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Case Report

Sebaceous cell carcinoma of the scalp in a young male: A rare case report

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ABSTRACT

Sebaceous carcinoma is a rare locally aggressive neoplasm of the sebaceous gland, which occurs frequently in elderly males. It constitutes 0.2 to 4.6% of all the malignant cutaneous tumours and occurs commonly in the periocular area. Scalp is a rare site for this tumor. Here we report a case of sebaceous carcinoma in a young 28 years old male patient who presented with firm to hard exophytic growth over right posterior occipital region with enlarged suboccipital and post auricular lymph nodes. Wide local excision was done and a diagnosis of sebaceous carcinoma of the scalp with lymph node metastasis was rendered after histopathological and Immunohistochemical examination.

1. Introduction

Sebaceous carcinoma (SC) is a rare neoplasm of the sebaceous gland.[1] Though an increased frequency is seen in the Asian population, its frequency in males and adults is less frequent.[2] Sebaceous carcinoma is found to occur commonly in the periocular area than in other sites, its occurrence over the scalp is less frequently reported.[3] It constitutes 0.2 to 4.6% of all the malignant cutaneous tumours and has a predilection in the elderly population.[4] The majority of sebaceous carcinomas arise from de novo mutations but those associated with Muir-Torre syndrome have a loss of mismatch repair gene expression and exhibit microsatellite instability, while those arising sporadically do not exhibit loss of mismatch repair or microsatellite instability.[5] Final diagnoses rely on histopathology and clinically, it may mimic other neoplasms like squamous or basal cell carcinoma.[3] Thus sebaceous cell carcinoma poses a diagnostic challenge and is usually associated with a poorer prognosis.[6]

2. Case Report

A 28 years old male presented with a lesion on the scalp for one year which had started growing rapidly for which he was operated 2 months ago in a private hospital and a diagnosis of sebaceous cell carcinoma was made histologically. However, the lesion reappeared after one month over the right posterior occipital region. On physical examination, a firm to hard cystic swelling was noted having an irregular, exophytic growth pattern with ulcerated appearance over right posterior occipital region. Simultaneously enlarged suboccipital and post auricular lymph nodes were also noted. Wide local excision of the lesion was done. Excised skin with the cyst, the suboccipital and posterior auricular lymph nodes, the excised periosteum and excised Calvarium bone were received in our department for the histopathological examination. The gross examination revealed a single, greyish-brown, skin covered tissue piece showing two

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nodular growth protuberances.(Figure 1 A) The larger Protuberance measured 2.8x2.5 cm in size, while the smaller protuberance measured 2 X1.8 cm. The external surface showed ulcero-proliferative and nodular growth, while on the cut section greyish-white areas with spots of haemorrhages were noted.(Figure 1 B) Tissue labelled as post auricular lymph node measured 2.5x1.8x1.5cm, which on cut section showed homogenous white areas. Tissue labelled as suboccipital lymph node measured 1.3x0.9x0.5cm, which on cut section showed homogenous white areas with foci of necrosis. Two membranous tissue pieces labelled as periosteum measuring 6x5cm and 3x1cm were also received. Several sections were taken and slides were reported. Microscopically the sections from the main tissue showed sheets, nests and lobules of atypical cells separated by fibrous septa. These atypical cells show marked pleomorphism, an enlarged nucleus with increased N:C ratio, vesicular nuclei, prominent nucleoli and multivacuolated cytoplasm (sebocytes). Focal area shows pleomorphic basaloid cells. (Figure 2 A to C) An extensive area of necrosis consisting of 30-40% of the section examined was seen, while clear cells consist of 40-50% of the section examined. No squamous connection or squamous metaplasia was seen. Sections from the ulcerated area show infiltration of mixed inflammatory cells in the superficial area with sheets of tumor cells in the deeper area. (Figure 2 D) The lympho-vascular and neurovascular invasion was also noted. All the margins were free and uninvolved. Suboccipital lymph node showed tumour deposits of the main tumour. Specimen labelled as post auricular lymph node showed 2 lymph nodes, both of which show tumour deposits. (Figure 3 A & B) Membranous tissue labelled as periosteum shows fibro collagenous and cartilaginous tissue which was also infiltrated by tumour cells. Immunohistochemical markers for EMA, AR, CD117, S-100 and HMB45 were done. EMA show diffuse cytoplasmic positivity in tumor cells with focal membranous positivity while AR shows diffuse intense nuclear positivity in the tumor cells. CD117 was negative in tumor cells. (Figure 4 A to D) The tumor cells were negative for S-100 and HMB45. (Figure 5 A & B) Thus a final diagnosis of sebaceous carcinoma of the scalp with lymph node metastasis was rendered histologically.

