CASE REPORT

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Case Report of a Local Recurrence of Spindle Cell Embryonal Rhabdomyosarcoma

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ABSTRACT

Introduction: Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children and adolescents. Spindle cell RMS is a rare variant of embryonal RMS that has a predilection for young males. **Aim:** We are presenting here a case of a local recurrence of an embryonal variant of the spindle cell RMS in a 19-year-old male. **Case report:** In this report it is described the study of patient with local recurrence of spindle cell embryonal RMS of the left testis after left orchiectomy and adjuvant chemotherapy. Computed tomography of the abdomen was used to evaluate the tumor. The recurrent mass was about 7,5cm and the patient was operated and discharged after 6 days in a good condition. Six months after the operation the patient had a new recurrence of RMS in the left *retroperitoneal* space. **Conclusion:** RMS is a malignant tumor of mesenchymal origin that is treated by a combination of surgery, chemotherapy, and radiation. However, up to one-third of patients experience recurrence.

Keywords: Rhabdomyosarcoma, embryonal, spindle, recurrence.

1. INTRODUCTION

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in children and adolescents, accounting for approximately 5% of all pediatric cancers and about one-half of all soft tissue sarcomas. It is the third most common extracranial solid tumor in children (1). Rhabdomyosarcoma types include embryonal rhabdomyosarcoma (approximately 60%), alveolar (approximately 20%), pleomorphic (approximately 10%), and spindle (approximately 10%) (2-4). Spindle cell RMS is a rare variant of embryonal RMS that is associated with a favorable outcome, in comparison with other subtypes. However, in the adult population, there does not appear to be any prognostic advantage (5).

2. AIM

The aim of this article is to present reports of a case of a local recurrence of an embryonal variant of the spindle cell RMS in a 19-year-old male.

3. CASE REPORT

A 19-year-old male underwent a total left orchiectomy 2 years ago with a diagnosis of a spindle cell variant of embryonal rhabdomyosarcoma of the left testis. He received adjuvant chemotherapy. The patient had a repeat abdominal Computed Tomography (CT) scan 2 years after the operation, without having any other sign or symptom until then. The CT scan revealed a retroperitoneal mass measured 6,75cm x 5,27cm x 7,46cm in diameter, coming from the left spermatic cord (Figure 1 and 2). The surgery was made in the General University Hospital of Patras by a team of general and vascular surgeons. In the operation the tumor was found to be next to the left external iliac artery. It was decided to cut the left external iliac artery (Figure 3) and an anastomosis was made between left femoral artery and the left external iliac artery with a synthetic *polytetrafluoroethylene* (*PTFE*) graft (Gore-Tex 8mm x 50 mm) (Figure 4). The patient had no postoperative complication during his hospitalization and was discharged 6 days after the surgery in a good condition. Six months after the operation the patient had a new recurrence of RMS in the left *retroperitoneal* space.

