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Original contribution

Appendageal tumors and tumor-like lesions of the testis and paratestis: a 32-year experience at a single institution☆,☆☆

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Summary The testicular hilum and paratestis contain several embryologically diverse anatomic structures, including the spermatic cord, tunica vaginalis, epididymis, rete testis, and several other embryonic remnants. Several benign and malignant lesions arise from these morphologically distinct structures, and owing to their proximity, it is challenging to classify and subsequently stage these tumors. Herein, we conducted a retrospective review of the paratesticular appendageal and rete testis tumors and tumor-like lesions diagnosed at our department from 1985 to 2016. Soft-tissue lesions/tumors were excluded. A total of 146 paratesticular appendageal and rete testis tumors and tumor-like lesions were identified. Most were benign (n = 107; 73%). Adenomatoid tumor (26%) was the most common benign tumor, followed by different types of cysts (19%), mesothelial hyperplasia (18%), serous cystadenoma (5.5%), and rete testis adenoma (4%). Malignant lesions comprised 23% of the cases, with mesothelioma the most common (15%), followed by adenocarcinoma of the rete testis (4%), serous cystadenocarcinoma (2%), and papillary and clear cell adenocarcinoma of the epididymis (2%). Finally, serous borderline tumors and melanotic neuroectodermal tumor (retinal anlage tumors) comprised the remaining 4% of cases. In conclusion, a wide range of benign and malignant lesions can arise from the paratesticular region. Awareness of these lesions and their histologic spectrum is crucial to avoid diagnostic pitfalls and to allow pathologists to establish a correct diagnosis and subsequent treatment plan.

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1. Introduction

The paratesticular region and testicular hilum contain different anatomic structures that are embryologically diverse. These include the spermatic cord, tunica vaginalis, epididymis, rete testis, and several embryonic remnants, including the appendix testis (the hydatid of Morgagni), appendix epididymis, paradidymis, and vasa aberrantia. Tumors of these structures are rare and consequently have been reported mostly as case reports. Their rarity creates a significant challenge, especially among general pathologists who seldom encounter these lesions. Both benign and malignant lesions may occur, and owing to their proximity, it is often challenging to classify and stage these tumors.

A large proportion of the tumors arising in the paratesticular region are of soft-tissue origin, which tremendously expands the differential diagnosis. Another potential diagnostic difficulty is to differentiate primary appendageal and rete testis tumors from metastases from other organs, and in many instances, the diagnosis of a primary malignant lesion in this region becomes one of the diagnoses of exclusion. To expand on the limited published data and provide a basis for the differential diagnosis and management decisions, we reviewed our experience with lesions in this region and herein report the largest survey from a single tertiary care institution.

2. Materials and methods

After institutional review board approval, the pathology database at the Indiana University Health System was searched. Available archival materials of all benign and malignant paratesticular appendageal and rete testis tumors and tumor-like lesions, including incidentally diagnosed ones, from January 1, 1985, to December 31, 2016, were retrieved. The cases included resections performed at our institution and referral cases. Available clinical records of the patients were reviewed, and data were retrieved, including demographics, clinical presentation, pathology reports, and follow-up. All available original hematoxylin and eosin slides and immunohistochemical staining results were reviewed by two pathologists (K.I.A.-O. and M.T.I.). We defined an incidental finding as one separate from the primary concern for which resection or orchiectomy was performed. Detailed morphologic descriptions, immunohistochemical results, and molecular characteristics are not presented because of brevity, focus, and scope of this report.

3. Results

A total of 146 appendageal and rete testis tumors and tumor-like lesions were identified (Table 1). Benign lesions constituted 73% (107 of 146) of the cases with patients' ages ranging from 3 months to 76 years (Table 2).

Table 1 A summary of the testicular and paratesticular appendageal tumors and tumor-like lesions (in order of frequency).

Benign
Adenomatoid tumor
Various types of cysts
- Epididymal cyst
- Tunica albuginea cyst
- Müllerian cyst
- Epidermoid cyst
- Rete testis cyst
Reactive mesothelial hyperplasia
Serous cystadenoma
Rete testis adenoma (sertoliform cystadenoma/ cystadenofibroma)
Borderline tumors
Serous borderline tumor
Melanotic neuroectodermal tumor (retinal anlage tumor)
Malignant tumors
Malignant mesothelioma
Rete testis adenocarcinoma
Serous cystadenocarcinoma of the tunica albuginea
Papillary and clear cell adenocarcinoma of the epididymis