3. Discussion

Sebaceous cell carcinoma usually occurs in the periocular areas, while amongst the extraocular areas, the head and neck region is more common where the sebaceous glands are most abundant.[7] This tumour usually occurs in thesixth and seventh decade of life.[7] When occurring in the periocular region, it usually arises from Meibomian glands, which are modified sebaceous glands. The majority of sebaceous carcinomas arise from de novo mutations but may also develop from benign sebaceous neoplasms,

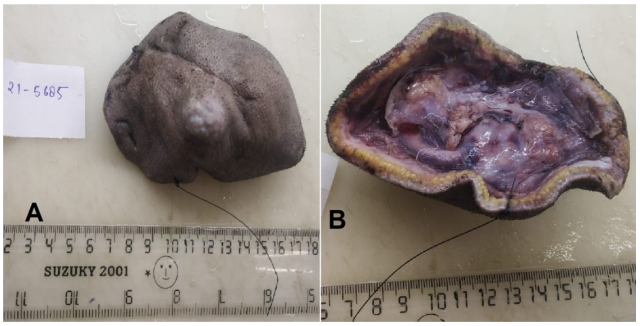

Fig. 1: A): Gross specimen showing a single, greyish-brown, skin covered tissue piece showing two nodular growth protuberances; **B):** Cut section of the tumour showing greyish-white areas with spots of haemorrhages

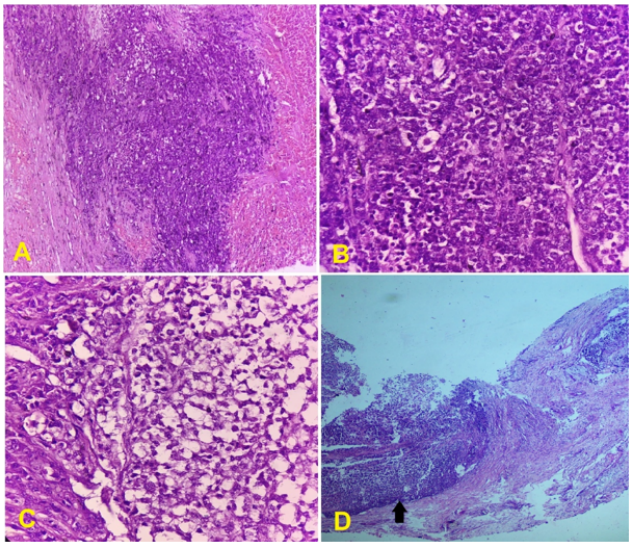

Fig. 2: A): Photomicrograph showing sheets of multivacuolated sebocytes admixed with basaloid cells. Areas of necrosis are also seen (H&Ex40); **B):** Photomicrograph showing sheets of atypical basaloid cells along with singly scattered multivacuolated sebocytes (H&E x100); **C):** Photomicrograph showing clusters of multivacuolated cells (sebocytes) with basaloid cells (H&E x400); **D:** Photomicrograph from ulcerated area shows infiltration of mixed inflammatory cells in the superficial area with sheets of tumor cells in the deeper area (Black arrow) (H&E x40)

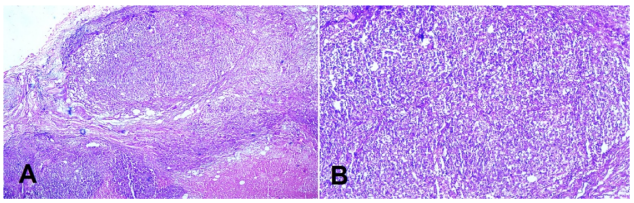

Fig. 3: A): Photomicrograph from lymph node show loss of lymph node architecture with presence of nodular aggregate and sheets of atypical cells along with areas of necrosis (H&E x40); **B):** Photomicrograph from lymph node show sheets of atypical cells with clear cytoplasm (Sebocytes). (H&E x100)

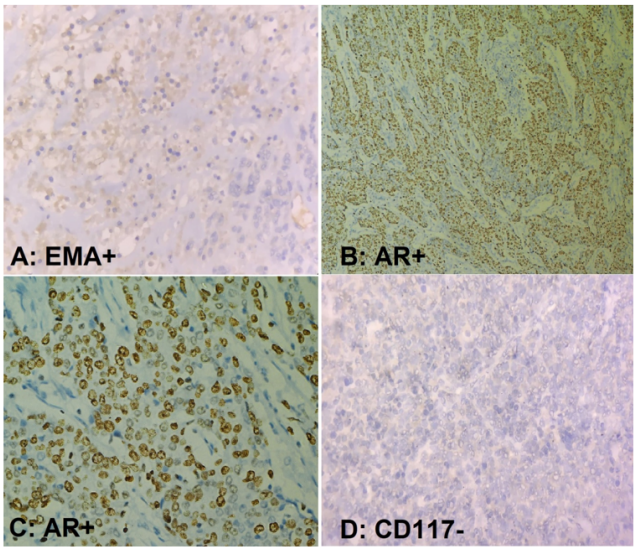

Fig. 4: A): Immunohistochemical staining for EMA: Photomicrograph showing positive cytoplasmic staining in tumor cells (IHCx400); **B):** Immunohistochemical staining for AR: Photomicrograph showing positive intense nuclear staining in tumor cells (IHCx100); **C):** Immunohistochemical staining for AR: Photomicrograph showing positive intense nuclear staining in tumor cells (IHCx400); **D):** Immunohistochemical staining for CD117: Photomicrograph showing tumor cells negative for CD117 staining (IHCx400)

