CASE REPORT

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Risk of contralateral testicular malignancy in patients with persistent Müllerian duct syndrome and primary testicular malignancy. A case report and a comprehensive review of literature

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Abstract

Introduction: Persistent Müllerian duct syndrome (PMDS) is a rare condition in phenotypic and karyotypic males characterized by retained Müllerian structures, such as the uterus and fallopian tubes, and is often associated with cryptorchidism and increased gonadal malignancy risk.

Aim: To evaluate the risk of contralateral testicular neoplasia in patients with PMDS presenting with primary testicular malignancy and to review the available literature on bilateral gonadal involvement.

Case study: We report a case of PMDS in a 30-year-old phenotypic and karyotypic male who presented with abdominal pain, weight loss, and a palpable abdominal mass. Imaging showed a large abdominopelvic mass with bilateral cryptorchidism, and surgery revealed an intra-abdominal right testicular tumor associated with Müllerian structures. Histopathology confirmed right testicular seminoma, while the grossly normal contralateral intra-abdominal testis showed focal gonadoblastoma and germ cell neoplasia in situ (GCNIS).

Discussion: This observation suggests the possibility of occult contralateral gonadal pathology in PMDS, particularly with bilateral cryptorchidism; however, as a single-case finding, it should be interpreted cautiously and not considered definitive evidence of bilateral risk. In our literature review, 63 reported cases of testicular malignancy in PMDS were identified, with bilateral involvement, including malignancy or GCNIS, described in a subset. Although this supports the possibility of contralateral pathology, the evidence is derived from highly selected reports and may be influenced by publication bias.

Conclusions: This case highlights the need for careful evaluation of both gonads in PMDS and consideration of occult contralateral pathology, while larger collaborative studies are needed to define its true incidence and guide evidence-based management.

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1. Introduction

Persistent Müllerian duct syndrome (PMDS) is a rare form of male pseudo-hermaphroditism characterized by the presence of Müllerian duct structures, including the uterus and fallopian tubes, in otherwise phenotypically and karyotypically male individuals. PMDS is frequently associated with cryptorchidism, which may predispose to gonadal malignancy.

We report a case of a 30-year-old phenotypically and karyotypically male with PMDS who presented with abdominal pain, weight loss, and a palpable abdominal mass. Imaging revealed a large abdominopelvic mass with bilateral cryptorchidism. Surgical exploration demonstrated an intra-abdominal right testicular tumor associated with Müllerian structures. Histopathological evaluation confirmed seminoma in the right testis. Notably, the contralateral intra-abdominal testis, which appeared grossly unremarkable, showed a focus of gonadoblastoma and germ cell neoplasia in situ (GCNIS) on detailed examination.

This observation raises the possibility of occult contralateral gonadal pathology in PMDS, particularly in the setting of bilateral cryptorchidism. However, given the limitations of single-case observations, this finding should be interpreted with caution and not be considered definitive evidence of bilateral risk.

2. Aim

We conducted a literature review and identified 63 reported cases of testicular malignancy in PMDS, among which bilateral involvement (including malignancy or GCNIS) was described in a subset of cases. While this suggests that contralateral pathology may occur, the available data are derived from highly selected reports and may be influenced by publication bias.

This case highlights the importance of thorough evaluation of both gonads in PMDS and suggests that the possibility of occult contralateral pathology should be considered. Larger studies and collaborative data are required to better define the true incidence and guide evidence-based management strategies.

3. Case study

A 30-year-old male presented with complaints of pain in the right lower abdomen, loss of appetite, and weight loss of 4 kg over the previous three months. He had no significant past medical history, no comorbidities, no addictions, and no family history of malignancy. Clinical examination revealed a palpable lump in the right iliac fossa. A contrast-enhanced CT (CECT) scan of the abdomen showed a heterogeneously enhancing mass lesion measuring 10.6 × 8.2 × 11.5 cm in the right iliac fossa, closely abutting the medial wall of the cecum and the right external iliac vessels, with the probable site of origin being the terminal ileum. There was no pelvic or retroperitoneal lymphadenopathy. Colonoscopy findings were normal. A transabdominal biopsy of the mass, performed elsewhere, revealed grade 3 neuroendocrine carcinoma. Therefore, with a working diagnosis of neuroendocrine carcinoma probably originating from the terminal ileum, he was referred to our hospital for further management.