Adenomatoid tumor (AT) was the most common benign tumor (38 cases; 35% of the benign lesions; 26% of total), occurring at a mean patient age of 44 years (range, 15–79 years) and with an average size of 1.3 cm (range, 0.5–3 cm). Most were left sided (24 cases), and most presented as mass lesions, although 8 were incidental. Macroscopically, most ATs appeared well circumscribed, with occasional exceptions, and showed firm, homogenous, white to tan yellow cut surfaces. Twenty-three tumors were in paratesticular locations, including the tunica albuginea, rete testis, hilar soft tissue, and epididymis; 10 partially involved the testicular parenchyma, and 5 were completely intratesticular, without evidence of extratesticular extension. Microscopically, 10 tumors showed a variably thickened capsule-like condensation of fibrous tissue around the tumors. The remaining tumors displayed less of this phenomenon. The tumor displayed solid, tubular, cord-like, nested, microcystic, macrocystic, trabecular, and focal spindle cell patterns. The cells had a moderate amount of eosinophilic to vacuolated cytoplasm with round to oval nuclei and indistinct nucleoli. The characteristic feature of thin bridging stroma within tubules was seen in more than half of the cases (Fig. 1A and B). Four tumors appeared to infiltrate into the surrounding tissue, including intratesticular growth (Fig. 1C). Tumor infarction, which caused exuberant fibroblastic proliferation and inflammatory reactions around the tumor, obscured the morphologic features and caused a diagnostic challenge in 7 consultation cases (Fig. 1D) [1].

Several different types of cysts (28 cases; 26% of the benign lesions; 19% of total) were the next most frequent benign lesions. Benign epididymal cysts (BECs) were the

Table 2 A summary of the benign testicular and paratesticular appendageal tumors and tumor-like lesions (73%; n = 107).

Type	No. of cases (% of benign; % of total)	Age range (mean), years	No. of incidental cases (%)	Size range (mean, cm)	Laterality (left/right)
Adenomatoid tumor	38 cases (35; 26)	15–79 (44)	8 (21%)	0.5–3 cm (1.3 cm)	24/13[a]
Various types of cysts	28 cases (26; 19)	7 m–77 (41)	15 (54%)		
- Benign epididymal cyst	21 cases	26–75 (41)	13 (62%)	0.3–5.4 (1.5 cm)	9/11[a]
- Benign cyst of tunica albuginea	4 cases	14–77 (40)	2 (50%)	0.2 cm[b]	1/3
- Müllerian cyst	2 cases	7 m–16	0 (0%)	N/A	0/1
- Rete testis cyst	1 case	77	0 (0%)	5.5 cm (–)	0/1
Reactive mesothelial hyperplasia	26 cases (25; 18)	3 m–75 (35)	24 (92%).	N/A	10/16
Serous cystadenoma	8 cases (8; 6)	35–73 (49)	0 (0%)	3.4–3.6 cm (3.5)	4/3[a]
Rete testis adenoma	6 cases (6; 4)	21–81 (33)	0 (0%)	1.1 cm[b]	0/4

N/A: not applicable, no grossly detected lesion.
[a] Laterality is unknown for one case.
[b] Size is available for one case.

most common (21 cases), occurring at a mean age of 41 years (range, 26–75 years) and averaging 1.5 cm (range, 0.3–5.4 cm) in diameter. BECs were incidentally found in 13 cases; of which, 10 patients had germ cell tumors and 3 had sex cord-stromal tumors. No side predilection was found. Macroscopically, the cysts were monoloculated and multiloculated. Microscopically, they were lined by flat to cuboidal and variably columnar ciliated cells with minimal to moderate amounts of eosinophilic cytoplasm and bland nuclei. The stroma was collagenous and lacked the features of the ovarian-type stroma. Next in sequencing were 4 benign cysts of the tunica albuginea (3 right and 1 left). Microscopically, three were unilocular, and one was multilocular. All were lined by flat mesothelial cells with minimal cytoplasm and dark nuclei. The stroma was densely collagenous. Two Müllerian cysts in a 7-month- and 16-year-old patient occurred on the right side. These cysts were lined by columnar ciliated epithelial cells without any specific stroma and thus were not classified as endometriotic cysts. One rete testis cyst that occurred in conjunction with a benign cyst of the tunica albuginea measured 5.5 cm in the greatest dimension. It showed a lining of attenuated rete epithelium.

Reactive mesothelial hyperplasia (RMH) occurred in 26 cases (25% of the benign lesions; 18% of total). All were incidental to benign (n = 15) or malignant (n = 11) lesions involving the testicular or paratesticular tissue. RMH was associated with germ cell tumors in 10 cases, whereas a single case was associated with embryonal rhabdomyosarcoma of the paratestis. Microscopically, RMH showed mesothelial cell proliferation on the surface of the tunica albuginea. None produced evidence of mass or showed infiltrative growth, and all had bland nuclear morphology, in conjunction with the polygonal profile and eosinophilic cytoplasm.

