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Metastatic Rhabdoid Variant of Malignant Melanoma of the Inguinal Lymphadenopathy with Unknown Primary Site – Case Report

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Abstract

Background: *Metastatic malignant melanoma is a very rare presentation and is known for cutaneous, nodal, visceral, bone and brain metastasis. It comprises of metastatic melanoma of unknown primary (MUP) and metastatic melanoma of a known primary (MKP). According to literature, MUP patients have a better prognosis than MKP patients because of its spontaneous regression of the primary site but the survival of MUP versus MKP, however is not consistent due to its heterogenous group. In this report, for this MUP type having some rare variant histology named rhabdoid which is also an aggressive and poor prognosis altogether leads to overall outcome was dismal.*

Case Report

We report a 68- year-old female who presented with a progressively increased swelling in the right sided groin region. On evaluation we could not detect the primary lesion. Surgery was done and biopsy showed metastatic malignant melanoma and also advised about adjuvant immunotherapy but the patient refuses to take due to the financial issues. After 5 months of follow up, she had recurrence in the right thigh, underwent excision which showed satellite lesion with same histology had high risk features suggested adjuvant radiation. Meanwhile she developed multiple lung metastasis, after discussing with tumour board we withhold the plan of radiation treatment and started palliative chemo therapy in view of disseminated spread. After 6 cycles of chemotherapy, she had recurrence in the opposite site, unfortunately her condition was deteriorated and succumbed her life. This shows that this rare variant of melanoma had very poor prognosis in spite of treatment that leads to impaired survival due to its aggressive nature of disease.

Introduction

In melanoma, skin is the most common site (56%) but 5% of patients may present with metastatic melanoma[1]. Of which, 80% are loco-regional mostly by lymphatic spread and remaining 20% are distant metastasis[2]. The typical malignant rhabdoid tumor is a paediatric malignancy, rhabdomyosarcomatous variant of Wilms tumor of kidney first described by Haas et al[3]. The presence of extra-renal malignant rhabdoid tumors is also seen in soft tissue as well as in various organs such as the brain, lung, liver, colon, ovary, uterus, and skin[4]. The presence of rhabdoid features in melanoma is not uncommon in metastatic melanoma but it is quite rare in primary lesions with only a few cases reported[5-7]. So, it comprises of metastatic melanoma of unknown primary (MUP) and metastatic melanoma of a known primary (MKP). The prognostic significance of the rhabdoid phenotype in melanoma is unknown, probably because those with metastatic melanoma already face a poor prognosis[6]. As this is a rare variant of poor prognosis, we made a case report of this to know the impact of survival.

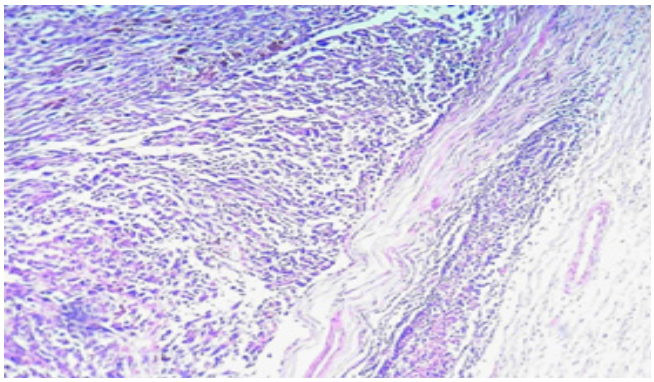
Methods and Materials

A 68- year-old female who presented with a progressively slowly increased swelling in the right sided groin region since 6 months associated with mild pain without any comorbidities. No other significant history to rule out the primary origin. On physical examination, ECOG score -2, no generalised lymphadenopathy except the localised right sided inguinal mass which was localised warmth, non-tender multiple matted firm of size 8cmx6cm, non-fluctuant with intact overlying skin with no redness or discharging tract. Her anogenital and pelvic examination showed normal. On skin examination, one blackish nodule presents over the forehead region since birth. Blood work

