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Case Report

Case report of high-grade pleomorphic sarcoma (rhabdomyosarcoma): a missed diagnosis in the non-healing post-traumatic wound of the elderly

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ABSTRACT

Rhabdomyosarcoma (RMS) is known to be common childhood soft tissue sarcoma (STS). RMS is infrequent in adults. STS constitute <1% of all adult solid malignant sarcomas and RMS accounts for 3% of all STS. The tumor is divided into three main subtypes-embryonal, alveolar, and pleomorphic (most common in adults). The most common primary sites are extremities. It's an aggressive lesion with a high rate of metastasis. The patient presented with a non-healing wound over a posterior-medial aspect of the right leg with features suggestive of cellulitis. The patient gave a history of fall 5 months ago for which he took treatment at a local doctor, he continued to have severe pain for which an orthopedic opinion was taken and advised conservative management for an un-displaced fracture of the fibula. He later developed swelling over the lower-middle part of his right leg 3 months ago for which an ultrasound was done suggestive of a large amount of fluid suspicious of hematoma secondary to A-V malformation, an attempt for drainage of fluid was made leading to an evacuation of blood clots further procedure was abandoned and the patient transferred to our center. CT-angio demonstrated it as a heterogeneously attenuating solid cystic lesion. After appropriate consents-wound exploration was done with the evacuation of soft gelatinous material with pus discharge and blood clots. HPE-suggestive of high-grade pleomorphic rhabdomyosarcoma. The patient was advised amputation but denied any further treatment. Post-operative recovery was uneventful with no recurrence for the first 6 months after which the patient did not follow up. Post-traumatic mass at extremities should be evaluated with a high index of suspicion of STS. In case of doubt, either a preoperative biopsy or an intraoperative frozen section is a safe practice.

Keywords: Soft tissue sarcoma, Rhabdomyosarcoma, Pleomorphic sarcoma, Malignancy

INTRODUCTION

Rhabdomyosarcoma (RMS) 1st described by Weber in 1854, is a malignant soft tissue cancer of the skeletal muscle origin.[1] Rhabdomyosarcoma (RMS) is the most common childhood soft tissue sarcoma, accounting for more than 50% of all Sarcomas of the soft tissues. RMS is uncommon in adulthood. RMS accounts for just 3% of all soft tissue sarcomas in adults, although accounting for 1% of all adult solid tumor malignancies.[2] Rhabdomyosarcoma is an aggressive malignant soft-tissue sarcoma that develops from undifferentiated mesenchymal cells.[3]

It has a propensity for progressive disease early in its progression.[4]

RMS is divided into three subtypes: embryonal, alveolar, and pleomorphic RMS (PRMS). The most common type of RMS to occur in adults is pleomorphic rhabdomyosarcoma.[2] The most common primary sites are extremities, trunk wall, and genitourinary organs. It most typically appears in deep soft tissues of the extremities.[5,6]

It has been found that males have a little predisposition to RMS.[8]

The diagnosis is based on histomorphology, immunohistochemical, and molecular characteristics. Metastasis is often found at diagnosis, commonly in the lungs, lymph nodes, and bone marrow.[7] The reason for consistently worse outcomes remains mostly unknown. One explanation for the reduced adult survival rates is an increase in the occurrence of negative prognostic variables such as unfavorable primary locations and greater rates of regional and distant dissemination. Because of these factors, adults are already from the time of diagnosis assigned to high-risk and very high-risk groups with inferior prognoses according to EpSSG (The European paediatric soft tissue sarcoma study group). Given the low incidence of pleomorphic rhabdomyosarcoma (PRMS), diversity in clinical presentation, tumor features, and prognosis, there is not much evidence to guide the care of adults with RMS. Surgery, radiation therapy, and chemotherapy are all part of the multimodal treatment strategy. A case report of pleomorphic rhabdomyosarcoma of the right thigh in a 70-year-old male is presented here.

CASE REPORT

A 70-year-old male presented with a non-healing wound associated with swelling over the posterior and medial aspect of a right leg along with features of cellulitis with diffuse ooze from the wound. The swelling was tense, and tender with local rise of temperature with surrounding erythema and areas of blackish discoloration with slough. The patient had a history of fall was 5 months ago and was treated by a local doctor but symptoms of pain and swelling aggravated. X-ray was done suggesting an un-displaced fracture of the lower third fibula for which no intervention was done.

Pain and swelling over the lower and middle part of his right leg continued for over 3 months. At a peripheral hospital, the swelling was evaluated with an ultrasound which showed a large amount of fluid suspicious of hematoma secondary to doubtful A-V malformation. He underwent a procedure where blood clots were evacuated but further procedure was abandoned and the patient was transferred to our center.