Other benign lesions included 8 serous cystadenomas (8% of the benign lesions; 6% of total), 5 in the paratestis (2 were papillary cystadenomas of epididymal origin and the remainder were of tunica vaginalis, efferent ductule, and or tunica albuginea origin). Two arose within the testis, and the exact origin of one is unclear. The average age of patients' was 49 years (range, 35–73 years). No laterality predilection was found. Microscopically, the cysts were composed of cuboidal to columnar ciliated epithelial cells with abundant pale cytoplasm and bland nuclei. The surrounding stroma was similar to the ovarian counterpart in 4 cases; however, the remaining 4 had collagenous stroma. Rete testis adenoma was seen in 6 cases (6% of the benign lesions; 4% of total), occurring at a mean patient age of 33 years (range, 21–81 years). All were right sided. Of the 6 cases, 3 were of the sertoliform cystadenoma type (Fig. 1E). They were centered in the rete testis and showed Sertoli-like cells with overlapping features with Sertoli cell tumors [2]. Two cases were cystadenofibromas and showed variable sized tubules and dilated cystic spaces embedded in a dense stroma. The tubules/cystic spaces were lined by flattened, columnar, or ciliated epithelium (Fig. 1F). The final case showed an epithelial component composed of noncystic glands compressed by abundant fibrocollagenous stroma, therefore designated as rete testis adenofibroma.

The remaining 27% (40 of 146) were malignant (n = 34) or borderline (n = 6) tumors (Tables 3 and 4). The patients' ages ranged from 13 to 47 years and 33–76 years, respectively. Malignant mesothelioma was the most common among this group (n = 22; 65% of the malignant lesions; 15% of total). It occurred mainly in older men, occurring at a mean age of 62 years (range, 42–76 years) and with mean size of 6.7 cm (range, 1.5–13 cm). Seven patients were diagnosed incidentally while being treated for recurrent hydrocele. Microscopically, two-thirds had pure epithelioid morphology with papillary, tubulopapillary, solid, or tubular architecture. The cells had moderate amounts of pale to eosinophilic cytoplasm with moderate to marked nuclear pleomorphism (Fig. 2A–D). The remaining one-third showed biphasic morphology with epithelioid and sarcomatoid (spindle cell) components. No purely sarcomatoid cases were identified.

