CASE REPORT

Transverse testicular ectopia with persistent mullerian duct syndrome

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SUMMARY

Persistent mullerian duct syndrome (PMDS) is a rare form of male pseudohermaphroditism caused by defects in synthesis or actions of mullerian inhibiting factor characterised by persistence of mullerian duct structures in a normal karyotype male. Transverse testicular ectopia (TTE) is a rare disease in which both testes are located in the same inguinal canal. We report a case of PMDS with TTE in a 1-year-old child who presented with non-palpable testis on right side with hernia on left side. Left herniotomy with bilateral trans-septal orchidopexy was done in this patient.

BACKGROUND

Persistent mullerian duct syndrome (PMDS) is a form of male disorder of sexual differentiation with persistence of mullerian duct structures characterised by the presence of uterus and fallopian tubes in a phenotypically and genotypically normal male.¹ Transverse testicular ectopia (TTE) is a rare form of testicular ectopia in which both testes are located in the same inguinal canal. We report a case of PMDS with TTE in a 1-year-old child.

CASE PRESENTATION

A 1-year-old male child presented to surgery outpatient department with complaints of swelling in left inguinal region and absent right testis since birth. The child was born by a full-term normal vaginal delivery. There was no history of any other congenital anomaly and was adequately immunised. There was no history of undescended testes or any congenital anomaly in the family. On examination, a swelling of size 1 × 1 cm was present in left inguinal region. Swelling was more apparent on standing or crying. Right testis was not palpable. There was no developmental delay and examination of other systems was within normal limits.

INVESTIGATIONS

Haemoglobin was 11.1 g/dL. Total leucocyte count was 6500/mm³.

TREATMENT

The patient was taken up for elective surgery with a diagnosis of left congenital hernia with right undescended testis. On exploration, left hernia sac had mullerian duct remnant with both the testes (figure 1). Left herniotomy with bilateral trans-septal subdartos orchidopexy was done under general anaesthesia.

OUTCOME AND FOLLOW-UP

Postoperative period was uneventful and the patient was discharged on postoperative day 2.

DISCUSSION

TTE is a rare form of ectopic testes. TTE was first reported as a finding at autopsy by Von Lenhossek in 1886.²

PMDS is caused by defects in synthesis or actions of mullerian inhibiting factor.³ It is characterised by normal male karyotype with persistent mullerian duct derivatives.

Jordan described TTE with PMDS for the first time in 1895.² PMDS with TTE is a rare anomaly that is usually encountered at the time of laparoscopy or while exploring for inguinal hernia. This anomaly combines abnormal testicular descent with regression of fetal mullerian structures.

Hutson *et al* classified PMDS into three groups.¹ Group A is the female type where testes are in position of normal ovaries. Group B is the male type with one testis in hernial sac or scrotum with the uterus and tubes. Group C is also male type with both testes found in same hernial sac with associated tubes and uterus. Our patient was Group C PMDS.

Figure 1 Intraoperative picture showing bilateral testes in the same inguinal canal with persistent mullerian duct structures.

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Most of the patients are diagnosed at the time of surgery. However, ultrasonography and MRI help in diagnosing it preoperatively. Hormonal assessment is not done routinely.⁴

In our case, imaging was not done and patient was taken for upfront surgery.

Treatment of PMDS is surgical and involves correction of cryptorchidism. Various methods have been described like fixation of testes on opposite side through suprapubic subcutaneous tunnel or herniotomy with trans-septal fixation of testes.⁵

Orchidectomy can be done if testes cannot be mobilised or if there is a risk of malignancy. Orchidectomy should be followed by lifelong supplementation of testosterone.

Laparoscopy can be used for diagnosis and management of undescended testes. Kamble *et al*¹ were the first to describe total laparoscopic repair for TTE with PMDS in 2015. Laparoscopy is now becoming the route of choice for management of such cases.

There are reports of seminoma, embryonal cell carcinoma, clear cell adenocarcinoma and yolk sac tumours in patients with PMDS.⁶ However, recent studies suggest the incidence of these is similar to that of a normal cryptorchid child.⁷

Removal of persistent müllerian duct structures is associated with high rate of injury to vas deferens. Hysterectomy is only

indicated when these structures limit the placement of testes in scrotum.⁸

In our case, herniotomy with trans-septal fixation of testes was done and the patient was kept on follow-up.

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Learning points

- Persistent müllerian duct syndrome with transverse testicular ectopia is a rare anomaly which combines abnormal testicular descent with failure of regression of fetal müllerian structures.
- Diagnosis is usually made at the time of surgery.
- Treatment involves correction of cryptorchidism.
- Removal of persistent müllerian structures is not mandatory and is associated with high rate of injury to vas deferens.

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