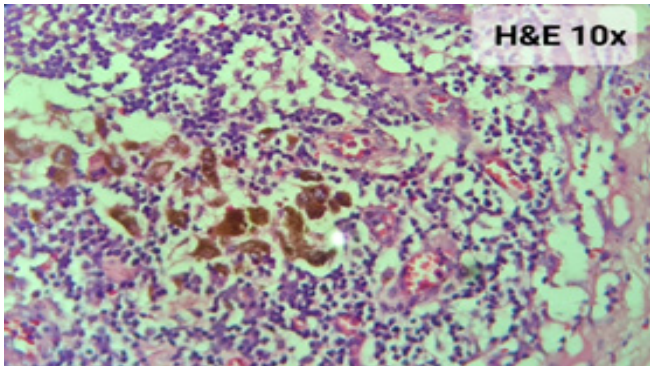
was unremarkable. MRI scan of abdomen and pelvis with plain and contrast showed multiple heterogenous metastatic necrotic right inguinal lymph nodes largest measuring 8.6x6.6cm.

Chest X-ray was unremarkable. PET-CT scan showed increased FDG uptake in multiple metastatic enlarged enhancing right inguinal and few right iliac lymph nodes, largest measuring 6.2x5cm, max SUV 25. Colonoscopy and Cystoscopy showed normal mucosa (did not show any evidence for a primary lesion). Biopsy showed metastatic deposits of rhabdoid melanoma, IHC confirmed and express S100, HMB45, Melan-A, Vimentin with ki-67 is up to 40% and CD45 is negative. Surgery was done with complete excision of ilioinguinal nodal dissection on right side under general anaesthesia. Post-op HPE report showed metastatic deposit of malignant melanoma of largest node size 6x3x2cm without extra nodal extension.

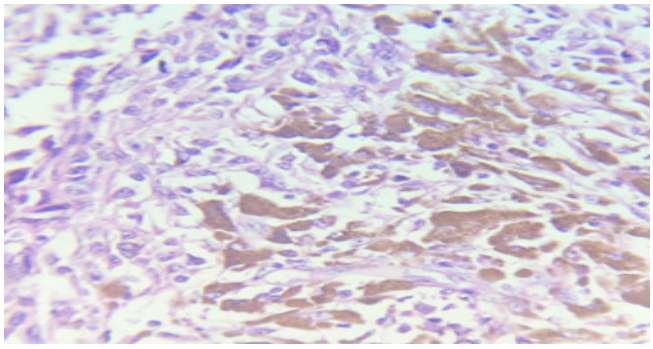
A) H&E 10X



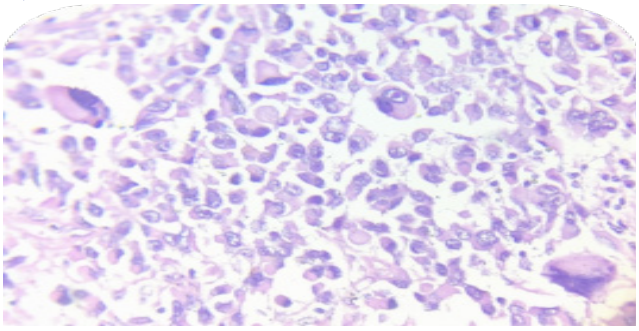
B) H&E 10X



C) H&E 40X



D) H&E 40X



Microscopy: Malignant Melanoma (Rhabdoid variant).

A & B: Metastatic deposit in lymph node.

C: Melanoma cell showing intracytoplasmic melanin pigment.

D: Rhabdoid type giant cells with prominent eosinophilic nucleoli.