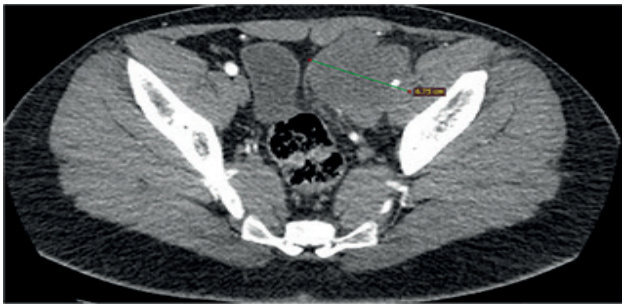

Figure 1. The patient's CT scan, showing the retroperitoneal mass

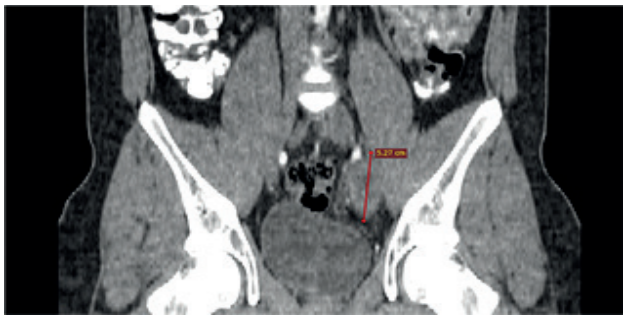

Figure 2. The patient's CT scan, showing the retroperitoneal mass

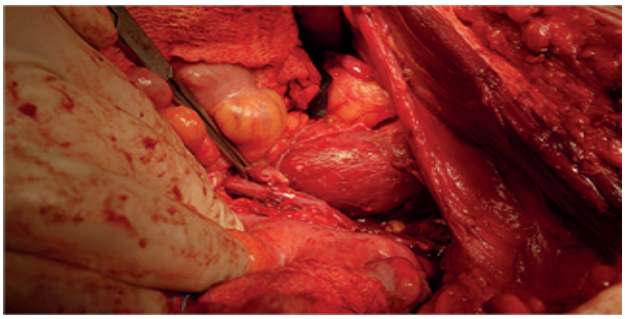

Figure 3. The retroperitoneal mass next to the cut left external iliac artery.

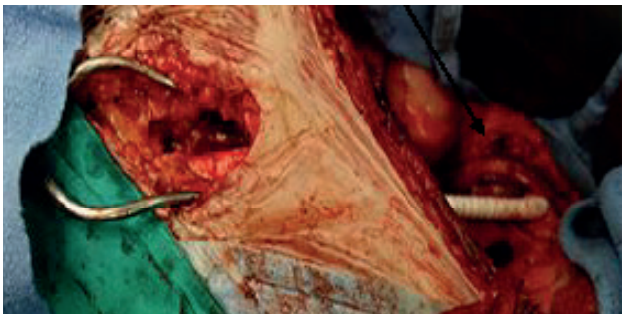

Figure 4. Anastomosis between left femoral artery and the left external iliac artery with a synthetic polytetrafluoroethylene (PTFE) graft.

4. DISCUSSION

The spindle cell variant of embryonal rhabdomyosarcoma was first recognized as a rare entity having a male predilection, propensity for occurrence in the paratesticular, and head and neck regions, and a low malignant potential, in 1992 by German-Italian Cooperative Soft Tissue Sarcoma Study. It accounts for 3-4.4 % of all subtypes of rhabdomyosarcoma (6-7). The tumours usually present as a gradually increasing painless, firm swelling (8-9). Even though embryonal RMS is common, the spindle cell variant is considered rare. RMS is treated by a combination of surgery, chemotherapy, and radiation. If the tumor is surgically inoperable, then initially radiotherapy and chemotherapy given to shrink a tumor followed by wide surgical excision of the tumor performed. Chemotherapeutic agents commonly used are vincristine, cyclophosphamide, dactinomycin, adriamycin, ifosfamide, and etoposide (5). The surgical management of patients with RMS is site-specific, and attempts for complete tumor resection should be made. Complete tumor resection is generally not advised if there is a significant risk for functional or cosmetic morbidity (for example, in head and neck RMS). Complete compartmental resection is not required for tumors arising within a muscle, as long as a minimum of 0.5 cm of margins is obtained (10). Although the majority of children with non-metastatic RMS now achieve complete tumor remission with current multidisciplinary treatment, which includes chemotherapy, radiotherapy and surgery, up to one-third of patients experience recurrence (11-13). Survival after recurrence is poor, and new salvage therapy strategies are needed (14-15).

5. CONCLUSION

The spindle cell variant is considered to be a rare type of rhabdomyosarcoma, that is treated by a combination of surgery, chemotherapy, and radiation. It is associated with a favorable outcome, in comparison with other subtypes. However, in the adult population, there does not appear to be any prognostic advantage. In addition, survival after recurrence is poor.

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