The patient underwent exploratory laparotomy. Intraoperative findings revealed a large solid-cystic lesion, approximately 10 × 10 cm in size, arising from the right testis and adherent to the anterior abdominal wall. Additionally, a rudimentary uterus was observed, with both testes connected to rudimentary fallopian tubes and a blind vaginal sac (Figure 1–4). The patient underwent primary cytoreductive surgery, including bilateral orchiectomy, resection of all rudimentary Müllerian structures including the uterus, both tubes, and

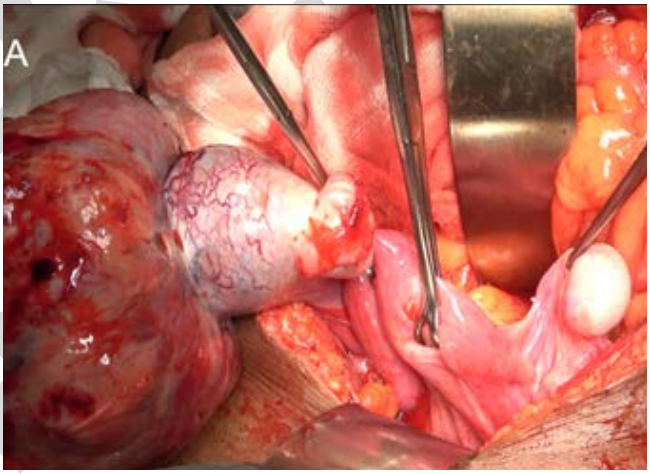

Figure 1. Intraoperative image with a rudimentary uterus, bilateral testicles associated with the fallopian tubes, resembling feminine ovaries. The large tumor is arising from the right testicle.

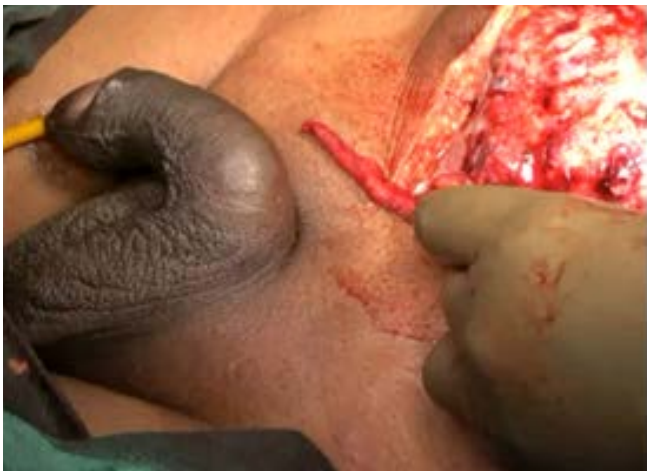

Figure 2. Well-developed scrotum and penis.

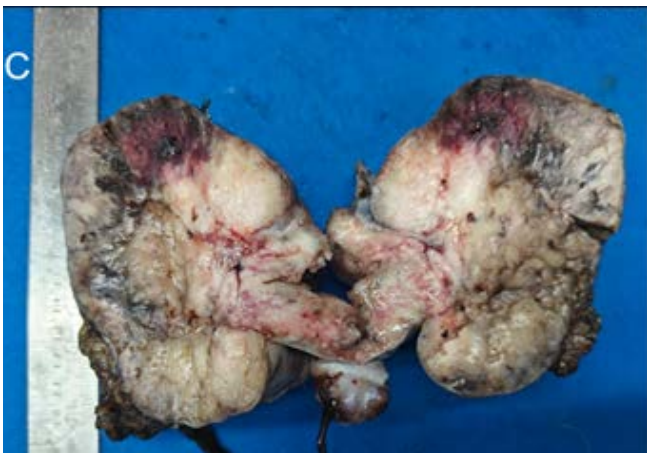

Figure 3. A cut section of the tumor arising from the right gonad.

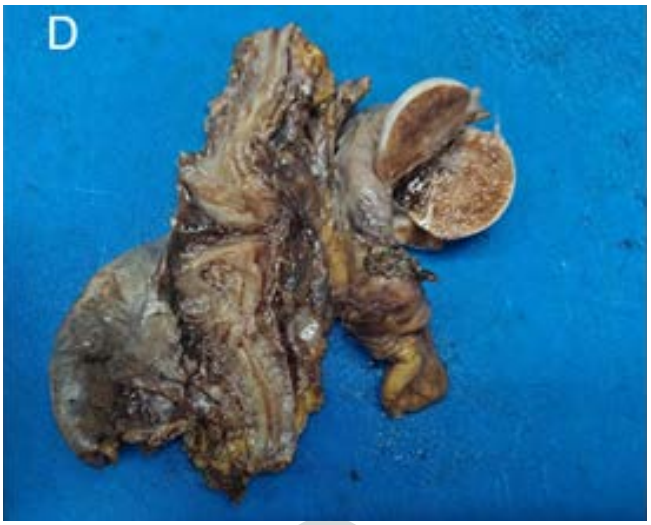

Figure 4. A coronal division of the rudimentary uterus and other Müllerian structures, along with the left-sided testis. Cut sectioned structures have been reflected superiorly.

rudimentary vagina, along with appendectomy, pelvic peritonectomy, and infracolic omentectomy.