The patient was immediately evaluated after assuring hemodynamic stability. CT Angiogram was done which was suggestive of a large 25.6×9.4×9.3 cm sized heterogeneously attenuating solid cystic lesion in the posterior compartment of the deep intermuscular plane of the entire right leg from just below knee up to 5 cm above the ankle. After appropriate consents-wound was explored. Intraoperatively soft gelatinous material with pus discharge and blood clots were evacuated. Entire muscles of the posterior compartment of the lower and middle third of the leg were found to be necrosed and replaced by gelatinous material.

Histopathology of soft tissue

High-grade pleomorphic sarcoma, showing myofibroblastic/myogenic differentiation. FNCLCC grade 3.

Immunohistochemistry of tumor cells

Diffuse positivity for SMA patchy moderate to strong NAC positivity for SAT82 whereas negative for Desmin and CD34.

Microscopy

A highly cellular tumor composed of markedly pleomorphic/anaplastic cells including several multinucleated giant cells with areas of stromal hyalinization in branching corded pattern, reminiscent of osteoid mitotic figures conspicuous up to 20/10 HFF including atypical forms.

Follow-up and outcome

A PET-CT scan was done to evaluate the response to the treatment. In this study, there was no evidence of abnormal hypermetabolism in the body on PET-CT. Post-operatively the wound healing process was excellent and after brief physiotherapeutic exercises, the patient started to walk with little to no impairment. The patient was advised further multimodality management for RMS but denied any further treatment. Post-operative recovery was uneventful with no recurrence for the first 6-8 months after which the patient did not follow up.

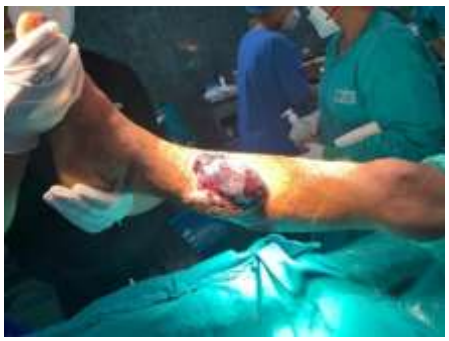

Figure 1: Pre-operative image of lesion.

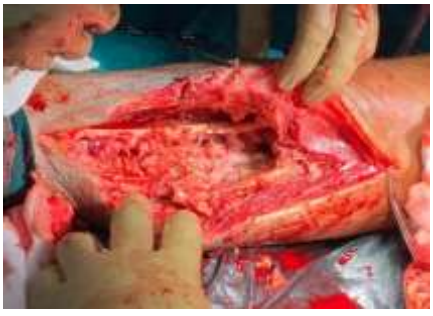

Figure 2: Intra-operative image after exploration.

Figure 3: Image of the resected specimen.

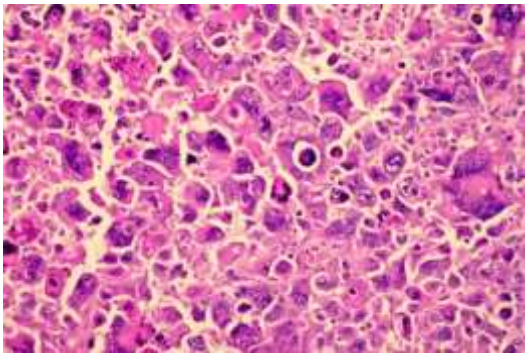

Figure 4: Histopathology image of the resected specimen.

Figure 5: Post-operative recovery of wound.

DISCUSSION

Adult rhabdomyosarcoma is a very rare case. Adult RMS differs from childhood RMS in terms of natural history, behavior, poor response to treatment, prognosis, and outcome.[5] The embryonal subtype is the most common, accounting for approximately 49% of all RMSs.[10] The alveolar subtype accounts for up to 30% of all RMSs.[10,11] In children and adolescents, embryonal and alveolar subtypes of RMS predominate. While in adults the pleomorphic type is the most common.[9] It predominantly occurs in the skeletal muscles of the thigh. It is histologically identical to a malignant fibrous histiocytoma, and the exact diagnosis is best accomplished using immunochemistry, as demonstrated in this case report.