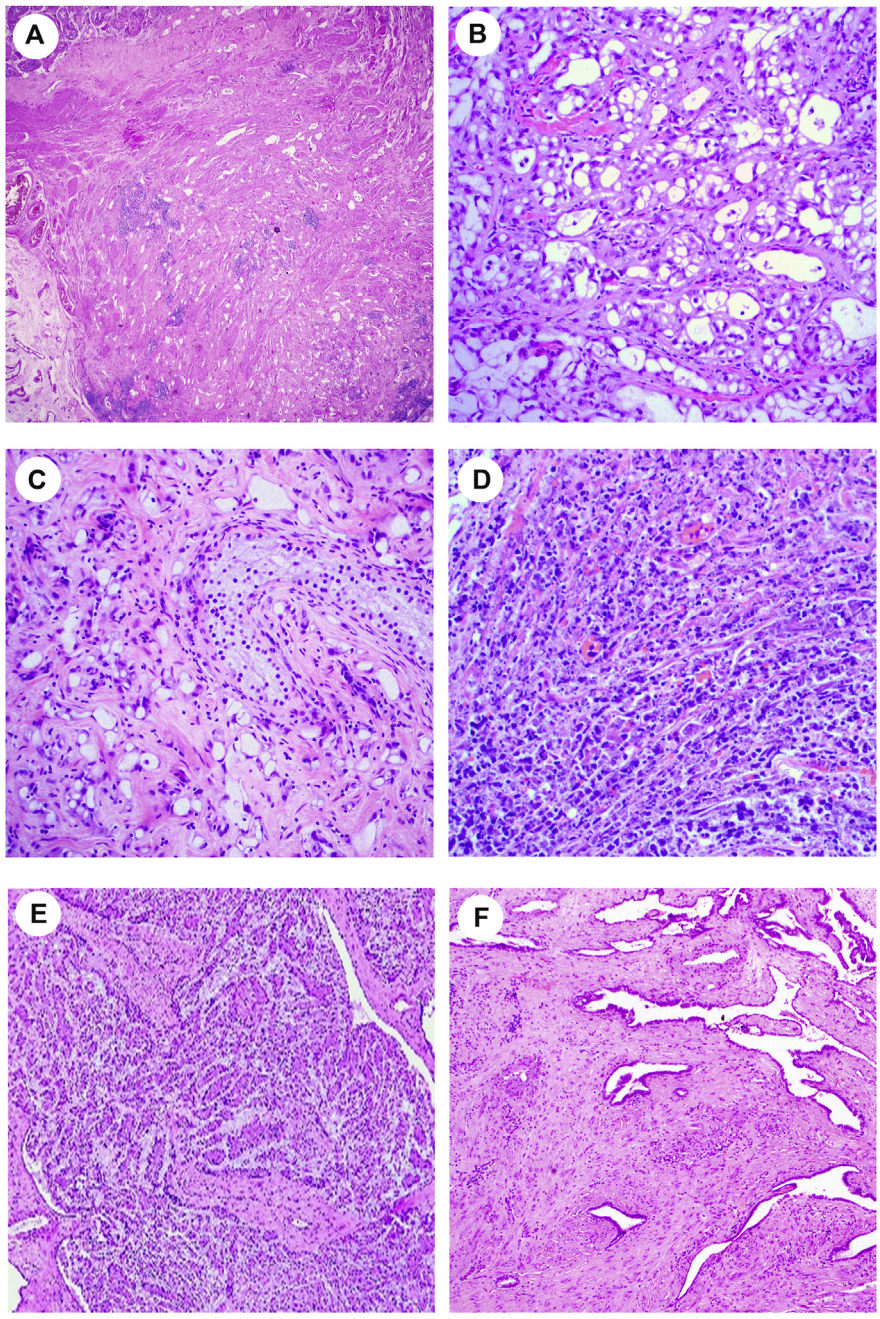

Fig. 1 Adenomatoid tumor displaying nodular proliferation (A) of tubular and microcystic growth patterns with prominent stroma (B). The cells have a minimal amount of eosinophilic cytoplasm with round to oval nuclei and indistinct nucleoli. A characteristic thin bridging stroma within tubules is seen (panel B). Intratesticular growth (C) and necrosis (D) are seen. Sertoliform cystadenoma of the rete testis showing tumor involving the cystically dilated rete testis composed of well-formed tubules, lined by columnar cells with eosinophilic cytoplasm (E). Rete testis adenofibroma showing variable sized glands with prominent stromal proliferation (F).

Six cases of adenocarcinoma of the rete testis (17% of the malignant lesions; 4% of total) occurred in men, occurring at a mean age of 64 years (range, 56−76 years). They presented as palpable masses. Grossly, the tumors had well-defined solid and cystic areas with tan yellow cut surfaces and were centered at the testicular hilum. Tumor size was available for three cases and measured 3.3 cm, 2 cm, and 9 cm. Microscopically, they displayed mostly glandular morphology that varied from slit-like lumens to well-formed glandular and tubular structures. Papillary and solid patterns were also formed. Tumor nests with eosinophilic to clear cells were seen in 2 cases, in addition to other less represented but unique patterns. Three of the tumors displayed geographic necrosis. The stroma ranged from fibrotic to partially hyalinized in all cases [3].

Table 3 A summary of the malignant testicular and paratesticular appendageal tumors and tumor-like lesions (23%; n = 34).

Type	Cases, no. (% of malignant; % of total)	Age range (mean), years	No. of incidental cases (%)	Size range (mean, cm)	Laterality (left/right)
Malignant mesothelioma	22 cases (65; 15)	42–76 (64)	0 (0%)	1.5–13 (6.7)	9/11
Adenocarcinoma of the rete testis	6 cases (17; 4)	56–76 (66)	1 (17%)	2-9 (4.8)[b]	2/4
Serous cystadenocarcinoma	3 cases (9; 2)	15–64 (37)	1 (33%)	N/A	2/1
Adenocarcinoma of the epididymis	3 cases (9; 2)	42–70 (67)	0 (0%)	9[a]	2/1

N/A: size is not available.
[a] Size is available for one case.
[b] Size is available for 3 cases.

Table 4 A summary of the borderline testicular appendageal tumors and tumor-like lesions (4%; n = 6).

Type	Cases, no. (% of borderline; % of total)	Age range (mean), years	No. of incidental cases (%)	Size range (mean, cm)	Laterality (left/right)
Serous borderline tumor	5 cases (83; 3.4)	42–76 (64)	0 (0%)	N/A	5/0
Melanotic neuroectodermal tumor (retinal anlage tumor)	1 case (17; 0.6)	56–76 (66)	1 (17%)	2–9	0/1

N/A: sizes are not available.

Our series also identified 3 cases of cystadenocarcinoma of the tunica albuginea (9% of the malignant lesions; 2% of total), occurring at a mean age of 37 years (range, 15–64 years). Two presented as masses, and the last was incidental in a hydrocelectomy specimen. All displayed low-grade papillary architecture similar to that of the female genital tract. Two of the tumors showed abundant psammoma bodies, whereas one had clear cell features (Fig. 3A and B).