Notably, the patient had been married for 7 years with a history of infertility and had a well-developed penis and bilateral empty scrotal sacs with absent testes. There was no history of consanguinity. Secondary sexual characteristics, including pubic hair, axillary hair, facial hair, deep voice, and male-pattern breast development, were present.

Histopathological analysis identified an 11 × 10 cm mass in the right testis, consistent with seminoma. Immunohistochemistry (IHC) demonstrated strong nuclear positivity for SALL-4 along with significant membranous positivity for CD117, further confirming the diagnosis of seminoma (Figure 5). The adjacent testicular parenchyma exhibited atrophied tubules and GCNIS. The

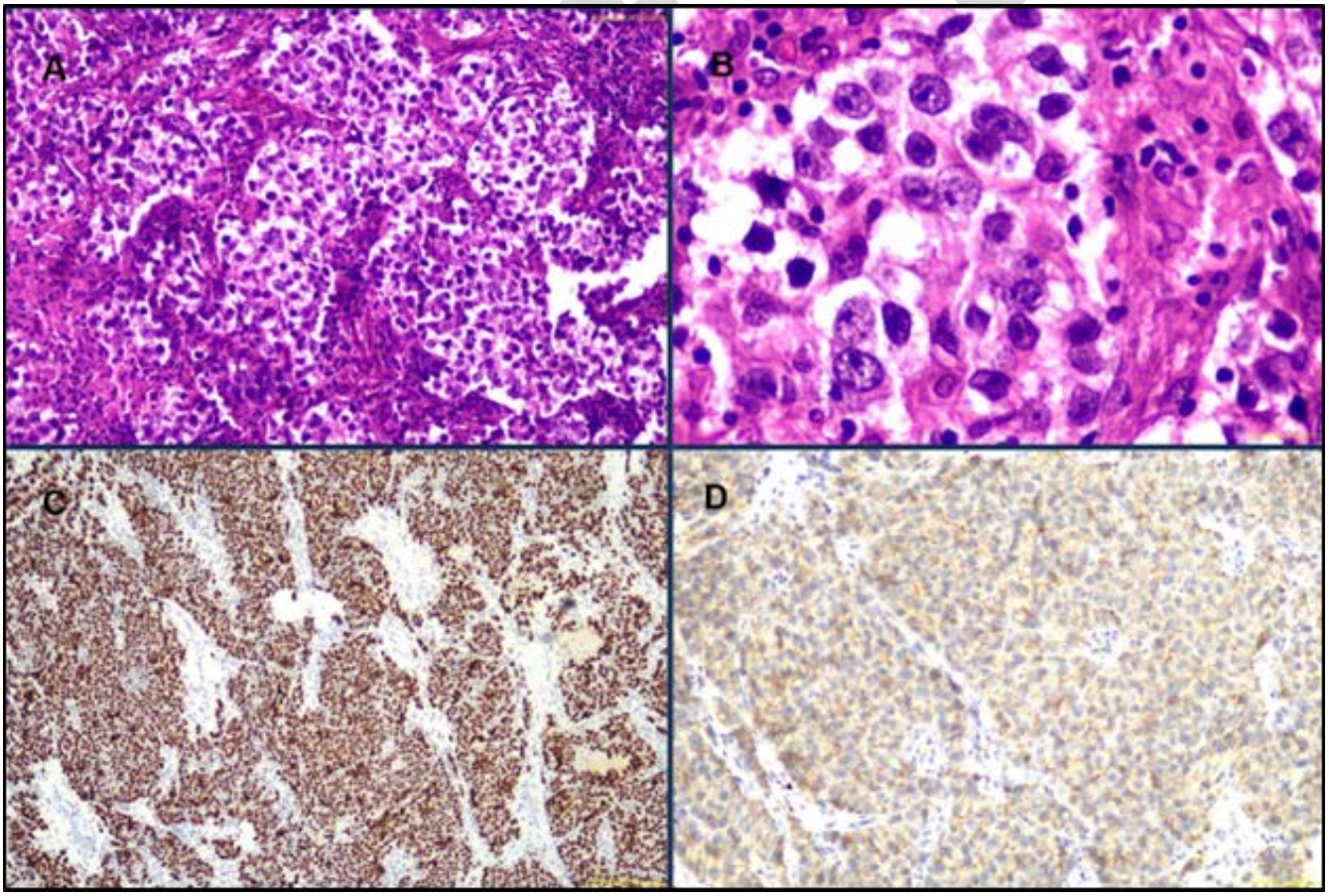

Figure 5. Histopathological examination of seminoma arising from right gonad (Testis): (A) Low power view (HE, 100x) showing nests of atypical cells surrounded by fibrous septae. (B) High power view (HE, 400x) showing nests of atypical cells with large nuclei, prominent nucleoli, and abundant clear cytoplasm. Surrounding fibrous septae show lymphomononuclear inflammation. (C) Low power view (SALL-4, 100x) showing strong nuclear positivity for SALL-4. (D) Low power view (CD117, 100x) showing moderate to strong membranous positivity for CD117.