Patients with pleomorphic RMS tend to present with a rapidly growing swelling in the extremities over a period of time. This kind of RMS often occurs between the fourth and sixth decades of life.[11,12] This is similar to our case, in which a 70-year-old guy had a growing tumor in his lower extremities over the course of six months. Our case is likewise typical of adult RMS, since the expanding mass was discovered in the most common location: the deep muscles of the leg. This is distinct from RMS cases in children, which can develop anywhere in the body.[13] Because of the small number of adult RMS cases, there is a scarcity of knowledge on treatment and prognosis. Adult RMS treatment strategies are derived from the intergroup rhabdomyosarcoma study group (IRS) or children's oncology group study (COG). It is vital to highlight that the IRS and COG-STS protocols are intended for RMS cases in children.
Previous studies have noted that adult RMS arises majorly in the extremities.[3,6] Nonetheless, the initial tumor site is still clinically significant, because tumors in the retroperitoneum, for example, can grow fairly big before causing signs and symptoms.[14] Metastases, tumor size, tumor resectability, and patient age at presentation are all prognostic variables. In both univariate and multivariate analyses of a case series from Memorial Sloan-Kettering, the relative number of extremities pleomorphic tumors rose with age, and survival declined with increasing age.[15] In that study, increasing age was not related with worse survival, regardless of whether it was treated as a continuous or dichotomous variable.[15]

There are still research on adult rhabdomyosarcoma that show inconsistent results regarding age as a predictor of survival.[16] Little et al and Esnaola et al found that increasing age was not related with worse survival, in contrast to recent studies by Dumont et al and Gerber et al, which found that age was relevant for overall survival in nonmetastatic adult patients.[4,16-18] The causes for this age impact are unknown, however it might be due to changes in histological subtype distribution or reduced adult patient tolerance for intense chemotherapy.[17] Previously, tumor size was linked to poor survival.[19]
An FNAC can be done to ascertain the diagnosis, especially when there is a strong suspicion of sarcoma as revealed by clinical features on imaging. In the case presented to our hospital, FNAC was not done as there was no suspicion of a sarcomatous change based on clinical and imaging findings.

If there is a suspicion of sarcoma, a frozen section can be done intra-operatively followed by wide excision based on oncological principles. However, the incidence of such a situation is extremely low, as in our case.
The gross pathology frequently reveals significant necrosis with localized hemorrhages as in our case showed similar findings and for final diagnosis, it requires

immunohistochemical and/or ultrastructural examination of sarcoma-type differentiation. Imaging is an essential component of diagnostics. Sarcoma on ultrasound usually shows a heterogeneous well-defined irregular mass of low to medium echogenicity.

CT scans can reveal soft tissue density and some enhancement with contrast. MRI is the most frequently used radiological modality, by virtue of the fact that it has extremely high soft tissue resolution. This enables a detailed evaluation of the distribution and depth of the mass.

Modern therapies like as chemotherapy and radiation are critical for initial cyto-reduction and metastatic disease elimination. Surgical techniques are indicated for low-risk tumors and might be explored as a therapeutic option for high-risk tumors following radiation or chemotherapy to manage the microscopic local residual illness. The specific treatment regimen is determined by the predicted risk of illness recurrence, prognostic variables, and a process known as risk-adapted therapy. Due to the unusual presentation of RMS, full excision of localized disease with function and limb-sparing strategy is appropriate as was done in this instance. In challenging cases when surgery is not an option, an initial diagnostic biopsy is recommended, followed by Neoadjuvant chemotherapy, followed by local treatment (i.e., radiation).[20]

Several studies have employed cyclophosphamide or ifosfamide, doxorubicin, and/or dactinomycin with or without vincristine or additional medicines such as cisplatin, carboplatin, and etoposide in multidrug chemotherapy regimens.

The 10-year overall, disease-free, and metastasis-free survival rates for 82 adult patients with RMS treated with chemotherapy regimens comprising vincristine and cyclophosphamide with dactinomycin or doxorubicin were 47%, 45%, and 59%, respectively, in retrospective research at MD Anderson.[17]

Another big retrospective research looked at 171 RMS patients, 37 of whom had PRMS. Surgery was the most common treatment for PRMS patients (74%), followed by radiation and chemotherapy. The average duration of follow-up was 28 months. The event-free survival rate and overall survival rate after five years are 29.9% and 53.4%, respectively.[2]

CONCLUSION

This case report discusses an exceptionally rarely seen cancer in an adult patient with rhabdomyosarcoma of the lower extremity which was misdiagnosed initially because of the atypical presentation. Therefore, in such cases, the delayed post-traumatic mass of extremities should be evaluated with a high index of suspicion of soft tissue sarcoma (STS). In case of doubt, either a preoperative biopsy or an intraoperative frozen section is a safer practice. Wide local excision with multi-multimodality treatment at a tertiary care center is the best option for such tumors

