Case Report

An unusual case of pleomorphic rhabdomyosarcoma of shoulder in an adult patient

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ABSTRACT

Rhabdomyosarcoma (RMS) is a highly malignant soft tissue sarcoma (STS). It is the most common childhood STS and is exceedingly rare in adults. The pleomorphic subtype affecting patient older than 45 years is the least common subtype. It is histologically similar to a malignant fibrous histiocytoma. Many pleomorphic RMSs (PRMSs) have been reclassified as fibrous histiocytomas, thereby making the diagnosis of PRMS more unusual. Here, we report a case of PRMS in a 45-year-old male who reported with the painless soft tissue swelling over the posterior aspect of left shoulder for 1 year. Magnetic resonance (MR) imaging and MR angiography showed a large well defined heterogeneously enhancing soft tissue mass lesion arising from posterior fibers of left Deltoid muscle. The patient received nine cycles of neoadjuvant chemotherapy with ifosfamide, epirubicin every 3 weeks. The patient underwent wide local excision of the tumor and received four cycles of adjuvant chemotherapy ifosfamide, epirubicin, and etoposide every 3 weeks. Locoregional adjuvant radiotherapy 66 Gy in 33 fractions was given by 3-dimensional conformal radiotherapy. Now the patient has a complete response on follow-up imaging 2 years after completion of radiotherapy. PRMS in adults has a significantly worse prognosis than that for other pleomorphic sarcomas with 12.5–50% of 1-year to 20-month disease-free survival and 27% rate of 5-year disease free survival. Thus, the correct and early diagnosis of PRMS is important.

Key words: Adults, chemotherapy, pleomorphic rhabdomyosarcoma, radiotherapy

INTRODUCTION

Rhabdomyosarcoma (RMS) is a highly malignant soft tissue sarcoma (STS) that arises from unsegmented, undifferentiated mesoderm, or myotome-derived skeletal muscle. RMS represents 5% of all childhood cancers.[1] It is the most common childhood STS accounting for more than 50% of cases diagnosed in childhood and adolescents.[2] Its annual incidence in the United States is 4.3 cases/million people younger than 20 years of age. Most cases occur in children younger than 10 years.[2] Although RMS may occur at any age, it is exceedingly rare in adults as shown by Weiss and Goldblum in 2001 (cited by Ferrari *et al*. and Saha *et al*.)[3,4] and their results revealed that in adults, STS constitute <1% of all malignancies and RMSs accounts for 3% of all STSs.[5] In children, it may occur at any site of the body, but the most frequently involved sites are the genitourinary, 31%; parameningeal, 25%; extremity, 13%; orbit, 9%; head and neck (excluding parameningeal tumors) 7%; retroperitoneum, 7%, trunk, 5%; and other sites 3%.[2]

Histologically RMS occurs as three subtypes embryonal (including botryoid), alveolar, and pleomorphic. Overall, the embryonal subtype is the most common subtype, making 49% of all RMSs. The alveolar subtype

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Access this article online

Quick Response Code:

Website: www.ccij-online.org

DOI: 10.4103/2278-0513.182060

Cite this article as: Azam M, Khurana R, Gupta A. An unusual case of pleomorphic rhabdomyosarcoma of shoulder in an adult patient. Clin Cancer Investig J 2016;5:274-7.

makes 30% of all RMSs and most commonly affects adolescents. The pleomorphic subtype is the least common subtype and almost exclusively affects patients older than 45 years.[6]

It is histologically similar to a malignant fibrous histiocytoma; in fact, many pleomorphic RMSs (PRMSs) have been reclassified as fibrous histiocytomas, making the diagnosis of PRMS an even rarer entity.[5]

CASE REPORT

A 45-year-old male presented to our institute with 1-year history of painless soft tissue swelling over the posterior aspect of left shoulder. The swelling progressively increased to 12 cm × 10 cm in size involving the left deltoid and lateral aspect of the upper third of arm. The swelling was firm, nontender and without any cystic consistency. Overlying skin was stretched, shiny, but the temperature was not raised. No engorged vessels were present on the swelling. The lump seems to be deep and fixed to the underlying structure. No lymph nodes were palpable in the axilla, elbow, and bilateral neck. There was no significant limitation of the shoulder joint movements. A clinical provisional diagnosis of STS was made. The patient had no comorbidities and had good performance status. Magnetic resonance (MR) imaging and MR angiography of shoulder joint showed a large well defined heterogeneously enhancing (T1 iso- to hypo-intense and T2 iso- to hyper-intense) soft tissue mass lesion measuring approximately 81 mm × 61 mm × 98 mm arising from posterior fibers of left deltoid muscle. The lesion was abutting proximal shaft of humerus. However adjacent cortical margin was intact. No involvement of the shoulder joint and neurovascular bundle was noted. Multiple feeding channels were seen supplying the neoplastic lesion arising from axillary and subclavian arteries and their branches [Figure 1]. A possible diagnosis of RMS was made based on the radiological findings. Whole body bone scan was negative for any bony metastasis. Fine-needle aspiration cytology from the swelling revealed spindle cell neoplasm. Histopathological examination (HPE) of the tru-cut biopsy specimen showed tumor composed of spindle cells as well as scattered bizarre cells with intervening myxoid areas. The tumor cells were moderately pleomorphic, with hyperchromatic nuclei, coarse chromatin, and moderate amount of eosinophilic cytoplasm. Immunohistochemistry (IHC) was positive for vimentin and desmin while negative for S-100, CD34, smooth muscle actin (SMA) and pan-CK [Figure 2]. HPE and IHC were consistent with spindle cell sarcoma possibly RMS. Metastatic workup with contrast enhanced computed tomography scan of thorax and abdomen were negative.