There were 3 cases of papillary and clear cell adenocarcinoma of the epididymis (9% of the malignant lesions; 2% of total) that presented at a mean age of 67 years (range, 42–70 years). The former showed papillary

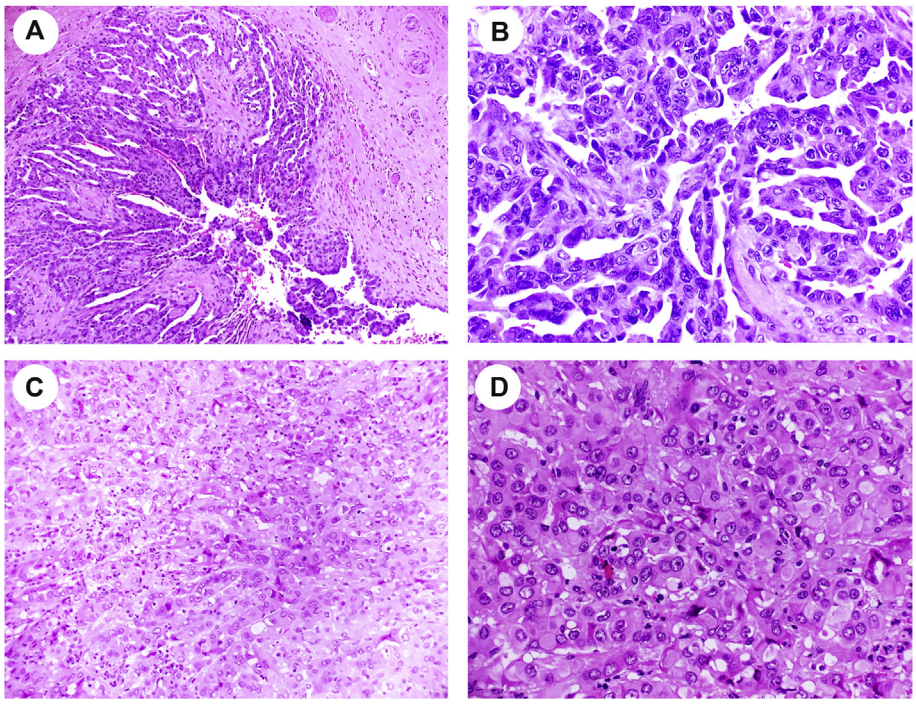

Fig. 2 Malignant mesothelioma displaying papillary (A and B) and solid (C and D) growth patterns, with epithelioid cells (C) and a variable degree of nuclear pleomorphism (D).

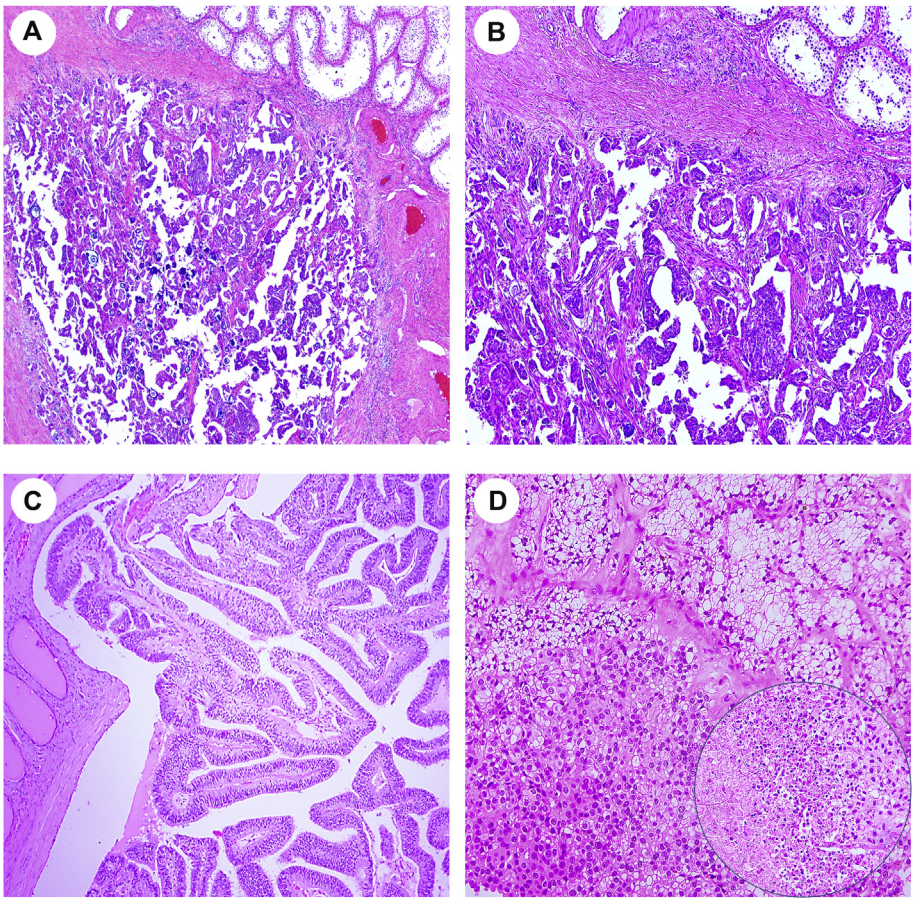

Fig. 3 A and B, Papillary serous carcinoma with psammomatous calcifications. C, Papillary adenocarcinoma of the epididymis showing papillary fronds lined by oval to columnar cells. D, Clear cell adenocarcinoma of the epididymis arising in a papillary cystadenoma and showing solid growth, with increased nuclear pleomorphism, and tumor necrosis (inset).