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REFERENCES

1. Chigurupati R, Alfatooni A, Myall RW, Hawkins D, Oda D. Orofacial rhabdomyosarcoma in neonates and young children: a review of literature and management of four cases. Oral Oncol. 2002;38(5):508-15.
2. Ferrari A, Dileo P, Casanova M, Bertulli R, Meazza C, Gandola L, et al. Rhabdomyosarcoma in adults. A retrospective analysis of 171 patients treated at a single institution. Cancer. 2003;98(3):571-80.
3. Ferrari A, Dileo P, Casanova M, Bertulli R, Meazza C, Gandola L, et al. Rhabdomyosarcoma in adults. A retrospective analysis of 171 patients treated at a single institution. Cancer. 2003;98(3):571-80.
4. Esnaola NF, Rubin BP, Baldini EH, Vasudevan N, Demetri GD, Fletcher CD, et al. Response to chemotherapy and predictors of survival in adult rhabdomyosarcoma. Ann Surg. 2001;234(2):215-23.
5. Khosla D, Sapkota S, Kapoor R, Kumar R, Sharma SC. Adult rhabdomyosarcoma: Clinical presentation, treatment, and outcome. J Cancer Res Ther. 2015;11(4):830-4.
6. Sultan I, Qaddoumi I, Yaser S, Rodriguez-Galindo C, Ferrari A. Comparing adult and pediatric rhabdomyosarcoma in the surveillance, epidemiology and end results program, 1973 to 2005: an analysis of 2,600 patients. J Clin Oncol. 2009;27(20):3391-7.
7. Simon JH, Paulino AC, Ritchie JM, Mayr NA, Buatti JM. Presentation, prognostic factors and patterns of failure in adult rhabdomyosarcoma. Sarcoma. 2003;7(1):1-7.
8. Wexler L, Helman L. Rhabdomyosarcoma and the undifferentiated sarcomas. In: Pizzo P, Poplack D, eds. Principles and Practice of Pediatric Oncology. 3rd ed. Philadelphia: Lippincott-Raven; 1997: 799-829.
9. Ghavimi F, Exelby PR, D'Angio GJ, Cham W, Lieberman PH, Tan C, Miké V, Murphy ML. Multidisciplinary treatment of embryonal rhabdomyosarcoma in children. Cancer. 1975;35(3):677-86.
10. McCarville MB, Spunt SL, Pappo AS. Rhabdomyosarcoma in pediatric patients: the good, the bad, and the unusual. AJR Am J Roentgenol. 2001;176(6):1563-9.
11. Brennan M, Casper E, Harrison L. Soft tissue sarcoma. In: DeVita V, eds. Cancer: principles and practice of oncology. 5th ed. Philadelphia: Lippincott-Raven; 1997: 1738-1788.

12. Jones TR, Norton MS, Johnstone PA, Kane E, Shin AY. Pleomorphic rhabdomyosarcoma in an adult forearm: a case report. J Hand Surg Am. 2002;27(1):154-9.
13. Parham DM, Barr FG. Classification of rhabdomyosarcoma and its molecular basis. Adv Anat Pathol. 2013;20(6):387-97.
14. Liu YT, Wang CW, Hong RL, Kuo SH. Prognostic Factors and Treatment Outcomes of Adult Patients With Rhabdomyosarcoma After Multimodality Treatment. Anticancer Res. 2019;39(3):1355-64.
15. Quaglia MP, Heller G, Ghavimi F, Casper ES, Vlamis V, Hajdu S, et al. The effect of age at diagnosis on outcome in rhabdomyosarcoma. Cancer. 1994;73(1):109-17.
16. Gerber NK, Wexler LH, Singer S, Alektiar KM, Keohan ML, Shi W, et al. Adult rhabdomyosarcoma survival improved with treatment on multimodality protocols. Int J Radiat Oncol Biol Phys. 2013;86(1):58-63.
17. Little DJ, Ballo MT, Zagars GK, Pisters PW, Patel SR, El-Naggar AK, et al. Adult rhabdomyosarcoma: outcome following multimodality treatment. Cancer. 2002;95(2):377-88.
18. Dumont SN, Araujo DM, Munsell MF, Salganick JA, Dumont AG, Raymond KA, et al. Management and outcome of 239 adolescent and adult rhabdomyosarcoma patients. Cancer Med. 2013;2(4):553-63.
19. Raney RB, Maurer HM, Anderson JR, Andrassy RJ, Donaldson SS, Qualman SJ, et al. The Intergroup Rhabdomyosarcoma Study Group (IRSG): Major Lessons From the IRS-I Through IRS-IV Studies as Background for the Current IRS-V Treatment Protocols. Sarcoma. 2001;5(1):9-15.
20. Cecchetto G, Bisogno G, De Corti F, Dall'Igna P, Inserra A, Ferrari A, et al. Biopsy or debulking surgery as initial surgery for locally advanced rhabdomyosarcomas in children?: the experience of the Italian Cooperative Group studies. Cancer. 2007;110(11):2561-7.

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