Advised immunotherapy but patient is not willing to take in view of financial constraints. On close follow up, after 5 months she got right thigh swelling and FNAC shows adnexal neoplasm, treated with wide local excision. Her postop HPE report satellite lesion shows recurrent rhabdoid melanoma with high grade, intratumoral lymphocytic infiltration, extent upto subcutaneous tissue. We planned for adjuvant radiation, hypofractionation with a dose of 48Gy/20# having 2.4Gy per fraction.

Before starting radiation, she had intractable cough, on chest X-Ray shows multiple lung metstasis. On multidisciplinary discussion, we abandoded the radiation in view of distant metastasis, counselled regarding the disease status. We have started palliative chemotherapy with Injection Dacarbazine and Injection Cisplatin under health schemes as patient is not affordable for immunotherapy. After completion of 6 cycles of chemotherapy, she had generalized fatigue, not taking orally and managed symptomatically. Unfortunately, her general condition deteriorated and also she got recurrence in the left inguinal region and expired.

Results

After careful examination and evaluation, the case was then diagnosed as a metastatic melanoma of unknown primary with Rhabdoid variant showed as more aggressive variant with recurrence in spite of surgery, having high risk features and also lung metastasis received palliative chemotherapy as patient is not affordable for immunotherapy. Again, she had recurrence in the opposite site and succumbed her life with a span of 11 months after diagnosis.

Discussion

Rhabdoid melanoma was first described by Bittesini et al. in 1992. Since then, over 40 cases of metastatic or recurrent rhabdoid melanoma have been reported as per literature[8]. Our

case is a metastatic rhabdoid variant of malignant melanoma of the inguinal lymphadenopathy with unknown primary site having recurrent and metastatic features.

The typical malignant rhabdoid tumor of wilms tumor is a paediatric malignancy occurring in the kidney, first described by Haas et al3. Presently, the malignant rhabdoid tumor is recognized as a separate entity due to a lack of rhabdomyoblastic ultrastructural features. Since first described in kidney, later on cases reported in extra-renal malignant rhabdoid tumors like soft tissue as well as in various organs such as the brain, lung, liver, colon, ovary, uterus, and skin[4]. In our case, these features are seen in ilio-inguinal lymph nodal metastatic rhabdoid variant of malignant melanoma metastasis with unknown primary origin as well as recurrent after surgery with satellite nodules and metastasis to lungs also.

Furthermore, Chung et al[9] analyzed the clinico-pathological features of 31 specimens from 29 cases of metastatic rhabdoid melanoma and reported that 61% of the specimens were exclusively amelanotic and included 15 tumors with purely rhabdoid features. In both primary and metastatic cases, rhabdoid melanoma also tended to be amelanotic[10]. Although the mechanisms are still unknown, melanoma cells may lack the ability to produce melanin in the process of rhabdoid transdifferentiation. In this case, HMB-45 staining was clearly positive and made the diagnosis of melanoma easier. In the review of metastatic rhabdoid melanoma by Chung et al, 41% of specimens were positive for HMB-45.

These type of rhabdoid melanomas to follow an aggressive course because they behave like renal and extra-renal rhabdoid tumours. However, based on the previous review, rhabdoid melanomas appear to behave no more aggressively than common melanomas[11]. A confounding factor in our case is the development of lung metastasis approximately within six months after the initial diagnosis. Because these are very rare and the number of reported cases is so small, additional research is needed to determine the prognosis of this disease with some degree of accuracy.

Rhabdoid melanoma is a rare variant of melanoma, and many pathological and biological problems have yet to be resolved. Recognition of this rare entity is important to avoid confusion both in diagnosis and in the subsequent direction of therapy.

Conclusion

Metastatic rhabdoid variant of malignant melanoma of the inguinal lymphadenopathy with unknown primary site is a rare aggressive tumor and is associated with a poor outcome in spite of aggressive treatment modalities. Due to its heterogeneous nature of tumor, predicting poor survival and thorough counselling regarding disease prognosis after diagnosis and also needs future direction of therapy to improve survival.

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