structures lined by oval to columnar cells with vacuolated cytoplasm, with occasional solid growth (Fig. 3C). The latter arose in a papillary cystadenoma and showed solid growth, with increased cellularity, nuclear pleomorphism, and tumor necrosis (Fig. 3D).

Of the low malignant potential tumors, 5 were serous borderline tumors (3.4% of total) and 1 (0.6% of total) was a melanotic neuroectodermal tumor (retinal anlage tumor), representing all the remaining 4% of the total cases. The former presented at a mean age of 39 years (range, 13−47 years). All were left sided. Most presented as masses (n = 4), but 1 was incidentally diagnosed in a hydrocelectomy specimen. Microscopically, the tumors were lined by stratified, ciliated serous-type epithelium and displayed papillary architecture within cystic spaces in a hierarchical branching pattern. Detached clusters of epithelium occurred along the luminal aspects of the papillae (Fig. 4A and B). No invasion occurred in the underlying collagenous stroma. One case showed psammomatous calcifications. These cases were identical to their ovarian counterparts. The melanotic neuroectodermal tumor (retinal anlage tumors) occurred in a 1-year-old patient. Microscopically, it displayed two types of tumor cells, small neuroblast-like cells and large cells surrounding the neuroblast-like cells. The large cells contained variable amounts of melanin pigment, occasionally obscuring the bland-appearing nuclei (Fig. 4C and D). No tumor capsule was present, and the tumor cells infiltrated into a sclerotic stroma.

4. Discussion

The diverse embryonic origins and histologic characteristics of the scrotal structures give rise to a wide morphologic spectrum of epithelial and nonepithelial lesions [4]. Most intrascrotal tumors are germ cell neoplasms. Those of the testicular ductular system and paratestis, however, are not uncommon [5]. They account for 7−10% of all intrascrotal tumors, and the proportion of malignancy is estimated to be 30% [6,7]. The relative infrequency and diverse appearance of these tumors can cause significant diagnostic challenges, especially for general pathologists [4,8]. We, therefore, reviewed the testicular surgical

