

Adult Pleomorphic Rhabdomyosarcoma of the Peroneal Compartment; Rare Case Report

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ABSTRACT

Pleomorphic Rhabdomyosarcoma (RMS) is a rare form of soft tissue sarcoma in adults. Due to the rarity of adult RMS, information regarding its clinical and biologic characteristics is very limited. I report a case of embryonal RMS of lower extremity involving the right leg peroneal compartment in a 37-year-old man. There was metastasis of the tumor to the ipsilateral inguinal lymph nodes at diagnosis. The patient underwent surgery with enbloc excision of the tumor from the peroneal compartment and ipsilateral inguinal lymphnode biopsy. Histopathological examination revealed high-grade pleomorphic RMS and postoperatively he under went chemotherapy and six cycles of radiotherapy.

Key words: Pleomorphic rhabdomyosarcoma, Adult, Peroneal compartment

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INTRODUCTION

Rhabdomyosarcoma (RMS) is a highly malignant mesenchymal tumor thought to originate from immature striated muscle. It is characterized by the presence of cells having an identifiable striated muscular differentiation with rhabdomyoblasts cells. The pleomorphic form is typically more frequent in adults and has a poor prognosis^[1,2]. RMSs are solid tumors that are common in children, representing 5% of all childhood cancers^[1,2,3,4]. In contrast, RMSs are rare in adults, with soft-tissue sarcomas making up fewer than 1% of malignancies in adults and RMSs accounting for 3% of all soft tissue sarcomas^[1].

In adults RMS is reported to predominantly affect adults aged more than 50 years especially affecting the extremities^[2]. The biological behavior and prognosis of adult pleomorphic rhabdomyosarcoma is currently poorly understood due to scarcity published information.

CASE REPORT

A 37 years old male driver presented to my office with history of rapidly progressive right leg swelling and pain over a 3 months duration attributed to a football injury. He had taken analgesics with a reduction in pain but swelling progressively increased in size.

Physical examination revealed swelling confined to the lateral compartment of the leg side (Figure 1). Swelling was firm in consistency and had associated inguinal lymphadenopathy, and reduced sensation in the peroneal nerve distribution.

Radiographs of right tibia and fibula showed only soft tissue swelling without bony involvement. His blood parameters were within normal range. The X rays of the tibia and fibula were normal. Chest and abdominal CT SCANS did not reveal any metastatic lesions.

He was admitted to The Nairobi Hospital, Kenya on 15/12/2017 for surgery. He was operated under general anesthesia with lateral incision. Intraoperatively tumor was confined to peroneal compartment and involved all the peroneal muscles. Enbloc excision of tumor was

done followed by inguinal lymph node biopsy. Grossly tumor appeared grey white to grey brown of size 15 × 7 × 5 cm and tubular in appearance (Figure 2). Histopathological examination showed a highly cellular tumour with typical features of pleomorphic RMS composed of hyperchromatic pleomorphic cells with areas of large multinucleated tumor giant cells having bizarre nuclei (Figure 3). A diagnosis of pleomorphic sarcoma with differentials of fibrosarcoma, rhabdomyosarcoma and synovial sarcoma was made. The histologic type was further confirmed by immunochemistry with the tumour being Vimentin positive and Desmin positive but negative for S100, AE₁/AE₃ and CD 117. Postoperatively he was referred to the radio-oncology unit where he undertook 6 cycles of radiotherapy and chemotherapy for 6 months. There was no local or systemic recurrence of symptoms with a follow-up of 1 year.

DISCUSSION

Adult presentation of RMS is very rare and especially at the age this patient presented. On literature search there is no reported case involving the peroneal compartment. The histological subtypes of rhabdomyosarcoma are embryonal, alveolar, and pleomorphic. The embryonal subtype is the commonest subtype, accounting for up to 49% of all RMSs^[5]. The alveolar subtype accounts for approximately 30% of all RMSs and most commonly affects adolescents^[2,5,7]. It is most commonly an intramuscular tumor in the soft tissues of the extremities with the thigh in the lower limb being a common site.

The pleomorphic subtype is the least common subtype and is reported to commonly affect patients older than 45 years^[2]. It arises most commonly in the skeletal muscles of the thigh. It is histologically similar to a malignant fibrous histiocytoma and the specific diagnosis is best done with immunochemistry as in this case report.

In adults with RMS, lymphadenopathy appears to be more common as is seen in this case. Specifically, 46% of patients had lymph node involvement at diagnosis in one series. Lymphadenopathy was most prominent in alveolar tumor subtypes and was least common in extremity tumors and pleomorphic tumor subtypes^[8,9]. Pulmonary metastases have been reported to be the most frequent site of distant disease.

The presence of metastases at presentation along with tumor size, tumor resectability, and patient age at presentation are also prognostic factors. In a series from Memorial Sloan-Kettering, the relative proportion of extremity and pleomorphic tumors increased with age, and survival decreased with increasing age in both univariate and multivariate analyses^[10]. In that series, increasing age was not associated with worse survival, regardless of whether it was treated as a continuous or dichotomous variable^[10].

The histologic subtype has a bearing on prognosis with the Embryonal tumors being associated with the most favorable outcome and pleomorphic, the least favorable^[8,9].

Treatment of patients with RMS is usually primarily surgery if excision is attainable, followed by radiation therapy and chemotherapy as was done in this case. Surgical resection is done for local disease control^[1,2,3]. Adjuvant chemotherapy and radiotherapy are routine for systemic disease control and to reduce chances of recurrence.

The overall 5-year survival rate in adults is reported at 31% ± 8% whereas 70% 5 year survival is recorded the paediatric population^[9]. The independent predictors of long-term survival are localized/locoregional disease at presentation and complete response to chemotherapy. In addition, female gender and tumor size appear to be associated with complete response to chemotherapy.

In a recent case report of a 48 year old female with pleomorphic RMS with a 6 year follow up the use of transcatheter intraarterial



Figure 1 Swelling of the peroneal compartment.



Figure 2 Intraoperative tumour.



Figure 3 Gross picture of the tumour after excision.

therapy involving high concentration delivery of chemotherapeutic drugs directly to the tumour and cryosurgery has given a very favorable outcome with limb preservation^[4]. Several sessions of cryosurgery were done at -160 to -180° C prior and after total surgical resection. That patient has been followed up for 6 years with no evidence of recurrence^[4]. This is a viable option that may become routine in the future.

CONCLUSION

The current treatment of rhabdomyosarcoma is multimodal involving surgery followed by several cycles of radiotherapy and chemotherapy. Newer treatment option including Trans Arterial Chemotherapy and Cryosurgery are novel therapies to consider. A thorough assessment and regular follow up is recommended for adult of RMS forms

that are associated less than 50% 5 year survival. The patient is on follow up for any recurrence.

ETHICAL STANDARDS

The patient gave the informed consent to the publication of the case study. There is no conflict of interest.

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