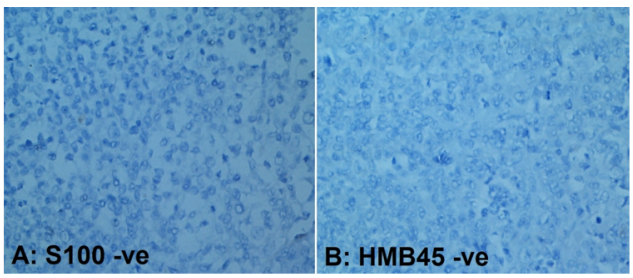

Fig. 5: A): Immunohistochemical staining for S-100: Photomicrograph showing negative staining in tumor cells for S-100 (IHCx400); **B):** Photomicrograph showing negative staining in tumor cells for HMB45 (IHCx400)

though uncommon.[8] Sebaceous carcinomas associated with Muir-Torre syndrome have a loss of mismatch repair gene while those arising sporadically do not show loss of mismatch repair or any microsatellite instability.[8] Some studies showed association with the germline mutations in the MSH2 and MLH1 genes found on chromosomes 3p and 2p respectively.[9] Risk factors for the development of sebaceous cell carcinoma are advanced age, Asian or South Asian race, previous history of irradiation, Muir-Torre syndrome, and immunosuppression following renal transplantation.[10] A tumour of sebaceous glands usually starts as a solitary, firm to hard, slow-growing erythematous or sometimes pale yellow-coloured nodule mainly over the head and neck and, less commonly over the trunk or genitals.[11] The biopsy is essential for the diagnosis of sebaceous cell carcinoma. Histopathologically, the tumour is composed of multiple irregular lobules of various sizes composed of sebocytes separated by a fibrovascular stroma. These can be classified as well, moderate or poorly differentiated depending upon the relative proportion of mature appearing sebocytes which are multivacuolated cells and undifferentiated basaloid cells along with taking into consideration the degree of pleomorphism, necrosis and mitosis.[12] Tumour cells in sebaceous carcinoma are often large and may show squamoid changes. In this case, it should be differentiated from squamous cell carcinoma with clear cell changes which does not show multi-vacuolations as sebocytes. Sometimes, tumour cells show basaloid differentiation with inconspicuous lipidization, and the tumour must be distinguished from basal cell carcinoma with sebaceous differentiation which shows peripheral palisading of basaloid cells and focal differentiation of multivacuolated sebocytes. Immunohistochemical staining for EMA can differentiate sebaceous carcinoma from basal cell carcinoma and squamous cell carcinoma. Ruling out any possibility of metastasis is also essential.[1,13] sebaceous carcinoma can be differentiated with malignant melanoma by negative staining for HMB45 in sebaceous carcinoma. Complete surgical excision is the treatment of choice while the local recurrence rates range from 9 to 36%, which tends to occur within 5 years.[14]

In our case, the disease presentation was in the second decade, at a young age which differs from the usual elderly age presentation. He had a negative family history of any associated or previous malignancy. The patient developed tumour relapse after an excision biopsy which indicates the locally aggressive nature of this tumour. The patient was then successfully treated with wide local excision and was kept under a follow-up. Now after one year of his operation, he is doing fine without any recurrence or metastasis. No chemotherapy or radiotherapy was given.

Sebaceous carcinoma is an important differential to be kept in mind while dealing with cutaneous lesions both clinically and histopathologically. The timely and accurate diagnosis helps in the initiation of an early treatment while for recurrent tumors, adjuvant radiotherapy may help to improve the overall outcome.

4. Conclusion

As sebaceous carcinoma presents with a varied clinical and histopathological appearance, a delayed diagnosis or misdiagnosis is not uncommon. Misdiagnosis as a benign mimic may lead to unwanted morbidity and mortality. Thus, both the clinician and pathologist should be aware of the clinicopathological features of these lesions, maintaining a careful and vigilant approach for an accurate diagnosis,

preventing any unwanted misdiagnosis for initiation of proper management and treatment and further follow up.

5. Source of Funding

None.

6. Conflict of Interest

None.

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