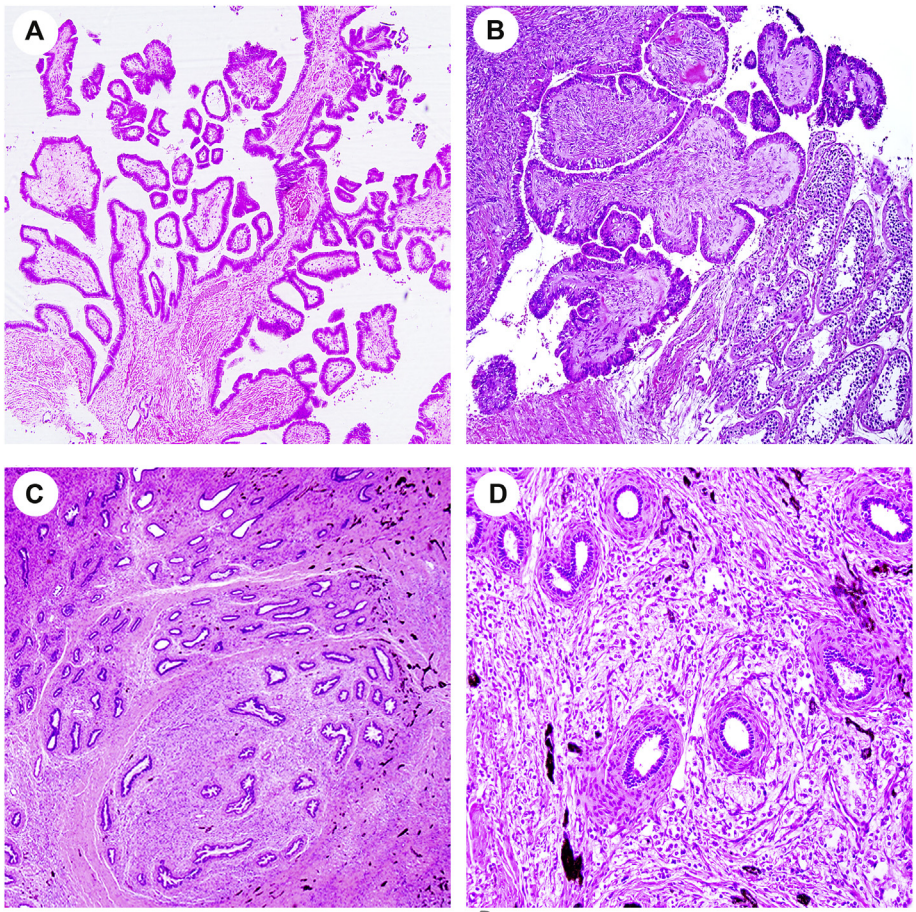

Fig. 4 Borderline tumors. A and B, Serous borderline tumor showing hierarchical branching of papillae. C and D, Melanotic neuroectodermal tumor (retinal anlage tumor) showing a dual population of neuroblastic-like cells and surrounding large melanin-containing cells. The tumor infiltrated between epididymal tubules.

specimens received by our department over the course of 32 years and found 146 cases of benign and malignant rete testis and appendageal tumors and tumor-like lesions.

In our series, benign lesions accounted for 73% of all tumors and occurred mostly between the third and fifth decades of life. AT was the single most common benign neoplasm (35% of the benign lesions) and involved the epididymis and other paratesticular structures, although 5 were exclusively intratesticular. Paratesticular tumor constituted the second most common tumor, ranking below only lipoma and occurring between the ages of 15 and 79 years (mean, 44 years), although a younger age at presentation has been reported [9,10[9,10,11]. It may present either as a painless scrotal mass or at autopsy [8]. Eight cases of AT (21%) were diagnosed incidentally in orchiectomy specimens, thus highlighting its importance in the differential diagnosis of testicular lesions. ATs may morphologically be confused with epithelial and/or spindle cell tumors, although the diagnosis is relatively straightforward. Our series included several consult cases received owing to unusual morphologic features, especially necrosis, and increased cellularity, which caused diagnostic challenges. The presence of the thread-like bridging strands, a distinctive feature of ATs, aided in the diagnosis, and the feature was present in more than half of our cases [12]. Reactivity for mesothelial markers is helpful, as is positivity for L1CAM, in difficult cases [13,14].

Next in frequency were various cysts. There has not been any reported data regarding their frequency; however, they accounted for 26% (28 cases) of the benign lesions in our experience. BECs were the most common (21 cases). Surgical intervention for BEC was performed for 8 cases, and 13 were incidental. Benign mesothelial cysts of the tunica albuginea were the next in frequency (n = 4), followed by two cysts of Müllerian remnant origin, one epidermoid cyst, and one rete testis cyst. The extratesticular epidermoid cyst is very rare, with only a few cases being reported [15–17].

RMH is a non-neoplastic lesion that presents in association with a variety of benign and malignant lesions of the testis and paratesticular tissue [8,18,19]. In our series, RMH occurred in 18% of the benign lesions (26 cases), 15 cases of which were secondary to benign lesions, including hydrocele, hematocele, and inflammatory processes, whereas the remaining 11 were secondary to malignant

lesions; of which, germ cell tumors were the most common. Exuberant mesothelial proliferation may be mistaken for mesothelioma. The key to the distinction is the absence of a mass lesion and invasive growth, with a pattern of extension into the tunica vaginalis stroma or tunica albuginea along a linear front [20].

Serous cystadenoma is a benign tumor that can arise from different paratesticular structures, including the epididymis and tunica vaginalis. Papillary serous cystadenoma of the epididymis usually occurs in young adults, but the age range is broad (16–76 years). It is usually unilateral; however, bilateral cases are strongly associated with von Hippel-Lindau (VHL) disease [21–23]. We had 2 cases of papillary serous cystadenoma of the epididymis. Both were unilateral and occurred in a 35- and 52-year-old patient. None had any stigmata of VHL disease. We also had 6 cases of serous cystadenomas, which occurred at a mean age of 49 years (range, 35–73 years). Two were intratesticular, 1 was within the tunica albuginea, 1 was within the efferent ductules, 1 was within the tunica vaginalis, and the exact origin of one was unclear.

Serous cystadenomas are slightly more common than mucinous cystadenomas in general; however, no mucinous cystadenoma was identified at our institution.

Rete testis adenomas are benign tumors that can have a wide spectrum of growth patterns [4]. Cystadenoma of the rete testis, including sertoliform, adenofibroma, and cystadenofibroma, represents an uncommon distinctive benign tumor and accounted for 6% (6 cases) of the benign lesions in our series, with an age ranged from 21 to 81 years in our series. The overlapping morphology and immunohistochemical profile between Sertoli cell tumors and sertoliform cystadenoma make the differentiation between these 2 entities challenging [2].

