Case Report

Primary Cutaneous Alveolar Rhabdomyosarcoma, an Uncommon Entity

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ABSTRACT

Although secondary rhabdomyosarcoma (RMS) of the skin is not uncommon, the primary type is rare. RMS arising on subcutaneous tissue in an adult is sorely uncommon. It usually affects the skin of the face. Among the four histologic subtypes, the alveolar variant is the least frequently presented variant. Here, the authors introduce a patient with primary cutaneous alveolar RMS of the scalp. The tumor manifested as an immense plaque-like lesion confined to the scalp. The diagnosis was made by histological and immunohistochemical features of the biopsied specimen. Immunohistochemistry stains revealed that the tumor cells were positive for Desmin and negative for HMB45 and CD34. Surgery combined with adjuvant chemotherapy seems to be the best treatment. If complete resection of the lesion is not possible, radiotherapy is recommended. This case demonstrates that primary cutaneous RMS should be considered in the assessment of peculiar malignant tumors of the skin, and proper ancillary studies will be helpful.

KEYWORDS: *Adult, cutaneous, primary, rhabdomyosarcoma*

INTRODUCTION

Rhabdomyosarcoma (RMS) enumerates for almost 5%–8% of pediatric neoplasms but <3% of cases arise in adults. It is a virulent mesenchymal tumor with the capability of striated muscle differentiation. RMS is divided into three basic variants according to histological manifestations: Embryonal RMS (containing the spindle cell and botryoid subtypes), alveolar RMS, and pleomorphic RMS.[1] Subsidiary lower popular variants have also been explained, containing sclerosing and epithelioid RMSs.[2,3] RMSs do not have the specifications for the mentioned variants alluded to as RMS not otherwise specified. The alveolar rhabdomyosarcoma (ARMS) is the most offensive subtype of RMS.[4] The majority of RMSs arise in profound sites. Indeed a primary ARMS of the skin is rare. Of 682 RMS s gathered at the MD Anderson center, only 0.7% of the cases were related to the skin.[5] Here, we introduce a case of cutaneous ARMS that manifested in a strange manner and made a diagnostic dilemma. Cutaneous RMS should stimulate an inspection for cryptic limbs or splanchnic neoplasm, which was not extant in the presented patient.

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CASE REPORT

A 50-year-old male referred to our dermatology ward with a dolorous indurated plaque on the scalp that he had for 8 months [Figure 1]. The lesion had grown rapidly. There were no accompanying systemic symptoms. General physical examination was unremarkable and lymphadenopathies were not detected. Scalp examination revealed a large and indurated plaque which extended from the forehead to the occipital area. The lesion was ulcerated and discharge was evident. It was very foul-smelling. Following biopsy histological sections revealed an ulcerated neoplasm sub utmost of the dermis [Figure 2] and extending to the hypodermis. The tumor was a compound of relatively allotropic cells with scant to ample eosinophilic cytoplasm, large pleomorphic nuclei, and eminent nucleoli. The tumor

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cells were arranged in the alveolar pattern. A few cells showed a rhabdoid feature. Cross striation was not found. A large number of mitoses were seen and atypical ones were found. Immunohistochemistry (IHC) stains revealed that the tumor cells were positive for Desmin and negative for HMB45 and CD34 [Figure 3]. The patient was a foreign national and due to his financial constraint, no further staining was performed. According to the histological and ancillary findings, the diagnosis of ARMS was confirmed. The patient was admitted to the hospital, antibiotics (regarding secondary bacterial infection) were given, and oncology counseling was requested. However, the patient refused treatment and was discharged with personal consent. Unfortunately, the patient was lost for follow-up.