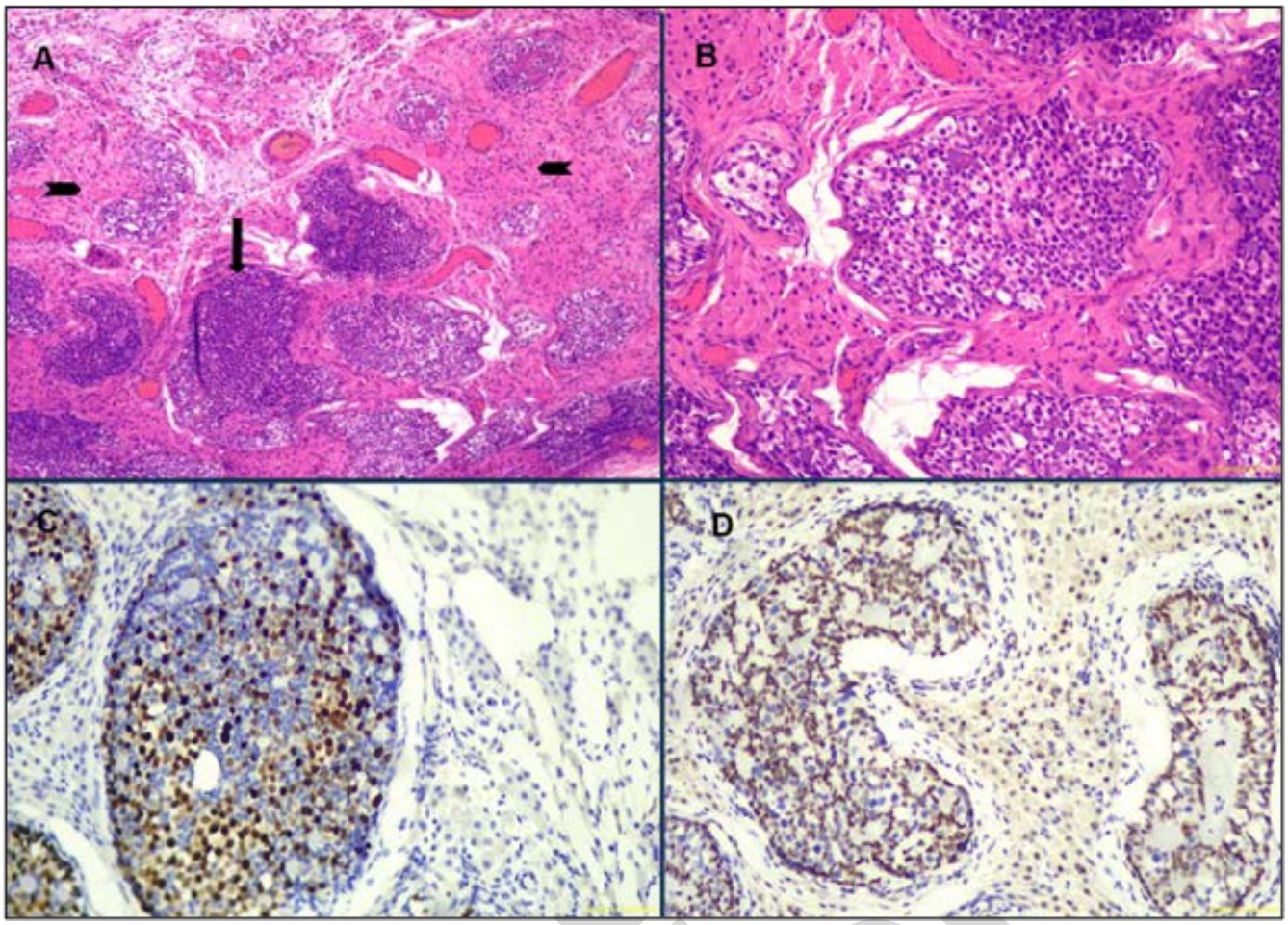

Figure 6. Gonadoblastoma arising from left gonad: (A) Low power view (HE, 100x) showing nests of atypical cells (arrow), along with areas showing Leydig cell hyperplasia (arrowhead). (B) High power view (HE, 400x) showing nests of atypical cells, with a mixture of large, atypical germ cells and small stromal cells. (C) Low power view (SALL-4, 100x) highlighting larger germ cells with strong nuclear positivity for SALL-4. (D) Low power view (SF-1, 100x) highlighting stromal cells.

