LETTER TO THE EDITOR

Alveolar rhabdomyosarcoma of the tunica vaginalis presenting as a tender hydrocele

Rhabdomyosarcoma (RMS) is a soft tissue malignancy originating from primitive mesenchymal cells that differentiate into skeletal muscle tissue. Histologically, RMS is divided into two categories: embryonal and alveolar. Paratesticular RMS (including RMS originating from the spermatic cord, epididymis, tunica vaginalis, appendices of the testis, and epididymis) is highly aggressive and exceedingly uncommon in adult patients (the median age at diagnosis being 8 years). Less than 10% of paratesticular RMS is of the alveolar subtype, which carries an even poorer prognosis.

We report here the case of a 22-year-old male presenting with right scrotal pain and swelling, the scrotum having been rapidly enlarging over a period of 3 weeks. His condition had been treated elsewhere as epididymitis, without any response to antibiotic therapy. Physical examination detected a right hydrocele with mild tenderness. Ultrasonography displayed several confluent nodules arising from the right tunica vaginalis and anechoic fluid surrounding the right testis (Fig. 1A). Levels of serum beta-human chorionic gonadotropin and alpha-fetoprotein were normal.

Because of the abnormal ultrasonographic findings and lack of response to antibiotics, tumor was considered, and a surgical exploration using a scrotal approach was undertaken, revealing clear fluid and nodular thickening of the tunica. A part of the tunica was resected for intraoperative frozen section examination, demonstrating a malignant tumor (with a high suspicion of sarcoma). Thus, a right radical inguinal orchiectomy was performed.

Macroscopically, the tunica was diffusely thickened, with several nodules arising from the tunica (Fig. 1B). Microscopic examination revealed a small round cell tumor with an alveolar-like structure (Fig. 1C). Immuno-histochemically, the tumor cells were positive for desmin (Fig. 1D) and bcl-2, and negative for SMA, EMA, CK5/6, PCK, CD34, CR, S-100, and ALK-1. Based on these findings, a pathologic diagnosis of primary alveolar RMS of the right tunica vaginalis was made. Adjuvant chemotherapy with a vincristine/dactinomycin/cyclophosphamide (VAC) regimen was provided postoperatively. The patient has since remained disease-free for 3 years.

To the best of our knowledge, only four cases of primary sarcoma of the tunica vaginalis have been described in the English-language literature, with two being embryonal RMS [1,2], one leiomyosarcoma [3], and one malignant mesothelioma [4]. Our patient is the first case of primary alveolar RMS of the tunica. Based on these five cases, we conclude that sarcoma of the tunica most commonly presents as a symptomatic or asymptomatic hydrocele. Although there are many causes of hydrocele, sarcoma of the tunica is most highly probable on account of the rapidly enlarging hydrocele without response to antibiotics, nodules arising from the tunica, and thickening or calcification of the tunica [1-4]. However, the definitive diagnosis depends on pathologic analysis.

Due to the rarity of adult paratesticular RMS, its optimal management remains to be elucidated. However, the pediatric protocol may be followed, which includes a selective combination of surgical resection, radiotherapy (for residual disease), and chemotherapy [5]. Radical orchiectomy may act as the cornerstone for local control. Ipsilateral retroperitoneal lymph node dissection may be considered for Stages II, III, and IV disease, regardless of the preoperative radiologic findings, and adjuvant chemotherapy based on a VAC regimen is advantageous to survival in all stages. We treated our patient following the same protocol (radical orchiectomy plus adjuvant...
chemotherapy). This treatment strategy induced a stable response and, 3 years after initial diagnosis, no relapse.

References


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