The estimated incidence of paratesticular malignant lesions is 30%, which approximately compares with what we found [7]. The majority of malignancies occurred between the fourth to sixth decades of life. Among them, malignant mesothelioma was the most common. It represented 15% of all paratesticular tumors and 65% of the malignant ones in our series. Our cases fell within the reported range (42–76 years), although younger patients have been reported [4]. In most of the reported cases, hydrocele was the most common presenting feature and accounted for 32% of our cases [24]. Malignant mesothelioma is an aggressive malignant tumor, with high recurrence (52%) and a mortality (40%) rate [24]. We found both purely epithelioid and biphasic tumors, with a ratio of 3:1. None were purely sarcomatoid. Malignant mesothelioma has challenging morphologic features that overlap to a degree with those of sex cord-stromal tumors, appendageal malignant tumors, and metastatic tumors if purely tubulopapillary [25]. Multifocal involvement of the tunica vaginalis, lack of immunoreactivity for steroidogenic factor 1 and PAX-8 (usually), the absence of epithelial cell adhesion molecule (EpCAM) expression, and the clinical history are helpful in the resolution of these differential considerations.

Adenocarcinoma of the rete testis followed malignant mesothelioma in terms of frequency. It accounted for 17% (n = 6) of malignant lesions in our series. We recently reported our detailed experience with these tumors, and therefore, they will not be further discussed here [3]. Next in frequency was cystadenocarcinoma of the tunica albuginea (n = 3; 9% of the malignant lesions). Prior reports document them in patients aged 16–42 years (mean, 31 years) who presented with testicular fullness, mass, or hydrocele [4,26,27]. We found a wider age range (15–64 years; mean, 37 years), slightly older than that reported in the literature. This rare malignant tumor shows a high rate of recurrence and metastatic potential [4,26]. The diagnosis of Müllerian-type tumors required careful evaluation and appropriate immunohistochemical support. Many of them develop from the appendix testis, which may be the gross epicenter, or from the surface of the tunica vaginalis through the process of Müllerian metaplasia. Common nuclear positivity for sex hormone receptors and PAX-8 is the supportive finding.

Adenocarcinoma of the epididymis is another extremely rare tumor, with less than 20 cases reported in the literature [5,28,29]. Two-thirds of the reported tumors occurred at an age older than 50 years (range, 27–81 years). The vast majority of tumors are not associated with VHL syndrome, contrasting with papillary cystadenoma of the epididymis [28]. Histologically, tumors show variable growth patterns, including tubular, papillary, tubulopapillary, cystopapillary, or solid patterns. The cytoplasm is clear, amphophilic, or eosinophilic [29]. Despite the overlapping features with papillary cystadenoma of the epididymis, the presence of increased mitotic figures, nuclear pleomorphism, necrosis, and/or invasive growth should direct the diagnosis toward adenocarcinoma. Tumors are frequently positive for EMA, pan cytokeratin, and CK7 immunostaining and are negative for prostate markers, S100, vimentin, alpha-fetoprotein, and calretinin [29]. In our series, it accounted for 9% (n = 3) of the malignant cases and presented in the seventh and eighth decades. Two cases were of pure papillary architecture, whereas one arose from clear cell papillary cystadenoma of the epididymis.

The rare borderline tumors represented the remaining 4% cases. We identified 5 serous borderline tumors in patients aged 13–47 years (mean, 39 years), which contrast with the age range of 14–77 years (mean, 56 years) in the literature [30]. All our cases were left sided, and one was diagnosed incidentally in a hydrocelectomy specimen. These tumors may grow to a large size, and thorough sampling is required to evaluate for invasion. Stromal invasion of $\leq$5 mm does not have prognostic significant, but greater invasion should be recognized as *low-grade serous carcinoma* as minimal invasion in stroma is permitted (up to 5 mm) in these lesions [31].

Melanotic neuroectodermal tumor of infancy is a rare, rapidly expansile tumor of neural crest origin, with a high recurrence rate [32]. Most of the tumors occur in the head

and neck region of infants, with <5% in the paratestis [33]. In our series, there was a case of melanotic neuroectodermal tumor in a one-year-old infant who was treated by excisional biopsy. The experience with this tumor in the epididymis and/or paratestis is limited. There is a low percentage of malignant behavior (14%) with metastasis to adjacent lymph nodes, and therefore, here, we considered it as a borderline lesion. In the literature, the three reported patients with metastasis were treated with lymphadenectomy and were alive after 28–48 months of follow-up [34].

In conclusion, the majority of paratesticular appendageal and rete testis tumors are benign. AT represents the majority of cases, followed by mesothelial hyperplasia and various types of cysts. On the other hand, malignant mesothelioma is the most common malignant tumor, followed by adenocarcinoma of the rete testis. Awareness of these seldom encountered entities by surgical pathologists is warranted and may yield substantial benefits for patients in establishing an accurate diagnosis and appropriate clinical management.

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