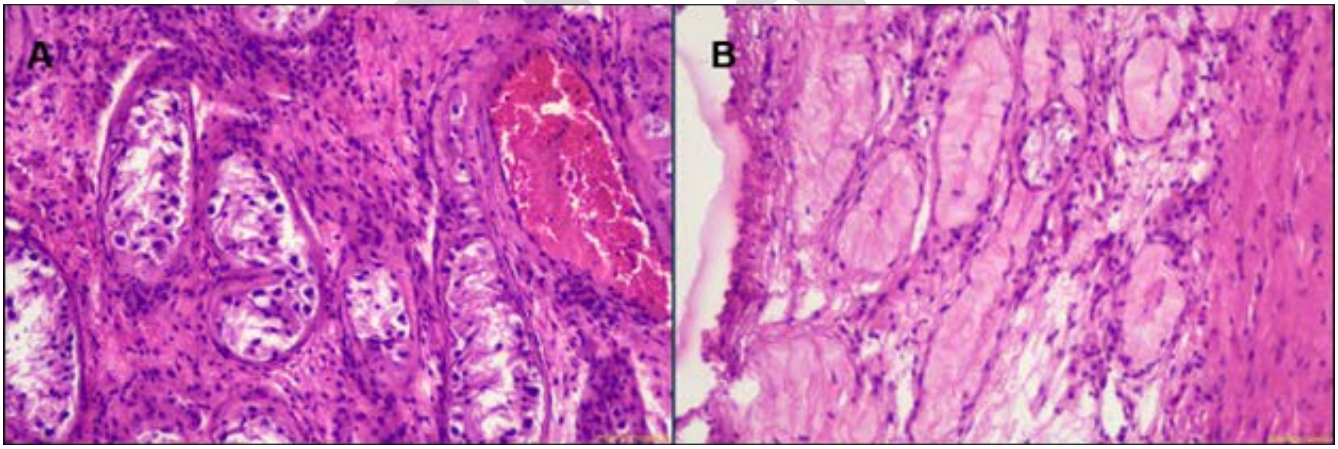

Figure 7. shows GCNIS and atrophic hyalinized tubules in left gonad. Panel A. Low power view (H&E, 100X) showing seminiferous tubules with atypical germ cells. The cells show high Nuclear: Cytoplasmic ratio, nuclear enlargement, hyperchromasia, prominent nucleoli and moderate cytoplasm. Panel B. Low power view (H&E, 100X) showing atrophic and hyalinized seminiferous tubules.

rudimentary uterus showed atrophic changes with no remarkable pathology. The left testis showed an atrophied gonad with a 1.2 cm focus of gonadoblastoma and surrounding GCNIS (Figures 6 and 7). Extensive grossing of the gonad did not reveal any ovarian tissue or dysgenetic gonad. The pelvic peritoneum and omentum were free of tumor. Conventional cytogenetic study revealed a normal male karyotype (46,XY) in all metaphases (Figure 8). He received a single cycle of carboplatin-based chemotherapy and is doing well at 1-year follow-up after surgery. He is currently on maintenance testosterone therapy.

PMDS has three primary clinical presentations. Type A is characterized by bilateral cryptorchidism,

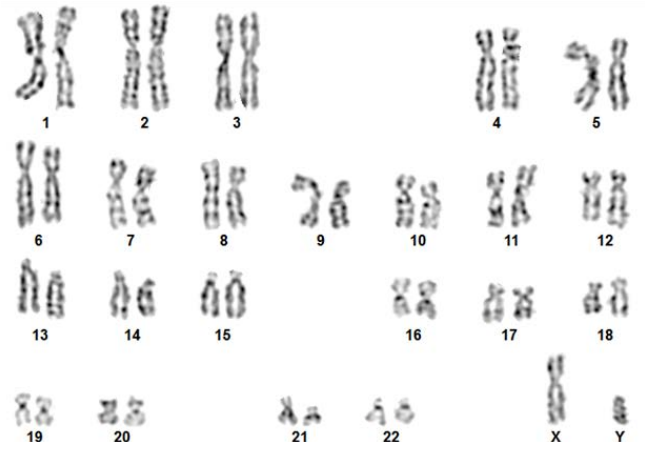

Figure 8. Karyotype analysis. Both stimulated and unstimulated cultures of peripheral blood were analyzed, followed by metaphase cell preparation, GTG banding, and karyotype analysis. Twenty metaphase cells were counted, and fourteen were karyotyped and analyzed according to ISCN 2020 guidelines, with a band resolution of 400. The result was a normal male karyotype (46,XY) with no evidence of clonal, numerical, or structural abnormalities.

with the testes situated in the pelvic region adjacent to the fallopian tubes and uterus, analogous to the normal position of the ovaries. Type B involves unilateral cryptorchidism, where one testis is found in an inguinal hernia along with a fallopian tube and uterus, referred to as *hernia uteri inguinalis*. Type C is known as transverse testicular ectopia, in which both testes and portions of the Müllerian structures herniate into a single processus vaginalis.[1] In the published literature, bilateral testicular malignancy or GCNIS was reported in approximately 17% (n = 11) of PMDS patients with testicular cancer; however, this proportion is derived from a highly selected group of reported cases.

