Pinna with synchronous chronic lymphocytic leukemia (CLL)/small lymphocytic lymphoma (SLL) - A rare presentation of Amelanotic melanoma (Desmoplastic variant)

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ABSTRACT

Malignant melanoma is a neoplasm derived from cutaneous melanocytes, that frequently occurs in the elderly and, may metastasize loco-regionally or distally. B-cell chronic lymphocytic leukemia/small lymphocytic B-cell lymphoma (CLL/B-SLL) is a neoplasm of B-cell lymphocytes that also occurs frequently in the older population as an asymptomatic elevation of the white blood cell count (WBC) and has an overall good prognosis.

There is minimal data on simultaneous occurrence of CLL and melanoma. In this report, we describe the unusual case of a 74 years old man who was diagnosed as desmoplastic amelanotic melanoma, pinna in an unsuspected second clinical setting, with incidental synchronous detection of chronic lymphocytic leukemia (CLL)/small lymphocytic lymphoma (SLL) , highlighting the need for workup of such chance associations. Dense inflammatory granulation tissue in lesions of pinna need to be followed up, so as not to miss desmoplastic variant of amelanotic melanoma as in this case.

Keywords- amelanotic melanoma (AM), desmoplastic variant, chronic lymphocytic leukemia(CLL), small lymphocytic lymphoma(SLL),lymph node, standardized incidence ratio (SIR)

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Conflict of interest: None

INTRODUCTION

Malignant melanoma is a neoplasm derived from cutaneous melanocytes, seen more frequently in the elderly and, may metastasize loco-regionally or distally. An increased risk of CLL, second primary melanoma, cancers of the prostate, soft tissue etc is being reported in cases of malignant melanoma during follow up. [1] However synchronous occurrence of such tumors in the same is even less common.[2] We present a rare case of amelanotic

melanoma AM (desmoplastic variant) with synchronously diagnosed CLL/SLL in a patient presenting with ulcerated lesion of the pinna, with a cervical lymphnode.

CASE REPORT

A 74 years old man resident of Dehradun presented with ulcerated lesion on the right ear, with history of frequent bleeding. History of an incision biopsy done 2 years ago for a pinpoint lesion, on the same site was given. The report mentioned the presence of an atypical cell in a background of inflammatory granulation tissue and an advice for a follow up biopsy.

On examination at the time of presentation, it was a 1.5 cm sized, ulcerated lesion on the anterior helix. There were no systemic complaints. The pinna was partially excised with the clinical diagnosis of pyogenic granuloma/ chondrodermatitis, with reconconstruction surgery. The solitary upper deep cervical lymph node detected at surgery was sent for histopathological examination.

The specimen received, showed an ulcerated lesion in the helix anteriorly with focal cartilage destruction (Figure1)

