Adult Paratesticular Rhabdomyosarcoma Presenting as Acute Intestinal Obstruction—A Case Report

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Abstract Paratesticular rhabdomyosarcoma is a rare intra-scrotal tumor localized in the tunica vaginalis, epididymis, or spermatic cord. We report one such case in a 19-year-old man presenting with painless scrotal mass and features of acute intestinal obstruction. On exploratory laparotomy, a small nodule in sigmoid mesentery was found to be the cause of obstruction. The nodule was excised and orchidectomy was performed. Histopathological examination of both the specimens revealed features of embryonal rhabdomyosarcoma. The patient was treated with chemotherapy but died after 3 months.

Keywords Rhabdomyosarcoma · Paratesticular · Intestinal obstruction

Introduction

Paratesticular rhabdomyosarcoma is an uncommon lesion mainly seen in children. Adult paratesticular rhabdomyosarcoma is extremely rare and has high malignant potential. It spreads rapidly and thus needs to be diagnosed accurately and treated early [1]. We report a case of paratesticular rhabdomyosarcoma in an adult with unusual presentation as acute intestinal obstruction.

Case Report

A 19-year-old man presented to the emergency department with complaints of distension, colicky pain in the abdomen, and obstipation for past 4 days. On examination, the abdomen was found to be soft but distended with visible peristalsis. The patient also had a left-sided scrotal mass that he noticed 1 month ago. It was nontender, firm, nodular, and measuring 6 cm × 5 cm. The testis was not palpable, separate from the mass. Fine-needle aspiration (FNA) of the mass had been performed previously and a possibility of small round cell tumor was suggested, but the patient did not take any treatment. Serum Human Chorionic Gonadotrophin (HCG), alpha-fetoprotein, and Lactic dehydrogenase (LDH) levels were normal.

On emergency laparotomy, small bowel was distended and terminal ileum was adherent to the root of sigmoid mesentery. On decompression of small gut, a small nodule was seen in the root of mesentery kinking the terminal ileum. The nodule was excised, gut was decompressed, and inguinal orchidectomy was performed.

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Fig. 1 Gross photograph of scrotal mass showing a tumor in the paratesticular region along with normal testis

On gross examination, the mesenteric nodule was found to be gray white on cut surface. Cut section of orchidectomy specimen revealed a gray white fleshy homogenous tumor, measuring 6.0 cm×4.0 cm×2.5 cm, in paratesticular region compressing the testis (Fig. 1). Microsections from both the specimens were similar in appearance and revealed a cellular sarcoma with small round to spindle-shaped cells having scanty cytoplasm and high mitotic activity (Fig. 2). The tumor was seen within thin-walled vessels and infiltrated between seminiferous tubules. Although cross-striations were not evident, overall appearance was that of embryonal rhabdomyosarcoma. On immunohistochemistry, the tumor was found to be positive for desmin and myogenin and diagnosis was confirmed. After recovery, the patient was given adjuvant chemotherapy but he died after 3 months.

Fig. 2 Photomicrograph of a tumor showing small round to spindle-shaped cells with hyperchromatic nuclei and scanty cytoplasm (H&E 400×)

Discussion

Paratesticular rhabdomyosarcoma usually presents as an asymptomatic scrotal mass and misdiagnosed as a vaginal hydrocele or testicular tumor. It has no specific tumor markers. Its various histological subtypes are embryonal, pleomorphic, alveolar, and mixed types [1]. Embryonal rhabdomyosarcomas typically arise in the head and neck region or in the genitourinary tract although they may occur at any primary site. They are the most common malignant tumors of the spermatic cord [2]. Spindle cell variant is most frequently observed at the paratesticular site [3]. Origin of paratesticular rhabdomyosarcoma is thought to be either from the normal striated muscle present in the gubernaculum testis and cremasteric muscle or from undifferentiated mesenchymal (stem) cells. Occasionally, teratoma of the testis with abundant striated muscle component may pose a diagnostic dilemma. Although both the tumors occur at young age and are malignant, treatment for both is different. Therefore, they have to be differentiated by a careful search for other tissue components of teratoma [4]. The reported 5-year survival for paratesticular rhabdomyosarcoma is 30 % [5]. Distant spread of the tumor is most frequently to pelvic, para-aortic, and sometimes mediastinal lymph nodes. The second most frequent site is lungs [4]. Surprisingly in our case, these organs were spared but the patient presented with stage IV disease (i.e. distant metastasis), whose prognosis is extremely poor.

Conclusion

Paratesticular rhabdomyosarcoma is a rare clinical entity in adults. Its awareness, early diagnosis, and intervention need to be emphasized since it grows and metastasizes rapidly leading to poor prognosis.

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