



Metastatic Sebaceous Carcinoma– A Rare Presentation

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Abstract

Sebaceous carcinoma is one of the rare malignancies encountered in clinical practice. They are very aggressive because of their pagetoid type of spread and also because of vascular and perineural invasion. With these features it forms a therapeutic challenge. Here, we present a rare case of sebaceous carcinoma in a 62 year old female with metastases in both parotid and abdomen 6 months after surgical resection of primary in the eyelid.

Keywords: Sebaceous carcinoma, Malignancy, Eyelid, Parotid.

Introduction

Sebaceous carcinoma is a rare, potentially aggressive, malignant tumour of sebaceous gland that arises either ocularly or extraocularly (25%) and diagnosis is often delayed because of its varied clinical presentation. Prevalence varies from 0.05% to 0.7% of all the skin cancers.¹

Head and Neck area being rich in sebaceous glands causes a high incidence of sebaceous carcinoma compared to other areas. Extraorbital sebaceous carcinoma is less aggressive when compared with periorbital because of a reduced tendency for regional metastasis (1.4% for extraorbital vs. 4.4% for periorbital).²The parotid gland forms frequent site of origin because of presence of pluripotential cells or ectopic sebaceous glands. Metastasis to parotid gland has also been reported.³ Here, we present a rare case

of sebaceous carcinoma where metastasis has occurred to parotid and abdomen.

Case Report

A 62 year old female presented with a painful left cheek and neck swelling (figure 1) since 6 months and pain in the left side of the abdomen since 6 months. She had a swelling in the left upper eyelid one year back for which she was operated (figure 2) in a local eye hospital and the post-operative biopsy report showed it to be a sebaceous carcinoma (figure 3). There was no history of any dental procedures in the recent past and she is non-diabetic, non-hypertensive. Her sleep pattern was disturbed because of the pain and also mild alteration in the bowel pattern was present.

On general examination, pallor, clubbing and lymphadenopathy were noted. On inspection, two

oval swellings were noted which were approximately 3cm x 4cm size in the left parotid region and 4cm x 5cm approximately in the neck region corresponding to level II,III,IV and were having well-defined edges. Skin over the swelling appeared red and tense. On palpation, edges were irregular and consistency was hard. Swelling could be moved in both vertical and horizontal direction. It didn't mold on palpation. It was non-reducible, non-compressible, non-fluctuant and non-pulsatile. Skin over the swelling was fixed. Sternocleidomastoid was palpable over the swelling. On auscultation, no bruits heard. Transillumination was negative.

Taking note the previous history of sebaceous carcinoma and its aggressive nature, a possibility of metastasis was suspected and a PET-CT was done. It showed increased uptake in the left parotid region, left side of the neck in level II, III & IV regions and left para-colic region (figure 4). As the neck swelling was unresectable, a minilaparotomy was done and a sample sent for HPE which confirmed it as a metastasis. Also FNAC of neck swelling and parotid were done separately and they also showed it as metastases. Patient was put on chemotherapy + radiotherapy and was advised to be on regular follow-up.



Figure 1



Figure 2

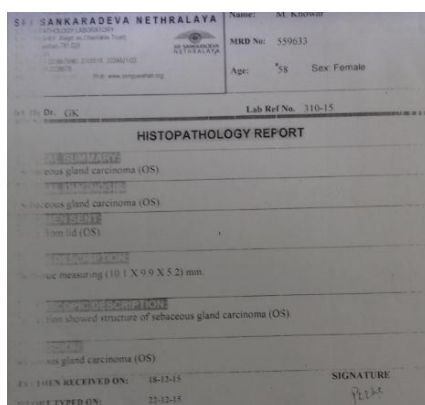


Figure 3

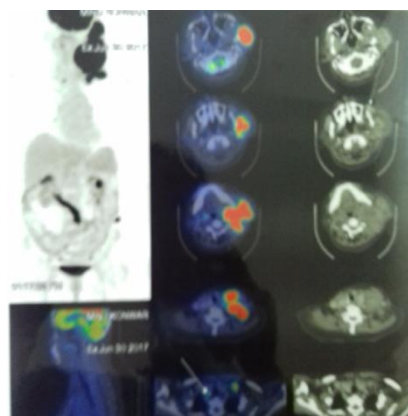


Figure 4

Discussion

Sebaceous carcinoma (SC) is well-known for its aggressive nature. It arises from adnexal epithelium of the meibomian or tarsal glands which are modified sebaceous glands. It can be ocular or extra-ocular. It most commonly occurs at

60-80 years of age with slight female preponderance.⁴ Of ocular SC, upper eyelid forms the most common site due to high intensity of meibomian glands.

Extraocular SC accounts for 25% of all SC cases.⁵ Head and neck region being rich in sebaceous

glands, incidence remains high. Parotid gland forms one of the frequent site because of the presence of pluripotent cells. Also parotid contains ectopic sebaceous glands formed by internal displacement of ectodermal tissue during embryogenesis. Regional and distant metastases rates have been reported in up to 21% of cases.⁶In our case, the distant metastasis has occurred to abdomen. Histologic features include marked atypia with undifferentiated cells having more eosinophilic cytoplasm and pagetoid cells staining for lipid instead of mucopolysaccharides.⁷

Exact cause of SC is unknown but some cases are found to be associated with Muir-Torre syndrome (MTS)⁸ and some with previous irradiation history.⁹Frequent multicentric origin and pagetoid spread makes the tumour recur after conventional 5-6mm margin surgical excision or Mohs micrographic surgery. Local recurrence after surgical excision with frozen section control tends to occur within 5 years in approximately 9% to 36% of patients and as early as 3 months to 35 months after Mohs micrographic surgery.¹⁰This makes prognosis worse as was the situation in our case.

Metastasis occurs mostly through lymphatic, hematogenous and by lacrimal secretory and excretory systems. Lymph nodes primarily involved include preauricular and/or cervical. Secondary parotid masses have also been reported earlier.¹¹Metastatic disease is associated with a 50% to 67% 5-year mortality rate.¹²

Treatment options include radiotherapy for both primary and secondary invasion to parotid and cervical lymph nodes. A total dose of 45-63 Gy in 4-7 weeks is advised¹³and was given to our patient. A metastatic disease can be treated by chemotherapy also. 5-fluorouracil, cisplatin and docetaxel combination has been administered for tumours not amenable to surgery with successful outcome.¹⁴The same regimen was followed in our case. Regional lymph node metastasis can be treated by radical neck dissection if resectable or else chemotherapy and radiotherapy can be planned either alone or in combination. In our

patient, the morbidity was decreased but it was not completely disease-free after 6 months follow-up.

Conclusion

As most common extraocular site being parotid and metastasis involves regional lymph nodes most commonly, sebaceous carcinoma can be considered as one of the differential diagnoses in cases of parotid and neck masses. Also, Muir-Torre Syndrome needs to be differentiated when diagnosed.

Conflicts of Interest: None

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