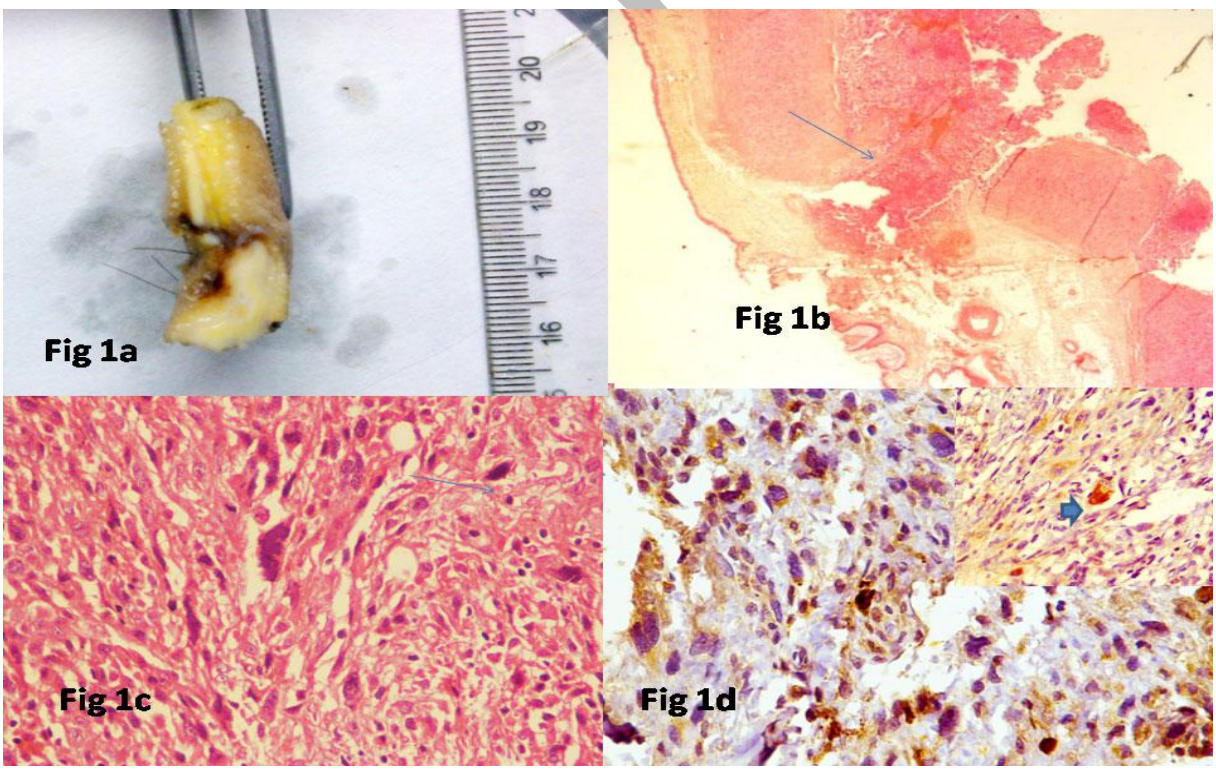

Figure1a. Cut section of the pinna showing ulcerated helix and cartilage destruction

Figure1b. Microphotograph showing tumour infiltrating across the cartilage (H&E 2x).

Figure1c.Microphotograph showing predominantly spindle cell component, with marked pleomorphism [arrow] (H&E 40x)

Figure1d. Microphotograph showing S-100 positivity [inset shows HMB-45 positivity] (DBS, RTU), HMB 45 (Dako, RTU)

Microscopic sections showed localized proliferation of ovoid to spindle shaped cells with abundant tapering cytoplasm with intervening clusters of few large epithelioid like cells atypical mitosis admixed with fair number of multinucleated histiocytic cells in rich vascular network. These cells were arranged in bundles and fascicles. No intracellular pigment was noted. The overlying epidermis was ulcerated at most places with underlying mixed inflammatory infiltrate. This lesion was seen infiltrating across the cartilage focally. All peripheral resected margins were free of tumor. On IHC, both the spindle and epithelioid cells were vimentin (DBS, RTU), HMB 45 (Dako, RTU), EMA and S-100 (Biocare,RTU) positive (Figure 1). These findings were suggestive of desmoplastic variant of amelanotic spindle cell melanoma with mixed pattern (Clarke's IV/V).

Cervical lymph node measured 2.5 x 2.0 x 1.0 cm with grey-white firm smooth cut surface. The sections showed near total effacement of lymphoid architecture showing proliferation of lymphoid cells, larger than mature lymphocytes with angulated nuclear borders with coarse chromatin and scanty cytoplasm with infrequent mitoses, and formation of a few proliferation centres and perinodal spread. (Fig.2) CD20 stain showed diffuse positivity. Hemogram showed Total leucocyte count (TLC) 17,770 cells per mm3 with 70% lymphocytes and a fair number of smudge cells. Serum protein electrophoresis was normal. Flow cytometry was advised .It showed CD5 and CD38 negative, CD23 dim to moderate positivity in 87.4% cells. CD10 and CD20 were dimly positive in 99.8% and 86.4% cells respectively. A final diagnosis of malignant amelanotic melanoma, spindle cell /desmoplastic variant, with synchronous

atypical SLL/CLL was made and was referred to higher centre for further management.