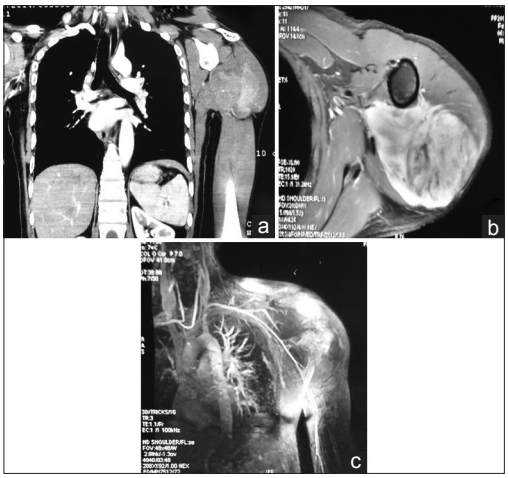

Figure 1: Magnetic resonance left shoulder. (a) T1-weighted coronal section showing iso- to hypo-intense soft tissue mass lesion. (b) T2-weighted axial section showing iso- to hyper-intense soft tissue mass lesion. (c) Magnetic resonance spectroscopy image showing multiple feeding channels supplying the neoplastic lesion arising from axillary and subclavian arteries and their branches

The tumor was staged according to the inter-group RMS study IV (IRS-IV) pretreatment staging system as Stage-3 and IRS Clinical Grouping Classification (after resection) as Group IIA.

Neoadjuvant chemotherapy (NACT) followed by wide local excision (WLE), adjuvant chemotherapy and adjuvant radiotherapy planned in view of the large tumor to facilitate the limb-sparing surgery with a clear margin. The patient received nine cycles of 3 weekly NACT with ifosfamide (2.0 g/m^2) day 1–4, epirubicin (60 mg/m^2) day 1. The patient underwent WLE of the tumor. Peroperatively, the mass was seen in the posterior compartment of the shoulder involving triceps and radial nerve and giving into the axilla. Radial nerve was sacrificed which was engulfed in tumor. Postoperative HPE showed 8.0 cm × 7.6 cm × 7.0 cm tumor. The tumor cells were moderately pleomorphic, with hyperchromatic nuclei, coarse chromatin, and moderate amount of pale eosinophilic cytoplasm. All surgical margins except anterior margin were clear of tumor cells. The anterior margin was close (<2 mm away from tumor). Nerve margins were clear. IHC showed a positive reaction for vimentin and desmin while negative for S-100, CD34 and SMA. HPE and IHC were consistent with RMS. The patient then received 4 cycles of adjuvant chemotherapy ifosfamide (2.0 g/m^2) day 1–4, epirubicin (60 mg/m^2) day 1, and etoposide (100 mg/m^2) day 1 every 3 weeks. A dose of 66 Gy adjuvant radiotherapy by three-dimensional conformal radiotherapy [Figure 3] in conventional fractionation with shrinking field technique was given after proper immobilization of the limb. Six megavolt X-ray photons were used. Now the patient has a complete response on follow-up imaging 2 years after completion of radiotherapy.

