Pediatrik Cerrahi Dergisi, 12:52-53, 1998

Perianal rhabdomyosarcoma mimicking rectal prolapsus in an infant: An unusual case

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Özet

Rektal prolapsusu taklit eden perianal rabdomiyosarkom olgusu

Rabdomiyosarkom çocukluk çağının en sık karşıla-şılan sarkom türü iken, primer pararektal rabdomiyo-sarkom nadirdir. Çocuklarda bu tümör perianal polipoid kitle gibi görülür ve sıklıkla yanlış tanı alır. Biz, rektal prolapsus yanlış tanısı ile gönderilen bir perianal embriyonal rabdomiyosarkom olgusunu sunuyoruz. Tümör eksize edilerek cilt primer kapatılmış ve sorunsuz iyileşmiştir. Tanı histolojik olarak doğrulanmıştır.

Anahtar kelimeler: *Perianal rabdomiyosarkom*

Summary

Rhabdomyosarcoma is the most common childhood sarcoma, whereas, primary pararectal rhabdomyosarcoma is very rare. In children, the tumour presents as a perianal polypoid mass, and is often initially misdiagnosed. We present an infant with perianal embryonal rhabdomyosarcoma, which was misdiagnosed as a rectal prolapsus and was transferred to the department of pediatric surgery for further treatment of rectal prolapsus, which was irreducible. The tumour was excised; wound closed primarily and healed well rapidly. The diagnosis was confirmed by histologic examination.

Key words: *Perianal rhabdomyosarcoma*

Introduction

Rhabdomyosarcoma (RMS) may present in the perianal area and involve the external anal sphincter and pelvic floor muscles [4]. The cases represent 2 % of all cases of childhood RMS. The tumour may present as a subcutaneous mass or as a verrucous superficial tumour [2,3]. Diagnosis may be delayed because of the fact that a mass may be misdiagnosed as a perineal abscess, and the verrucous appearance may be confused with viral condylomata.

Case Report

An 11 months old boy was admitted to Zeynep Kamil Hospital, with a perianal mass. Two months prior to admission, the perianal mass was noticed by his family. This was misdiagnosed as a rectal prolapsus by a general practitioner and he was transferred to the department of pediatric surgery for further treatment as an irreducible rectal prolapsus.

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The tumour was a red, rubbery, uniform, 4x5x6 cm polypoid mass originating from a 2 cm base on the right lateral side of the perianal verge. The lesion had ulcerations and bleeding (Figure 1). Rectal examination revealed no internal macroscopic or palpable tumour. Laboratory examinations were within normal limits. Bone marrow biopsy as well as ultrasound, CT and bone scanning were negative.

The tumour was excised with a round incision from the buttock into the anal verge including part of the anal sphincter under general anesthesia. Wound was closed primarily and healed well without wound detachment. Histopathologically, the tumour was composed of poorly differentiated small round or oval cells with hyperchromatic nuclei, with varying degrees of cellularity. Some of the tumor cells were PAS (periodic acid-schiff) positive, revealing glycogen content.

Immunohistochemically Desmin was strongly positive. The histopathologic diagnosis was embryonal RMS and the patient was referred for chemotherapy.

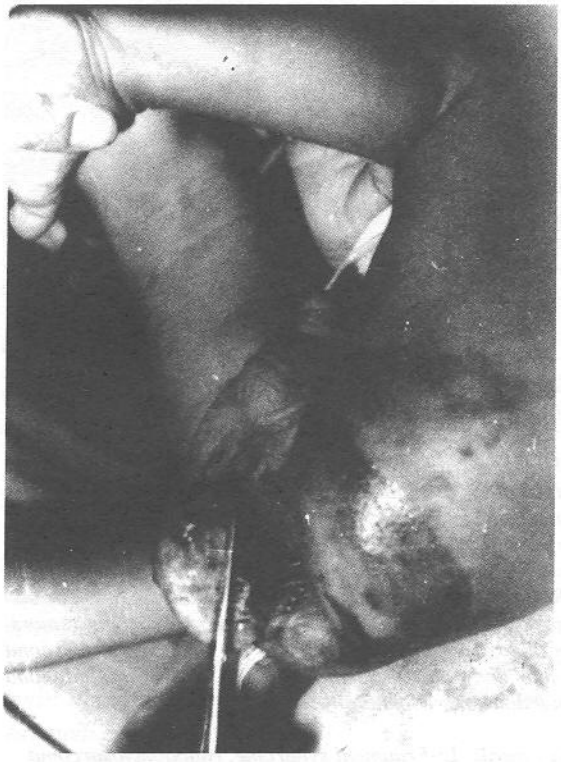

Figure 1. Polypoid tumor presenting at the anal margin.

Discussion

Although RMS can occur in mesench-ymal tissue in any part of the body, the perianal region is an extremely rare site for the primary lesion [1,2,3]. Only 14 cases, including eight adults and six infants, have been reported in the medical literature until 1983 [2].

In 1986, Moir et al, have reported another perianal RMS case in an infant [3]. Our recent experience of a 11-month-old infant with perianal RMS treated by local excision with chemotherapy is the eighth reported childhood case with long-term survival.

Signs and symptoms of RMS depend on the location of primary tumour and metastases. However, perianal RMS is not easy to diagnose in its early stages. Previously reported cases were misdiagnosed as a papilloma or fibroma [3] or abscess [2] or hemangioma [5]. Our patient was also misdiagnosed as having a rectal prolapsus for two months.

Current treatment of childhood RMS tends to reduce extended surgery and other treatment procedures to protect vital organs and to preserve normal growth and development have gained popularity [2]. The surgical tendency is to leave the wound edges open for secondary healing [3]. However, in our case, the wound had been closed primarily after the tumour excision.

The wound healed rapidly in seven days without any complications like hematoma, infection or detachment. We think that if the wound healing was not good after primary repair, it would be detached and secondary healing would occur. We must give a chance for primary wound healing in these patients.

In conclusion, early diagnostic studies and careful histological review of a perianal mass is mandatory. Also RMS should be taken into consideration in the differential diagnosis of the other perianal lesions.

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