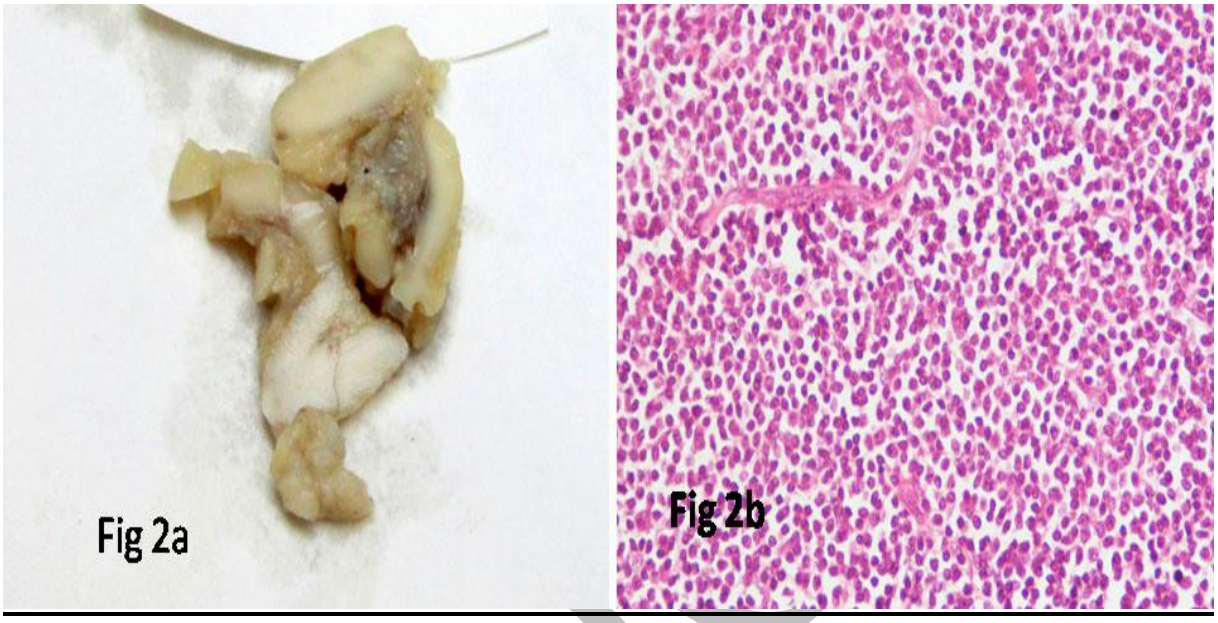

Fig.2a. Gross appearance of cervical lymph node, grey white smooth homogenous cutsurface

Fig.2b. Microphotograph showing diffuse effacement by monotonous lymphoid cells (H&E 10x)

DISCUSSION

Patients with malignant melanoma, have an increased risk for developing metachronous secondary malignancies, especially CLL/B-SLL. Patients with MM had a standardized incidence ratio (SIR), of 1.9 to 2.3 for later development of CLL/B-SLL with higher risk in males and people older than 50 years.[1,3]

Possible factors attributed for increased rates of concurrent MM with CLL/B-SLL are, increased clinical surveillance, shared genetic abnormalities on chromosome 9p21q, (changes in the INK4 locus with p16, an essential tumor suppressor protein) and environmental factors e.g exposure to chemicals i.e pesticides and hair dyes as well as exposure to the 280-315 nm wavelengths of sunlight (UV). UV is thought to decrease skin hapten reactivity, inhibit proper antigen presentations, and reduce T-cell activity,

resulting in a greater risk of developing lymphoid and cutaneous malignancies. [4]

An increased risk of both MM and CLL has been reported in higher socioeconomic, white-collar workers, possibly due to increased recreational sun exposure.[5] The external ear represents a site with high UV exposure and thin skin overlying the cartilage. This may make the external ear more prone to develop MM.[4,5] Use of hair dye and UV light could be a possible cause in our case, who was a resident of hills of dehradun.

Concurrent diagnosis of melanoma and CLL in 62 patients with 25% having melanoma deposit in lymphnode has been reported.[6] But in this case lymph-node detected at the time of surgery showed only SLL without any melanoma deposit.
20% of all melanomas arise in head and neck, and 7-15% cases occur in the vicinity of the ear, most often the helix with Clarke's level IV or V in significant number.[7,8]

Early diagnosis is often challenging in desmoplastic melanoma because it is often amelanotic and has a predominant dermal component, and variation in the amount of intratumoral cellularity and fibrosis as in this case. The previous slides were reviewed and showed presence of a few atypical cells in background of fibroblast rich inflammatory granulation tissue. Some tumors may even show mixed features of both desmoplastic melanoma and nondesmoplastic melanoma. Hence, desmoplastic melanoma is now being sub classified into pure and mixed types increasing the need for IHC as in this case .[9] AM is more common on face and ears than the trunk and ulceration is also common, constituting (8%).[10]

Desmoplastic amelanotic melanoma, however, tend to be HMB-45 negative although universally S100 positive, adding to the diagnostic difficulty, as in this case.[11] This case also showed more intense and diffuse positivity for S-100 as compared to HMB-45.

CONCLUSION

Ulcerating lesions on ear can be deceptive clinically and diagnosis becomes difficult in cases of amelanotic spindle/desmoplastic cell lesion in the absence of IHC. Any presence of atypical cell needs to be followed by repeat confirmatory biopsy. Mild lymphocytosis needs to be considered with concern and such cases need to be investigated fully with

IHC/ flow cytometry especially in the setting of melanoma.

List of Abbreviations

AM amelanotic melanoma
CLL chronic lymphocytic leukemia
IHC immunohistochemistry
MM malignant melanoma
SLL small lymphocytic lymphoma
UV ultraviolet

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of this written consent is available for review by the Editor-in-Chief of this journal.

Authors' contributions

NS scrutinized the microscopy findings, conceptualized the case report and revised the basic manuscript.

PS performed the literature search and wrote the basic manuscript.

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