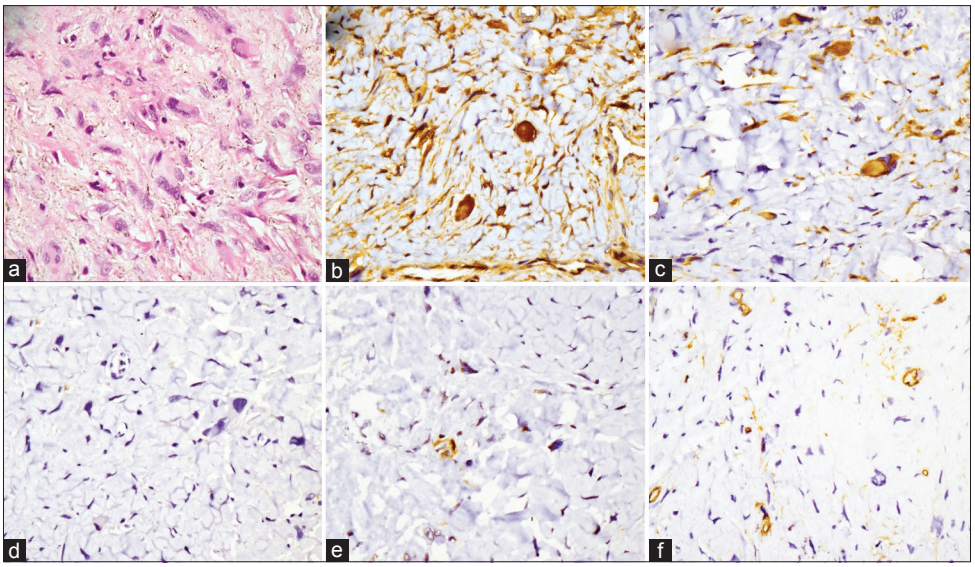

Figure 2: (a) Section shows singly scattered rhabdomyoblasts with eccentric pleomorphic nuclei and eosinophilic cytoplasm (H and E, ×400). (b) Rhabdomyoblasts show positivity for vimentin (IHC, ×400). (c) Rhabdomyoblasts show positivity for desmin (IHC, ×400). (d) Rhabdomyoblasts show negativity for S-100 (IHC, ×400). (e) Rhabdomyoblasts show negativity for smooth muscle actin (IHC, ×400). (f) Rhabdomyoblasts show negativity for CD34 (IHC, ×400)

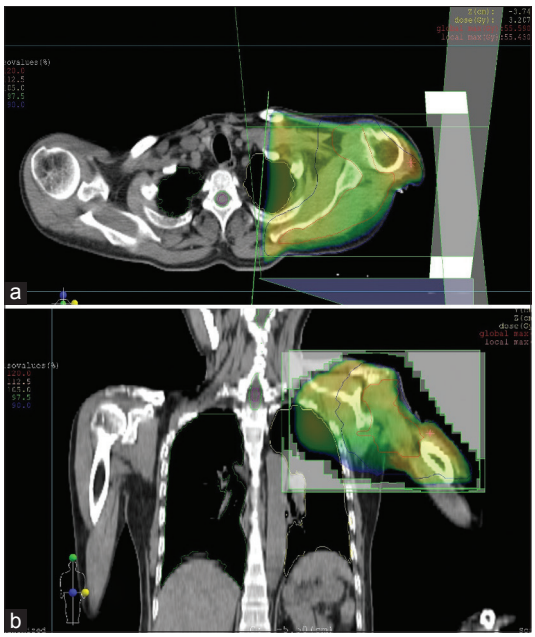

Figure 3: Three-dimensional conformal radiotherapy dose distribution. (a) Axial view. (b) Coronal view

DISCUSSION

RMSs are the most common childhood STS, accounting for 5% of all pediatric malignancies. Until adolescence, RMS are still among the most common STS, but they become more infrequent with older age and are rarely seen in patients who are older than age 45 years.[6,7] Pleomorphic is the least common of all, occurs in extremities and generally seen in elderly.[5]

Historically, stout first introduced PRMS into the literature in 1946 as "classical" RMS.[8] Histology of pleomorphic subtype is composed chiefly of spindle cells in a haphazard arrangement. These cells are generally large and show considerable variation in appearance. The characteristic feature of this tumor is large bizarre shape cells with nuclei situated in the expanded end of the cell.

In 1958, Horn and Enterline outlined four subtypes of RMS and called the classical ones "Pleomorphic RMS." In the largest case series of Furlong *et al*. in 2001, they reported only three cases of PRMS involving the upper extremity.[9]

There are differences in the anatomic sites that are involved and the prognosis in pediatric and adult patient groups. Parameningeal and extremity tumors tend to have a bad outcome compared to other locations. The diagnosis of pediatric-type sarcomas in adults is often challenging because of the unusual contextual clinical setting and morphologic features. Immunohistochemical studies have greatly facilitated this process.

Long-term follow-up results for RMS in adults are few. This is the reason why experiences from childhood RMS are extrapolated widely in adult patients. The treatment for adults who have RMS is not standardized thus having an impact on the overall survival.[10]

CONCLUSION

Adult RMS though a rare disease is quite heterogeneous. The biological behavior of adult RMS is poorly understood. Localized RMS should, therefore, be treated aggressively with multimodality approach consisting of surgery, radiotherapy, and chemotherapy. The primary aim of management should be a complete cure and maintaining the quality of life with

emphasis on preservation of function and cosmesis. Although studies on adult RMS are very few, they indicated that radiotherapy definitely improves local control but did not transform into overall survival. Careful long-term follow-up is necessary due to high risk of recurrence.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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