Discussion

Although secondary RMS of the skin is not uncommon, the primary type is rare. Primary cutaneous ARMS enumerates <1% of RMS. It usually affects the skin of the face.[6] It is claimed that in comparison with the embryonal type, ARMS has a more offensive course and its prognosis is poorer.[7] Schmidt *et al*.,[6] emphasized that the diagnosis of ARMS should be made only if there is an alveolar pattern. There is a bimodal age distribution in primary cutaneous RMS and it mostly affects men, a phenomenon that is also seen in deep soft-tissue type. The pathogenesis of this tumor is obscure. The occurrence of the tumor in areas of the body that are exposed to sunlight, such as the face and scalp, indicates that sunlight may be involved in the etiology of RMS. In the differential diagnosis of RMS, there are two groups of tumors, including tumors with small round cells and tumors with pleomorphic rhabdoid features. Thus, primary and metastatic neuroendocrine tumors, hematologic malignancies, Ewing's sarcoma/peripheral neuroectodermal tumor (PNET) family and neuroblastoma (especially in pediatric patients), eccrine carcinoma, malignant rhabdoid tumor of soft part, proximal type of epithelioid sarcoma, poorly differentiated myoepithelial carcinomas, and epithelioid angiosarcoma should be considered. Other diagnoses are ruled out based on the patient's age, the location of the tumor, and IHC findings.[1,8,9] In general, all of the mentioned tumors, except rare cases of Ewing's sarcoma/sPNET family, are negative for muscular markers. A literature review shows that all cases of cutaneous RMSs are positive for desmin, while the vast majority of them are positive for myogenin and myoD1.[10] Therefore, desmin can be used as the most sensitive marker in preliminary studies, and then myogenin or myoD1 can be used to confirm striated muscle differentiation. As it turns out, the diagnosis is established by histologic and IHC findings. In a challenging case, an electron microscope or fluorescence *in situ* hybridization is needed. Due to the rarity of the disease and limited follow-up of the patients, the prognosis is unclear. However, one study showed that prognosis depends on factors such as the location, size, site, histology, clinical stage of the lesion, and the age of the patient at the time of diagnosis.[11] Surgery combined with adjuvant chemotherapy seems to be the best treatment. If complete resection of the lesion is not possible, radiotherapy is recommended.

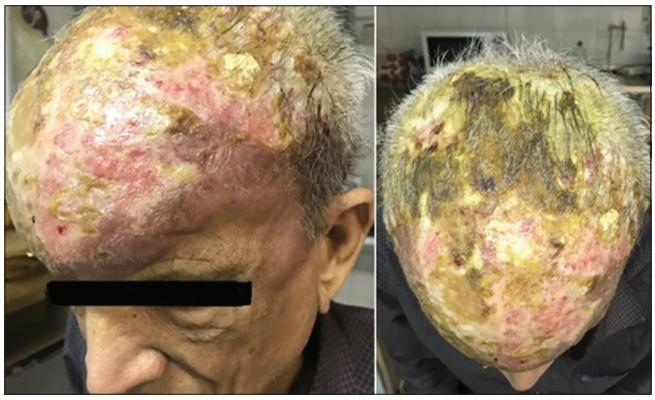

Figure 1: Shows indurated plaque on the scalp

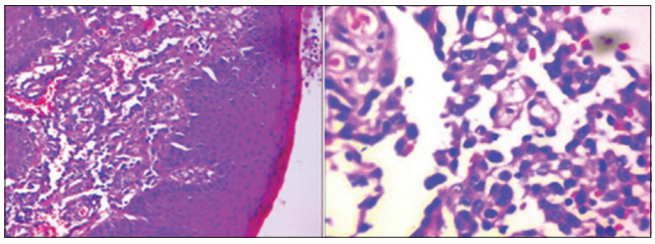

Figure 2: Sections reveals that the tumor is a compound of relatively pleomorphic cells with scant to ample eosinophilic cytoplasm arranged with the alveolar pattern

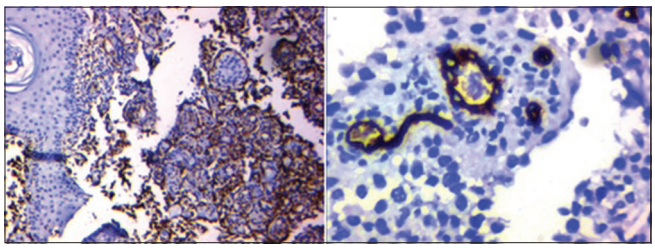

Figure 3: Tumor cells were positive for desmin and negative for CD34

Conclusion

Although primary cutaneous RMS is a rare entity, it should not be overlooked in the differential diagnosis of tumors with rhabdoid or small cell morphology. In the vast majority of cases, diagnosis is not possible without the IHC technique. The most useful markers for diagnosis are desmin, myogenin, and myoD1. Proper

diagnosis in these patients is essential to ensure correct and adequate therapy.

Authors' Contribution

All the authors contributed in the preparation of this research and article.

Declaration of patient consent

Patient's consent was obtained.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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