We searched PubMed for reports of persistent Müllerian duct syndrome associated with gonadal or Müllerian duct malignancy published between 1981 and July 2024 using predefined search terms: 'Persistent Müllerian Duct Syndrome' AND 'Testicular cancer' OR 'Gonadal malignancy' OR 'Gonadal tumor' OR 'Seminoma' OR 'Müllerian malignancy' OR 'Uterine cancer.' This search identified 125 articles. When multiple publications described the same patient, duplicate cases were identified by cross-referencing demographic, clinical, and pathological details and were counted only once. Bilateral involvement was defined as the presence of synchronous or metachronous malignancy or GCNIS affecting both testes, irrespective of histological concordance. Given the rarity of PMDS and the predominance of isolated case reports in the literature, a narrative review approach was adopted rather than a formal systematic review. Reports published in English language only were included; non-English reports were excluded.

After screening these articles, we identified 63 cases of testicular malignancy arising in PMDS: 38 seminoma, 15 mixed germ cell tumor (MGCT), 4 non-seminomatous germ cell tumor (NSGCT), 2 teratoma, and 4 with unspecified details. There were also 2 cases of GCNIS (including 1 bilateral), 2 cases of prostatic cancer, 4 cases of uterine cancer (1 uterine adenosarcoma, 2 clear cell adenocarcinomas, and 1 adenocarcinoma of remnant Müllerian ducts probably of endocervical origin), 1 leiomyoma of the uterus, 1 syringoid carcinoma of the scrotal skin, and 1 case of lymphoma.

Among the 63 cases of testicular malignancy, 53 were unilateral and 10 were bilateral. Eleven cases, including 10 cases of bilateral testicular malignancy and 1 case of bilateral GCNIS, are discussed in detail[2–11] (Table 1). The median age at presentation was 34 years (range 17–50 years). Clinical presentations included obstructive uropathy with hydroureteronephrosis in 2 patients, low abdominal pain in 1, abdominal mass in 2, testicular mass in 1, infertility in 1, acute urinary retention in 2, torsion of the testis in 1, and undescended testis in 1. PMDS type A was present in 9 cases and type B in 2 cases (Table 1). The median longest tumor diameter was 11 cm (range 7–20 cm). All patients had cryptorchidism. Neoadjuvant chemotherapy (NACT) was administered in 3 patients, adjuvant therapy in 3, and adjuvant radiotherapy along with chemotherapy in 2. Retroperitoneal nodal enlargement was seen in 2 patients, while distant metastasis in the form of pleural effusion was observed in 1 patient. Histologically, in the presenting or ipsilateral gonad, there were 3 cases of NSGCT, 7 cases of seminoma, and 1 case of microinvasive GCT. The contralateral gonad revealed 2 cases of NSGCT (although no residual tumor was found after NACT), 6 cases of seminoma, 1 case of mixed GCT, and 2 cases of GCNIS. No disease was reported at the last follow-up in 7 patients, with a median follow-up period of 30 months (range 6–88 months). Proper management of cryptorchidism, involving inguinal or abdominal exploration with orchidopexy during childhood, was noted in 3 cases; however, it was neglected in 7 cases. Details were missing in 1 report. Additionally, 1 patient had undergone inguinal hernia repair.

4. DISCUSSION

PMDS is an uncommon condition involving abnormalities of sexual differentiation due to disruption of the Müllerian inhibiting factor (MIF) pathway. This disruption prevents regression of the fetal Müllerian ducts, resulting in the development of a uterus, cervix, and fallopian tubes in an individual who is genetically and

Table 1. Reported cases of bilateral testicular malignancy or germ cell neoplasia in situ in patients with PMDS.

