Adult Alveolar Rhabdomyosarcoma on Extremity, Successful Treatment with Radiotherapy following Chemotherapy: Serial Case Report

Abstract

Alveolar rhabdomyosarcoma (RMS) is one of the four subtypes of RMS. Alveolar RMS is the rare type found in adults, with the worse prognosis. We report 2 serial cases of alveolar RMS on extremity which was only treated with subtotal excision or incision biopsy. We further gave a total dose of 70 Gy radiotherapy on the local tumor and 50 Gy prophylaxis dose on regional lymph node after 6 cycles of anthracyclines-based chemotherapy. Postradiotherapy, contrast computed tomography scan revealed no tumor mass left (complete response).

Keywords: *Alveolar rhabdomyosarcoma, chemotherapy, radiotherapy*

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Introduction

Rhabdomyosarcoma (RMS) is a rare soft-tissue tumor, comprises 1% of solid tumors and 3% of adult-type sarcomas. It is an aggressive tumor with high local recurrence and metastasis tendency. There are four subtypes: embryonic, alveolar, pleomorphic, and not otherwise specified (NOS). Embryonic is frequently encountered in children and adolescent, while pleomorphic and NOS RMS commonly occur in adults.[1,2] Alveolar subtype has the worst prognosis, with therapeutic controversies due to limited case reports and references' availability. The main modality for this disease includes surgery, radiotherapy, and chemotherapy.

Case Reports

Alveolar rhabdomyosarcoma on medial right femoral region

A 39 year old female came with fixated nodule on her medial right thigh. A subtotal excision was done, removing 70% of the initial tumor volume sized 6 cm × 4 cm × 3 cm. Histopathology examination supports alveolar RMS [Figure 1]. Tumor staging reveals stage 3 with Karnofsky score – 80%. One month following surgery, we delivered adjuvant chemoradiotherapy. Chemotherapy consisted of vincristine, dacarbazine, epirubicin, and cyclophosphamide for 6 cycles every 3 weeks.

Radiotherapy was delivered 12 weeks after, with 70 Gy dosages to the primary gross tumor (35 fractionation) and 50 Gy prophylactic dose on ipsilateral inguinal region (25 fractionation). Radiotherapy field margin is 5 cm from tumor bed, surgical scar and left over tumor margins. We also did 2 cm skin sparing from the radiation field. Thermoplastic Orfit® mask was used for area fixation. Radiotherapy was delivered with conventional 2-dimensional plan-parallel anteroposterior – posteroanterior technique using Cobalt-60 teleradiotherapy (Gammabeam 100-80, Best Theratronics®).

During the late evaluation, a small tumor mass was still palpable. This patient also encountered Grade 3 radiodermatitis with prominent edema, erythematous and oozing skin lesion, as well as hyperpigmentation on the margins of the radiotherapy area. Radiodermatitis was then managed with sterile physiologic saline solution compress twice daily for 15 minutes per session, followed by application of topical 1% silver sulfadiazine. Three months later, no more palpable mass and radiodermatitis lesion were found. Contrast computed tomography (CT) scan revealed no observable tumor mass, with only scar tissue on radiotherapy area [Figure 2].

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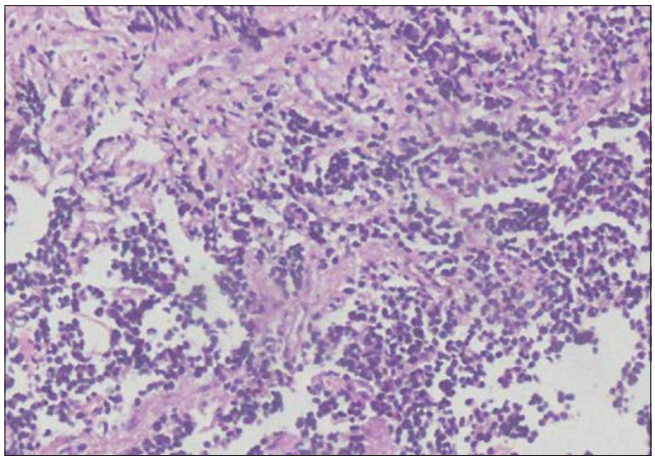

Figure 1: Histopathology showing: mesenchymal tumor cells, clumping in fibrovascular septae. Tumor cells have eosinophilic cytoplasm with round, hyperchromatic, uniform nucleus and prominent mitosis features (H and E, ×200)

