Primary paratesticular neuroblastoma: Report of 2 cases and review of the literature

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Abstract

Primary paratesticular neuroblastoma is a rare malignancy in children and has only been described in 6 reported cases in the literature. We report 2 additional cases and perform a review of the literature with a focus on the management of primary paratesticular neuroblastoma.

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Prepubertal testicular and paratesticular neoplasms comprise 1–2% of all pediatric solid tumors with an incidence of 0.5–2 per 100,000 children [1]. Paratesticular tumors arise from the complex anatomic region surrounding the testis and are particularly heterogeneous due to the diverse histologic elements in this region. About 75% of paratesticular neoplasms are benign, while the remainder represent a metastatic or primary malignant tumor such as rhabdomyosarcoma [2].

Neuroblastoma is the most common extracranial malignancy in children and rarely presents as a primary paratesticular tumor. Only 6 cases have been previously described in the literature. Here, we report 2 additional cases of primary paratesticular neuroblastoma and focus on its management based on the current evidence in the literature.

1. Case #1

A healthy 6-month-old boy was incidentally found to have a right paratesticular mass during a well-child visit and was referred for a further evaluation. He had previously exhibited an appropriate growth and development without any history of intermittent scrotal swelling or scrotal trauma. On physical examination, he was found to have a firm, smooth mass superior to his right testis. His abdominal and genital examinations were otherwise unremarkable. A scrotal ultrasound demonstrated a 2 × 2 cm heterogeneous, hyperchoic mass with an increased vascularity on color Doppler images and calcifications in the right suprasternal region (Fig. 1).

A scrotal exploration was performed via a right inguinal approach. Intraoperative findings included a well-circumscribed, red mass with vascular structures originating from his spermatic cord but no involvement of his testis or epididymis. After his spermatic cord was isolated and occluded, the paratesticular mass was delivered into the operative field and excised for pathologic examination. Frozen-section analysis demonstrated a small blue cell tumor, but a definite diagnosis could not be made and was deferred to permanent-section analysis. A right radical inguinal orchiectomy was performed without any complications. Pathologic examination demonstrated a poorly differentiated, stroma-poor neuroblastoma with an intermediate mitosis-karyorrhexis index (Fig. 2). The surgical margins were involved by the tumor, but his testis and spermatic cord did not demonstrate any evidence of malignancy.

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He was referred to Pediatric Oncology and found to have no evidence of metastatic disease on further staging with a CT of his chest, abdomen, pelvis, bone scan, MIBG scan, and bone marrow aspirates and biopsies. The tumor demonstrated no amplification of N-myc and a favorable histology based on the International Neuroblastoma Pathology Classification system. He was diagnosed with International Neuroblastoma Staging System (INSS) stage 1 disease and assigned to the Children’s Oncology Group (COG) low-risk group. He has since been followed with no evidence of disease at 10 years.

3. Discussion

With the exception of rhabdomyosarcoma, a majority of primary paratesticular malignancies are exceedingly rare in childhood and are only described in case reports and small series. Only 6 cases of primary paratesticular neuroblastoma have been previously reported in the literature [3–8]. Neuroblastoma more commonly metastasizes to the testis and paratesticular region with 26 cases being reported in the literature [9]. Several authors have proposed that primary paratesticular neuroblastoma originates from an ectopic adrenal rests in the spermatic cord [4,5,7]. Others have argued that it actually represents a metastatic lesion diagnosed after the spontaneous regression of its primary tumor or a multicentric site of disease rather than a true metastasis [7,10,11].

Both of our patients presented with a paratesticular mass at 6 months of age. All of the previously reported cases occurred in patients ranging from 1 day to 3 years of age with all but 1 case occurring in the first year of life. They often presented with a discrete paratesticular mass that was clearly separate from the testis except in 2 cases [3–8]. A scrotal ultrasound was performed in both of these cases and failed to identify a paratesticular mass [6,8]. In the remaining cases and our case #1, a scrotal ultrasound demonstrated a heterogeneous paratesticular mass with the variable presence of calcifications but did not provide any additional diagnostic information to physical examination [3–5,7]. While a scrotal ultrasound can reliably distinguish between intra- and extra-testicular masses, it does not allow for a definitive characterization of masses and cannot discriminate between benign and malignant paratesticular tumors [12]. Certain benign conditions can also mimic a paratesticular tumor on a scrotal ultrasound, such as a communicating hydrocele in the case described by Akramipour et al. [8]. Other mimics include a cryptorchid testis, epididymal cyst, epididymitis, hematoma, indirect inguinal hernia, polyorchidism,
pyocele, spermatocele, splenogonadal fusion, and varicocele [12]. We believe that a scrotal ultrasound may only be useful when the scrotal anatomy is unclear or there is a concern for involvement of adjacent structures on physical examination.

Both of our patients underwent a radical inguinal orchiectomy due to equivocal findings on frozen-section analysis. Frozen-section analysis, however, is not routinely performed and was only completed in the cases previously described by Encinas et al. and Calonge et al. [3–8]. It influenced the decision to perform a simple tumorectomy in the former case but not the latter due to an involvement of the testis [4,6]. The remaining patients underwent a simple tumorectomy with the exception of the patient described by Krieger et al., who presented with obvious multifocal disease and underwent a radical orchiectomy and retroperitoneal lymph node dissection followed by the administration of adjuvant radiation and chemotherapy due to a nodal involvement [3,5,7,8]. It influenced the decision to perform a simple tumorectomy in the former case but not the latter due to an involvement of the testis [4,6]. The remaining patients underwent a simple tumorectomy with the exception of the patient described by Krieger et al., who presented with obvious multifocal disease and underwent a radical orchiectomy and retroperitoneal lymph node dissection followed by the administration of adjuvant radiation and chemotherapy due to a nodal involvement [3,5,7,8].

Given the similar clinical presentation of rhabdomyosarcoma and other malignant mesenchymal tumors, we recommend that all patients undergo an exploration through an inguinal approach with the appropriate precautions followed for a possible underlying malignancy. The paratesticular mass should initially be excised and undergo frozen-section analysis to ensure its appropriate treatment.

Our patients and those described in the previously reported cases were all diagnosed with an INSS or Evan’s stage 1 disease and assigned to the COG low-risk group when determined except for the patient described by Krieger et al. All patients have demonstrated no evidence of disease with a follow-up ranging from 8 months to 10 years [3–8].

4. Conclusion

Primary paratesticular neuroblastoma is a rare malignancy that has only been reported in children. It often presents as a paratesticular mass in the first year of life. A scrotal ultrasound is of limited utility in the routine evaluation of paratesticular masses found on physical examination. All patients with a concerning mass should undergo an exploration through an inguinal approach and frozen-section analysis as well as a radical orchiectomy if there is any concern for multifocality, involvement of adjacent structures, or a primary malignant tumor such as rhabdomyosarcoma. A simple tumorectomy may otherwise be considered, provided that neuroblastoma is confirmed on frozen-section analysis. Based on the current evidence in the literature, primary paratesticular neuroblastoma most commonly presents with early-stage, low-risk disease and has an excellent prognosis.

Conflict of interest

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