No	Author, year (reference No)	Age, y	Clinical presentation	LDT at surgery, cm	Cryptorchidism	PMDS Type	Distant metastasis	NACT/ACT	Histological finding of resected testis	Histology of C/L gonad	Outcomes, months
1	Asthana, 2001 (2)	34	Bilateral PE, Obstructive uropathy	7 × 6	Yes	A	Pleural effusion	NACT (3 BEP)	NSGCT (preop cytology and markers). No tumor in gonad after NACT	NSGCT (preop cytology and markers). No tumor in gonad after NACT	NED, 36
2	Asthana, 2001 (2)	25	Bilateral OU	10 × 10	Yes	A	Nil	NACT (3 BEP)	NSGCT on preoperative markers. No tumor	NSGCT on preoperative markers. No tumor	NED, 18
3	Beatty, 2013 (3)	50	Low abdominal pain	NA	Yes	A	NA	ACT (2Carboplatin)	Seminoma	Seminoma	NA
4	Bucci, 2002 (4)	36	Testicular mass	NA	Yes	A	NA	NACT (BEP)	Seminoma	Seminoma	NA
5	Duenas, 2001 (5)	NA	Bilateral abdominal mass	NA	Yes	A	NA	Adj (Cisp/Cylophos + RT)	Seminoma	Seminoma	NED, 88
6	Eastham, 1992 (6)	34	Infertility	11 × 8	Yes	B	Nil	NA	YST	Mixed (Seminoma predominantly, smaller YST)	NA
7	Kazim, 1985 (7)	24	AUR	NA	Yes	A	NA	NA	Seminoma	Seminoma	NED, 30
8	Kulkarni, 1990 (8)	30	Lump in hypogastrium, right scrotum	15 × 10	Yes	B	Nil	Adj RT	Seminoma	Seminoma	NED, 24
9	Masereel, 1999 (9)	34	Torsion testis	20 × 14	Yes	A	Nil	ACT (4 BEP)	Seminoma	Seminoma	NED, 42
10	Modi, 2015 (10)	42	AUR	11 x 7	Yes	A	Nil	ACT (4BEP)	Seminoma right side	GCNIS left side	NED, 6
11	Williams, 1994 (11)	17	Undescended testis	NA	Yes	A	Nil	Nil	Microinvasive GCT	GCNIS	NA
12	Present case	30	Abdominal mass	10 × 10	Yes	A	nil	Adj ACT (1 Carboplatin)	Seminoma	Gonadoblastoma	NED, 12

Comments: y – year; LDT – longest tumor dimension; NACT – neoadjuvant chemotherapy; ADT – adjuvant chemotherapy; PMDS – persistent mullerian duct syndrome, RP – retroperitoneal; C/L – contralateral; PE – pleural effusion; BEP – bleomycin, etoposide, platinum regimen; NED – no evidence of disease; Adj – adjuvant; NSGCT – non seminomatous germ cell tumor; MGCT – mixed germ cell tumor NA – not available; OU – obstructive uropathy; AUR – acute urinary retention; Cisp/Cylophos – cisplatin/cyclophosphamide; GCNIS – germ cell neoplasia in situ; RT – radiotherapy.

phenotypically male. Management decisions in PMDS should therefore be individualized, taking into account patient age, fertility potential, anatomical location of the testes, feasibility of surveillance, and patient preference. While bilateral orchiectomy offers definitive oncologic risk reduction, it may not be appropriate in all clinical contexts.

4.1. Risk of malignancy in PMDS

Patients with PMDS have a markedly elevated risk of developing testicular tumors, linked to the condition's frequent association with undescended testes.[1] Earlier estimates suggested that the incidence of testicular cancer in PMDS was around 18%, comparable to the general risk seen in cryptorchidism.[12] However, a review by Picard et al. reported a higher incidence of 33% among PMDS patients aged 18 years and older, who experienced unilateral or bilateral malignant testicular degeneration.[1] The presence of undescended testes further amplifies the risk of testicular tumors in these individuals. Reports from Melman et al. and Manassero et al. indicate that early orchidopexy does not necessarily prevent testicular degeneration, implying that factors other than testicular malposition may contribute to malignant change in PMDS.[13,14] Seminoma is the most common type of testicular malignancy in PMDS; however, mixed germ cell tumors, choriocarcinomas, gonadoblastomas, and yolk sac tumors have also been documented.[12] Additionally, young patients may develop germ cell neoplasia in situ.[11]

Alternative strategies such as testis-sparing approaches with intraoperative biopsy and structured surveillance may be considered in selected patients, particularly when fertility preservation is a priority.

However, in PMDS, these approaches may be limited by the intra-abdominal location of the testes, technical challenges in access, and the difficulty of reliable long-term surveillance. In addition, many patients present with non-functional, atrophic gonads and limited reproductive potential, further reducing the benefit of organ preservation.