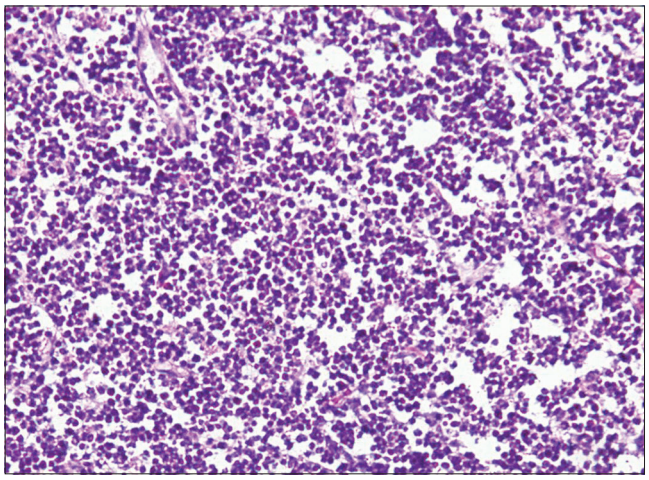

Figure 3: Histopathology showing: diffuse, dense oval to round mesenchymal tumor cells, separated by fibrovascular septae, with hyperchromatic nucleus and eosinophilic cytoplasm (H and E, ×200)

Alveolar rhabdomyosarcoma on right triceps region

A 27 year old female came with fixated nodule on her right triceps, sized 6 cm × 6.5 cm × 4 cm. Incision biopsy was done, and histopathology examination confirms alveolar RMS [Figure 3]. Staging reveals Stage 3 with Karnofsky score –70%. We treated this patient with chemoradiotherapy. Chemotherapy regimens consisted of doxorubicin, mesna, and haloxane every 3 weeks for 6 cycles with partial response result.

Radiotherapy was delivered 14 weeks postchemotherapy with similar technique, radiotherapy device, and fixation method with the first case. Dosage given to gross tumor of all compartment affected with the tumor infiltration was 70 Gy (35 fractionation), and prophylactic dose on ipsilateral axillar fossa was 50 Gy (25 fractionation).

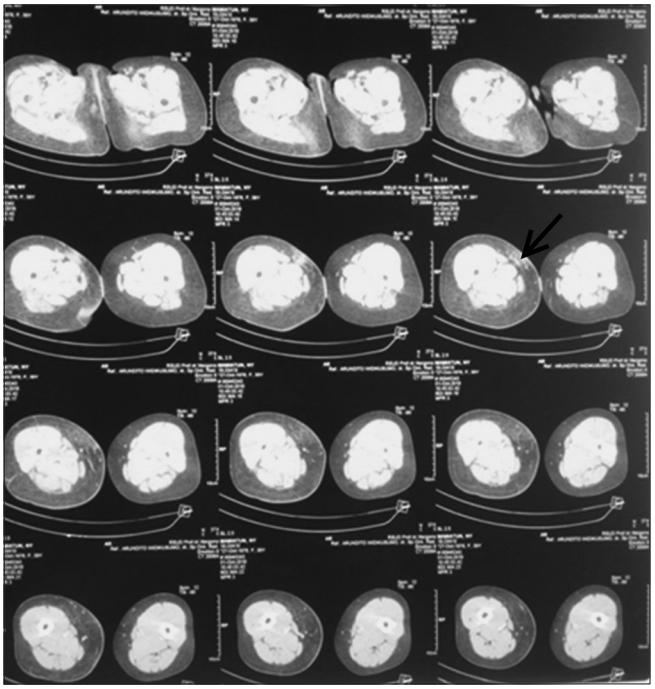

Figure 2: Image of postradiotherapy contrast computed tomography scan, no detectable tumor mass, with observable postradiotherapy cicatricial tissue

