Case Report

Rhabdomyosarcoma thigh in a 45-year-old male: A rare presentation

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ABSTRACT

Rhabdomyosarcoma (RMS) is a common childhood cancer, constituting more than 50% of all soft tissue sarcoma, but it is an uncommon neoplasm in adult. We reported a case of 45-year-old male patient presented with a huge swelling in his left thigh. Magnetic resonance imaging of thigh revealed soft tissue mass involving the deep muscular compartment. Core biopsy and immunohistochemistry confirmed it as an embryonal RMS. The tumor showed a complete response after three cycles of neoadjuvant chemotherapy (CT) and then was treated with three more cycle of CT followed by adjuvant radiotherapy. This case is being reported on account of its rarity at this age and nonsurgical treatment. Pertinent literature is being reviewed.

Key words: Complete response, neoadjuvant chemotherapy, rhabdomyosarcoma

INTRODUCTION

Rhabdomyosarcoma (RMS) is a common childhood cancer, constituting more than 50% of all soft tissue sarcomas (STS), although it is infrequent in adults. STS make up less than 1% of all adult malignancies and RMS accounts for 3% of all (STS).[1] RMS can occur within any mesenchymal tissue, although it has a predilection for the head and neck, genitourinary organs, retroperitoneum and extremities.[2] The embryonal subtype is the most common, representing up to 60-80% of tumors. Alveolar tumors are more common among adolescents, often arise in the extremities and carry a worse prognosis.[3]

CASE REPORT

The case we present here is about a 45-year male patient who presented with a gradually increasing swelling over lower part of left thigh for 4 months. On examination-a nontender mass 12 × 11 cm in size, circumferentially involving left thigh, without any skin or underlying bony fixity, was seen. There was no distal neurovascular deficit. No lymph node was palpable. Magnetic resonance imaging (MRI) of left thigh [Figure 1] shows a large lobulated infiltrating soft tissue mass involving circumferentially the deep muscular compartments of the thigh predominantly involving the vastas medialis, vastas intermedius, vastus lateralis and adductor magnus without any bony involvement extending up to the knee (Intergroup Rhabdomyosarcoma Study Group [IRSG] Stage-3). Ultrasonography (USG) guided fine-needle aspiration cytology from the swelling shows - small round cell tumor-embryonal RMS [Figure 2]. Core biopsy from the lesion confirmed the diagnosis of embryonal RMS. Immunohistochemistry study showed that the tumor was positive for desmin and myogenin [Figures 3 and 4]. Complete blood count, liver function test, renal function test, chest X-ray, USG whole abdomen, two-dimensional echocardiography was within normal limit. The patient was treated with neoadjuvent chemotherapy (CT) with injection mesna, doxorubicin, ifosfamide, and dacarbazine regimen. The tumor showed a complete response (disappearance of all target lesions. any pathological lymph nodes, whether target or nontarget, must have reduction in short axis to <10 mm) after three cycles of neoadjuvent CT according to the response evaluation criteria in solid tumor criteria.

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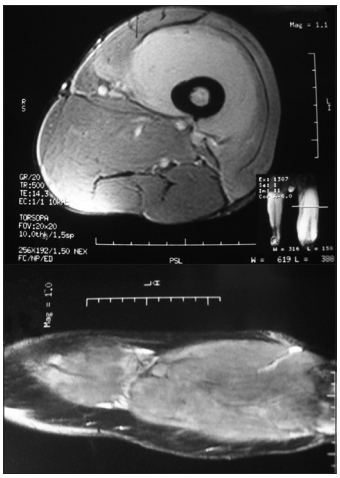

Figure 1: Prechemotherapy magnetic resonance imaging of left thigh showing a large lobulated soft tissue mass involving circumferentially the deep muscular compartments of left thigh predominantly involving the vastus medialis, vastus intermedius, vastus lateralis, and adductor magnus without any regional bony involvement

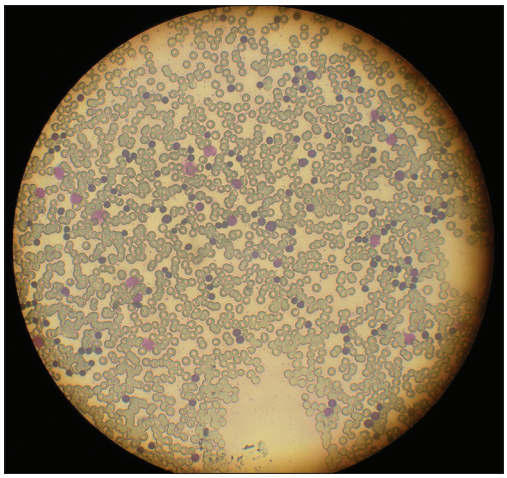

Figure 2: Cytosmears reveal high cellularity composed of dispersed population of neoplastic cells showing nucleomegaly with chromatin clumping and inspicuous nucleoli. The cytoplasm is scanty mitosis is seen. Numerous apoptotic cells are seen. Small round cell tumor-embryonal rhabdomyosarcoma (cytology smear, MGG stain, ×40)

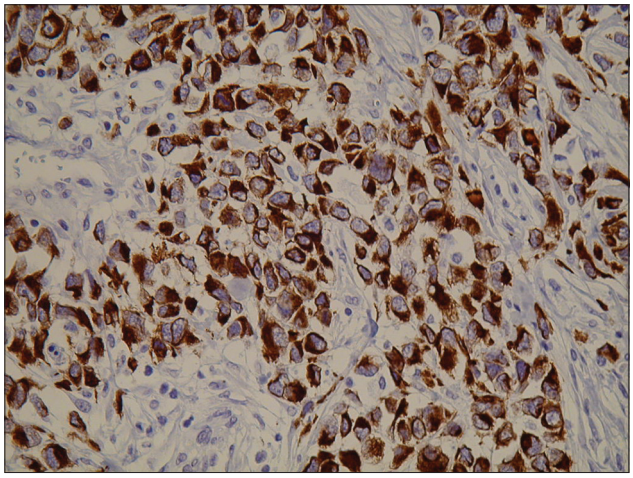

Figure 3: Immunohistochemistry study showing desmin positive (DAB, ×200) MGG stain, ×40)