4.2. Malignancies of Müllerian derivatives

Malignant degeneration of Müllerian derivatives is relatively rare. Gagliardi's literature review over the past 50 years identified 12 cases of malignancy among 300 reported cases of PMDS, involving individuals aged 4 to 68 years.[15] Reported malignancies include uterine adenocarcinoma, papillary cystadenocarcinoma, clear cell adenocarcinoma, adenosarcoma, endocervical adenocarcinoma, and squamous cell carcinoma. Farikullah et al. estimated that 3.1% to 8.4% of males with PMDS develop Müllerian malignancies.[16] Therefore, removal of Müllerian structures during surgical procedures is recommended whenever possible because of their intrinsic potential for malignant transformation, regardless of hormonal influence. Stripping the mucosal lining of Müllerian structures has also been shown to reduce the risk of malignant uterine degeneration.[17]

4.3. Risk of malignancy in the contralateral testis

The risk of malignancy in the contralateral testis in individuals with PMDS is significant, especially in cases of bilateral cryptorchidism. In our review, contralateral malignancy or GCNIS was identified in approximately 17% of reported PMDS patients with testicular cancer. However, this proportion is derived from a highly selected cohort of published cases and is likely influenced by referral and publication bias. It should therefore be interpreted cautiously and regarded as hypothesis-generating rather than a precise estimate of absolute risk. These findings highlight the possibility of occult premalignant or malignant pathology in the contralateral testis of PMDS patients with bilateral cryptorchidism rather than providing a precise estimate of absolute risk.

The abnormal intra-abdominal position of the testes results in prolonged exposure to a suboptimal environment, leading to atrophic and hyalinized seminiferous tubules. This altered environment, along with genetic factors, predisposes germ cells to neoplastic transformation, resulting in GCNIS and invasive germ cell tumor within the atrophic tubules.

GCNIS is the most common precursor to testicular malignancy. Pourkeramati et al. found that 23% of infertile men with intra-abdominal testes had GCNIS based on orchiectomy specimen examinations.[18] Additionally, Dieckmann et al. reported that approximately 50% of patients with GCNIS develop invasive testicular germ cell tumors within five years.[19] GCNIS is more prevalent in individuals with atrophic germinal epithelium and has a high incidence in those with undescended testes.[20] Routine hematoxylin and eosin (H&E) staining can miss GCNIS, necessitating the use of immunohistochemical staining for PLAP and OCT 3/4, which are frequently expressed in testicular malignancy.[21] Therefore, meticulous grossing and adjunctive immunohistochemistry are essential for accurate assessment, particularly in high-risk settings such as intra-abdominal testes.

Given that many patients with PMDS have infertility, cryptorchidism, intra-abdominal testes, and atrophic germinal tubules, GCNIS may occur more frequently in this population, although the true incidence remains uncertain. The abnormal intra-abdominal position of the testes appears to be associated with an adverse microenvironment that may contribute to malignant transformation of germ cells. If unaddressed, this condition can progress to malignancy. In our case, we identified GCNIS in both testes in the presence of atrophic and hyalinized germinal tubules.

Based on the available published reports, contralateral malignancy or GCNIS was identified in approximately 17% of reported PMDS patients with testicular cancer; however, this figure reflects a highly selected population and may be influenced by referral and publication bias. It is possible that careful and extensive grossing, along with immunohistochemistry, may identify additional occult lesions in selected cases. Given the atrophic and non-functioning nature of intra-abdominal testes in many PMDS patients, their potential for malignant transformation, and the challenges associated with reliable surveillance, bilateral orchiectomy, including contralateral orchiectomy in selected patients presenting with primary testicular malignancy, may be considered, particularly in adults with bilateral cryptorchidism and no reproductive potential. However, it must be acknowledged that the available evidence is derived predominantly from isolated case reports and small case series, which precludes precise risk stratification and limits the generalizability of management recommendations.

Accordingly, the observed proportion should be regarded as hypothesis-generating and underscores the need for systematic data collection through multicenter registries rather than serving as a definitive risk stratification metric. Given the rarity of PMDS and the predominance of case-based evidence, larger col-

laborative efforts, including multicenter registries and pooled analyses, are needed to better define the true risk of gonadal and contralateral malignancy. Such data will be essential to enable more reliable risk stratification and inform evidence-based, standardized management strategies in PMDS.

5. Conclusions

PMDS is associated with an increased risk of gonadal malignancy, particularly in the setting of cryptorchidism. This case highlights the possibility of occult contralateral pathology even in macroscopically normal testes, although this observation should be interpreted with caution. In selected patients – particularly those with bilateral intra-abdominal, non-functional testes and limited feasibility for surveillance 0150 bilateral orchiectomy may represent a reasonable risk-reduction strategy. However, management should be individualized, taking into account patient age, fertility potential, anatomical considerations, and patient preference. Given the rarity of PMDS and the predominance of case-based evidence, multicenter registries and pooled analyses are required to better define malignancy risk and guide standardized management.

CONFLICT OF INTEREST

None declared.

FUNDING

None declared.

ETHICS

The study has been approved by Bioethical Committee (IEC/2024/188).

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