Conservative Treatment of Serous Borderline Paratesticular Tumor in a Pediatric Patient

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Serous borderline tumors are rare neoplasms. Herein we report our conservative approach, whose rationale is neoplasm low-malignant potential. Tumor was removed under general anesthesia, and frozen section ruled out a germinal malignancy or a stromal tumor such as rhabdomyosarcoma. Ultrasound evaluation was initially performed every 3 months during the first 2 years, every 6 months during the next 3 years, and annually thereafter. After 8 years, the patient has not experienced any relapse, either clinical or ultrasonographical. In our opinion, conservative approach, whose final decision relies on intraoperative frozen section, represents the best option and does not jeopardize long-term oncological outcome. UROLOGY 89: 123–125, 2016. © 2016 Elsevier Inc.

Serous borderline tumor (SBT) of the testis represents a rare neoplasm; histologically, it resembles its ovarian counterpart.[1,2] Its oncological behavior is not well defined, given the paucity of literature on this topic. Case reports and small caseloads show that radical orchiectomy is the treatment most often used.[1-3] We describe herein our conservative approach and pathological characteristics of an SBT that occurred in a 14-year-old patient 8 years ago.

CASE PRESENTATION

A 14-year-old male presented to the emergency department complaining of testicular pain over the past 2 days. A careful objective examination revealed a 7-8 mm mass indissociable from the right didymus. After ultrasonographical evaluation (Fig. 1) and tumor marker panel serology, which was negative, an excisional biopsy was planned. On July 2007, the testicle was exposed, under general anesthesia, and the neoplasia, adherent to the tunica albuginea, was removed.

Macroscopical tumor aspect was polipoid; mass consistency was mucinous. The specimen was sent for a frozen section. This ruled out a germinal malignancy or a stromal tumor such as rabdomyosarcoma. Moreover, histological evaluation showed a tumor with papillae covered by neoplastic epithelium with no malignant features, thus orchiectomy was not performed. The tumor was localized at tunica albuginea level, hence, after removal, this was sewn, given the small mass diameter.

We evaluated the effectiveness and oncological safety of our conservative approach through a strict follow-up over the past 8 years. Ultrasound evaluation was initially performed every 3 months during the first 2 years, every 6 months during the next 3 years, and annually thereafter.

RESULTS

Pathological examination revealed a serous borderline papillary tumor (Fig. 2).

Immunohistochemical evaluation confirmed the preoperative diagnosis and demonstrated positivity for AE1/AE3, CK7, CA125, ER, PR, WT1, BER EP4, EMA, and PAX8 markers. Immunohistochemical examination was negative for calretinin, trombomodulin, caldesmon, CEA, D2 40, CK5/6, CDX2, CK20, CD117, CD30, alpha fetoprotein, beta hcg, desmin, and B72.3.

After 8 years, the patient has not experienced any relapse, either clinical or ultrasonographical.

COMMENT

Because literature did not produce cases of relapses or metastasis for SBT,[2,4,5] we had adopted a conservative approach, of which final decision would have been based on extemporaneous pathological examination.

SBTs are rare entities. Only four cases have been reported in literature as occurring in pediatric patients.[3,6-8] Paratesticular tumors can have malignant characteristics, especially in case of serous carcinoma or sarcoma.[5,9]

As paratesticular tumor, rhabdomyosarcoma is more frequent compared to SBT.[9] Rhabdomyosarcoma's standard

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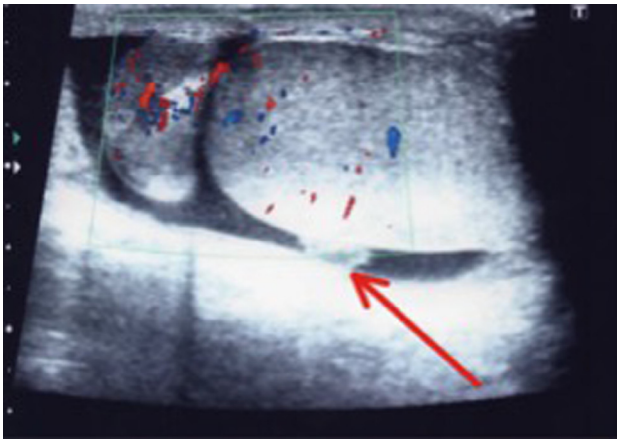

Figure 1. Ultrasonographical examination; red arrow indicates the tumor. (Color version available online.)