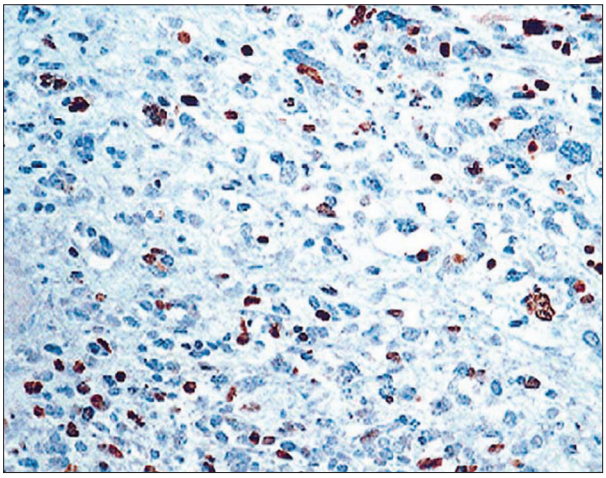

Figure 4: Immunohistochemistry study showing myogenin positive (DAB, ×200)

The MRI left thigh showed complete resolution of the prechemotherapy MRI findings of diffusely infiltrating soft tissue mass [Figure 5]. The patient was sent for surgical assessment, but as the post-chemotherapy (three cycles) MRI revealed no residual mass, no surgery was done by the surgeon. Patient completed six cycles of CT. 3 weeks after completion of 6th cycles of CT patient received external beam radiotherapy (EBRT). EBRT was given with Co-60 teletherapy machine by conventional technique using anterior-posterior-posterior-anterior portal. In Phase 1 50 Gy in 25 fractions was given to prechemotherapy volume with 5 cm proximal and distal margins and 2 cm circumferential margin, sparing a 2 cm strip of skin over the medial aspect of thigh (lymphatic corridoor). In Phase 2 prechemotherapy tumor volume with 2 cm margins were boosted to a total dose of 70 Gy in total 35 fractions. Now patient is under follow-up and he is free from any loco-regional recurrence or metastases for the last 1 year.

DISCUSSION

Rhabdomyosarcoma is a common childhood cancer, constituting more than 50% of all STS, most aged 3-12 years. Although, it is infrequent in adults: STS make up less than 1% of all adult malignancies and RMS accounts for 3% of all STS.[1] RMS can occur within mesenchimal tissue at any site, although it has a predilection for the head and neck, genitourinary organs, retroperitoneum, and extremities.[2] The embryonal subtype is the most common, representing up to 60-80% of tumors at above sites. Alveolar tumors are more common among adolescents, often arise in the extremities and carry a worse prognosis. On immunohistochemistry embryonal RMS may express vimentin, desmin, actin, myoglobin, myosin, creatine kinase

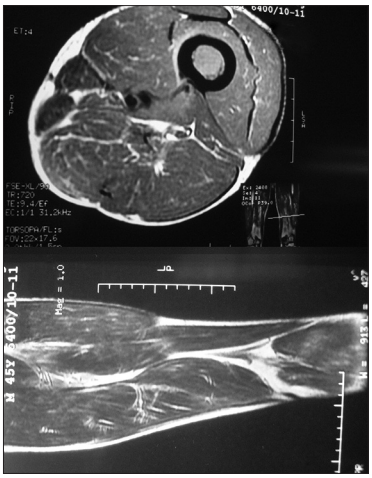

Figure 5: Magnetic resonance imaging (MRI) of left thigh after three cycle of chemotherapy showing mild hyperintensity (edema) in the periosseous muscles of the deeper compartment of the left thigh predominantly in the supracondylar region. When compared to the prechemotherapy MRI the diffusely infiltrating soft tissue mass has been completely resolved

M, titin, dystrophin and acetylcholine receptor antigen.[3] In intergroup RMS study-IV, the relative subtype proportions were embryonal, 70%; alveolar, 20%; and others, 10%.[4] Based on the results of studies like IRSG, generally accepted treatment guidelines for childhood RMS include gross total resection with preservation of function, systemic CT and radiation therapy for all but completely resected tumors of embryonal subtype.[4,5] Prognosis can be determined by stage, histological classification, age, and site of origin. Younger patients tend to have a more favorable prognosis. Histopathologicl subtype in adults with RMS appears to have no prognostic relevance. Embryonal RMSs with diffuse anaplasia may have a worse outcome than the other subsets of embryonal RMS. Parameningeal and extremity tumors tend to have a bad outcome compared to other locations, whereas orbital and paratesticular tumors tend to have a better one. Experiences from childhood RMS are extrapolated widely to adults with this disease and despite the use of multimodal therapy; the prognosis in older patients appears to be worse than in children.[6,7] Even in the IRSG studies, an adverse effect of increasing age on outcome has been documented.[4] Staging is accomplished by clinical evaluation (IRSG Stage) or surgicopathological evaluation (IRSG Group).[8] RMS in adults is very rare and literature regarding its management is limited. Detailed reports of multimodal treatment outcome, patterns of failure and prognostic factors in adult patients with RMS are few.[9,10]

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