells secrete Müllerian Inhibiting Substance (MIS), and Leydig cells produce testosterone. MIS causes complete regression of the müllerian ducts is caused by MIS, whereas maturation of spermatogonia occurs under influence of testosterone and paracrine and endocrine actions lead to male phenotype development. When the Y chromosome is absent, at around 11-13 weeks, the gonads differentiate into ovaries [3,4].

Sexual developmental disorders are one of the most fascinating conditions clinically encountered. There has been a rapid advancement in diagnosis of these conditions in recent years. Clinicians can now promptly make the accurate diagnosis, counsel the parents and give the particular therapeutic options [5].

Male pseudo-hermaphrodites are 46, XY genetic males with normal or mildly defective testes. This is due to deficiency in testosterone and di-hydro-testosterone production caused by a deficiency of 5-α reductase [6].

The problems with gender assignment in these cases associated with the social and psychological implications has led to delayed surgical genital reconstruction in some cases.

In our case report, a father of 2 children presented with the complaint of lower abdominal pain in the right pelvic region and a possibility of a neoplastic mass in mesentery was kept. The mass was excised during exploratory laparotomy and sent for section study. Surprisingly, histopathology revealed atretic uterus.

CONCLUSION

Intersex disorders are clinical problems which are complex and require a coordinated assessment and management by a group of specialists. This is a rapidly developing field with evolving active clinical work-up. Identifying the pathology leading to each disorder constitutes the future goal for better long term survival. CT in such a situation plays a supportive role in diagnosis of intersex disorders. Multiple factors are to be taken into consideration to assign sex with a complete informed parental discussion. Counselling should begin as soon as possible.

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