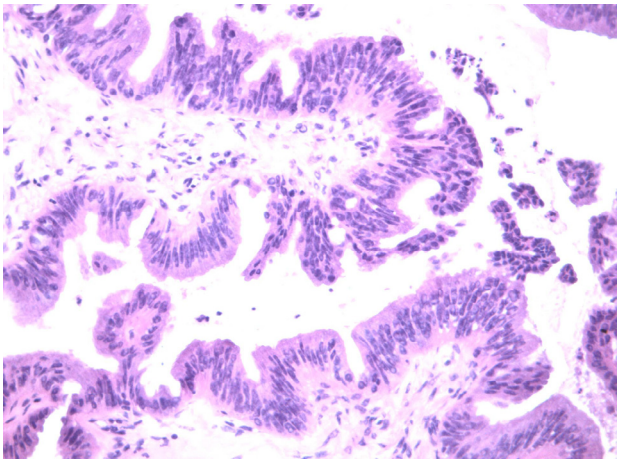

Figure 2. Papillae lined by stratified epithelial cells: epithelium with light atypia and a low mitotic activity (hematoxylin and eosin staining). (Color version available online.)

multimodal treatment relies on the local control of the primary site with radical orchiectomy, chemotherapy, and radiotherapy according to stage, histology, and age of the patient.[10] If the frozen section had shown rhabdomyosarcoma, hemiscrotectomy along with high ligation of the spermatic cord would have been performed. Because surgery alone produced approximately a 50% 2-year relapse-free survival,[11] systemic chemotherapy alone or combined chemo- and radiotherapy would have been adopted to maximize tumor control, thus resulting in increased morbidity.

Our case concerns a borderline tumor, whose malignant potential seems to be inconsistent.[1-5] Its treatment is classically reported to be total orchiectomy.[1-3] In the four previously reported pediatric cases, radical orchiectomy has been the treatment of choice in three[3,6,7] whereas one only describes tumor resection.[8]

Eight years follow-up of the present case confirms and supports the conservative approach given the low malignant SBT potential.

SBTs have been reported to occur in patients from 14 to 68 years of age, usually manifesting themselves as scrotal enlargement.[12] Their origin is still under debate. One theory affirms that ovarian-type epithelial tumors of the testis originate from Müllerian ducts remnants in paratesticular connective tissue, epididymis, and spermatic cord, whereas another states that SBT is the result of Müllerian metaplasia of the tunica vaginalis mesothelium. This latter theory is supported by frequent findings of metaplastic serous Müllerian epithelium in these tumors.[13] Hormonal influences concerning the risk of SBT developing in the paratestis are unknown; it is not clear if long-term topical testosterone may have a role.[5]

One of the key steps in the diagnosis of ovarian-type epithelial tumors is distinguishing them from clinically aggressive neoplasms such as tunica vaginalis mesothelioma, and carcinoma of the rete testis. Beyond histological features of mesothelioma, immunohistochemical evaluation of mesothelial markers such as D2-40, thrombomodulin, and calretinin can be helpful for assessing tumor nature.[14] Our pathological results are in keeping with those findings. Quite often, serous tumors show an opposite immunohistochemical pattern for those antigens and express ovarian epithelial tumor markers such as epithelial membrane antigen, CA-125, cytokeratin 7, CD15 (Leu-1), and Ber-EP4.[14] Because PAX8 expression in ovarian-type epithelial tumors is usually expressed, it represents a valuable tool when differentiating this neoplasm from malignant mesothelioma, that, on the contrary, rarely expresses it.[15]

CONCLUSION

Our experience in conservative approach to SBT has demonstrated safety over a long-term follow-up period. Because literature does not produce cases of metastasis or relapse for this kind of tumor, our conservative approach, in our opinion, represents the best option and does not jeopardize long-term oncological outcome.

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