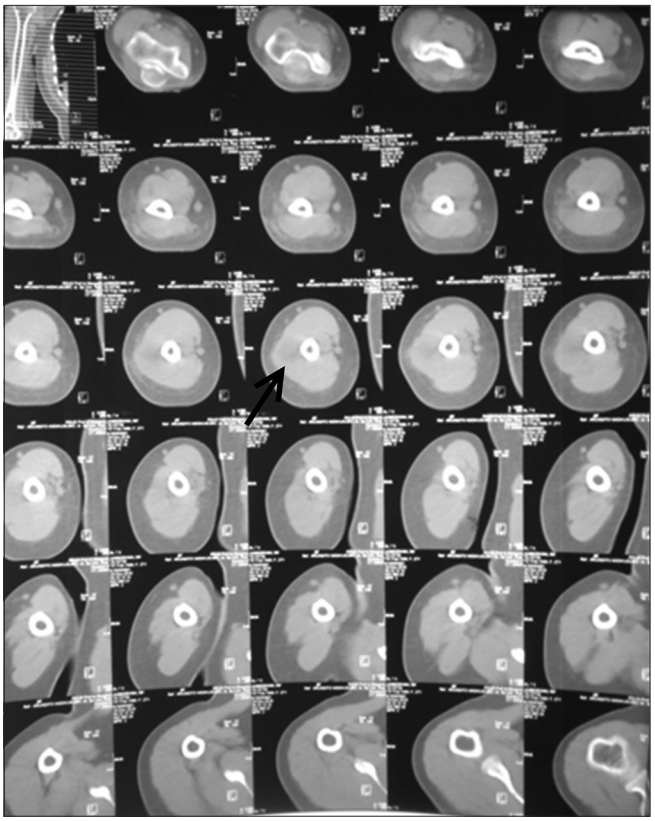

Figure 4: Postradiotherapy contrast computed tomography scan with no observable tumor on the triceps region. There is a minimal postradiotherapy edema

The tumor has 60%–70% volume regression after 40 Gy, and it completely disappeared after 60 Gy. Acute Grade 2 radiodermatitis occurred at 70 Gy dose and self-resolved 3 months after radiotherapy ended. Further, contrast CT scan found no observable tumor mass with only minimal edema on the radiotherapy area [Figure 4].

Discussion

Alveolar is the rarest adult RMS with the worst prognosis when compared to other subtypes. The most frequently affected area are head and neck as well as extremity.[2,3] Unlike children, the prognosis and outcome of adult RMS are very poor. The data from the United States Surveillance, Epidemiology, and End Result reveal that 5- and 10-year survival rate for adult RMS is only 27% and 21%, respectively, far lower when compared to 61% and 58%, respectively, in children.[2] However, those data did not further describe survival rate of RMS subtypes.

Alveolar RMS treatments consist of surgery, radiotherapy, and/or chemotherapy. Surgery is the mainstay therapy, and surgical margin status is therefore crucial. Tumor-negative surgical margin has a more favorable prognosis compared to positive surgical margin. A combination with adjuvant radiotherapy is the most popular treatment option, with better outcomes when compared to surgery alone. Preoperative and postoperative radiotherapy may both improve RMS clinical outcome.[3] Chemotherapy is rarely used in early RMS stage compared to radiotherapy; however, it is frequently used in advanced stage and RMS with metastasis.[3,4]

Both cases demonstrate large gross tumor volumes which cannot be operated to achieve tumor-negative surgical margin. Chemotherapy was delivered considering that alveolar RMS tumor volume is directly proportional to metastasis incidence. Esnaola *et al*.[4] reported that the tumor size of <5 cm, 5–10 cm, and >10 cm each carry the metastasis possibility of 13%, 31%, and 75%, respectively (P = 0.01). Therefore, anthracyclines-, actinomycin-d-, or ifosfamide-based chemotherapy should be delivered to these kinds of cases.[2,4,5]

Postsurgical radiotherapy may enhance failure-free survival,[6] and patients with inoperable tumor or postsurgery microscopic and macroscopic RMS are always treated with radiotherapy. Alveolar RMS has the most frequent lymph node metastasis.[3,7] Therefore, we decide to treat both cases with radiotherapy on the primary gross tumor and regional lymph node. Radiotherapy dose on gross tumor varied between 41 and 78 Gy (median 60 Gy), as for the lymph node varied between 40 and 75 Gy (median 50 Gy).[3] Kelly *et al*.[8] reported the delivery of 30 fraction radiotherapy (with conventional fractionation of 1.8–2 Gy/day) on paranasal sinus alveolar RMS. Furthermore, Saha *et al*.[9] deliver radiotherapy with 70 Gy total dose using plan-parallel anteroposterior–posteroanterior technique, following 6-cycle chemotherapy regimen consisting of mesna, doxorubicin, ifosfamide, and dacarbazine. Radiotherapy dose for extremity alveolar RMS in our policy is 66–70 Gy for primary gross tumor (with conventional fractionation of 2 Gy/fraction) and 50 Gy for prophylaxis on the regional lymph node. We suggest that radiotherapy should be delivered earlier, at 4–8 weeks instead of 12 and 14 weeks following chemotherapy.

Conclusion

Alveolar is the rarest and the worst RMS subtype in adults. In our cases, the treatment outcomes for inoperable alveolar RMS, treated with 70 Gy radiotherapy total dose on gross tumor and prophylactic dose of 50 Gy on regional lymph node following 6 cycles of anthracyclines-based chemotherapy had resulted in a favorable complete response.

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Conflicts of interest

There are no conflicts of interest.

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