A RARE CASE OF SEBACEOUS CARCINOMA OF SCALP WITH PAROTID AND NECK SECONDARIES

Chitra Subramanian[1], Saravanan Krishnasamy[2], Ashoka Chakravarthi Dhamodaran[3], Manoharan Appavu[4]

[1]Associate Professor, Department of General Surgery, Madurai Medical College, Madurai.
[2]Assistant Professor, Department of General Surgery, Madurai Medical College, Madurai.
[3]Assistant Professor, Department of General Surgery, Madurai Medical College, Madurai.
[4]Post Graduate, Department of General Surgery, Madurai Medical College, Madurai.

ABSTRACT

INTRODUCTION

Sebaceous carcinoma is a rare aggressive skin appendageal tumor mostly arising from periorbital region.[1,2,3,4] Extra orbital sebaceous carcinoma is extremely rare.[5] We report a case of 68yr old man presented as sebaceous carcinoma scalp with parotid and neck secondaries. Definitive diagnosis is only by histopathological examination. Adequate biopsy and early treatment must be the aim in managing this rare entity.

KEYWORDS

Sebaceous Carcinoma, Scalp, Muir-Torre Syndrome.

HOW TO CITE THIS ARTICLE: Chitra Subramanian, Saravanan Krishnasamy, Ashoka Chakravarthi Dhamodaran, Manoharan Appavu. "A Rare Case of Sebaceous Carcinoma of Scalp with Parotid and Neck Secondaries". Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 52, November 30, 2015; Page: 8699-8702,
DOI: 10.18410/jebmh/2015/1207

INTRODUCTION: Sebaceous carcinoma is a rare skin malignancy of the skin appendages, mostly seen in orbital region.[1,2,3,4] Extra orbital sebaceous carcinoma is very rare,[5] occurring mostly in head and neck.[5] Incidence of sebaceous carcinoma varies from 0.2% to 4.6% of all skin cancers. Most patients present in their 6th or 7th decade of life. Women tend to be affected more often than men. An increased frequency is seen in Asian population. No obvious etiology is found for sebaceous carcinoma. But few were associated with Muir-Torre syndrome. These tumors spread by lymphatic's to preauricular and cervical nodes. These tumors have high incidence of local recurrence (30%-40%) and regional metastasis (10%-15%). Definitive diagnosis is only by histo pathological examination. Early diagnosis and treatment is crucial in reducing morbidity and mortality associated with the tumor.

CASE HISTORY: A 68yr old male patient presented to our hospital on fourth week of April, 2015 with complaints of ulcerative growth over forehead for past 10 years. This growth initially started as a nodule which later on ulcerated. Growth is gradually increasing in size to attain present size. He developed a swelling in the right parotid region 4 months back. He also developed a swelling in the left parotid region 2 months back. Both these swelling were insidious in onset gradually increasing in size to attain present size, not associated with pain over the swelling, fever, increased salivation. No history of loss of weight. No significant past medical or surgical history. No history of malignancy in family.

He was not a diabetic, not a hypertensive, no history of tuberculosis.

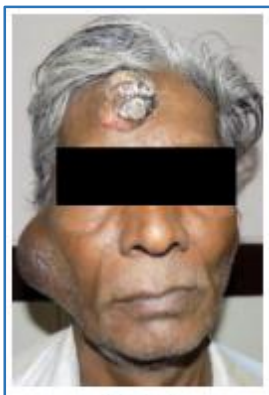


Fig. 1

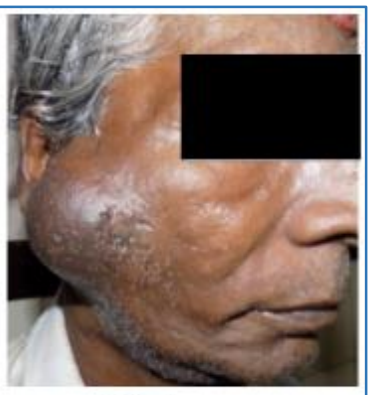


Fig. 2

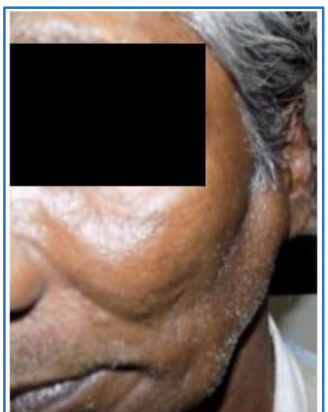


Fig. 3

Submission 23-11-2015, Peer Review 24-11-2015,
Acceptance 26-11-2015, Published 30-11-2015.
Corresponding Author:
Dr. Chitra Subramanian, Shri Prasanna Illam,
Model House No. 3, North 3RD Cross Street,
Anna Nagar, Madurai-625020, Tamilnadu, India.
E-mail: chitra.madurai@gmail.com
DOI: 10.18410/jebmh/2015/1207

On examination patient was moderately built and moderately nourished. There was an ulcero-proliferative growth over forehead of size about 5cmx5cm not fixed to deeper structures, not bleeding on touch. Right parotid was enlarged for about 8cmx6cm which was hard and fixed to deeper structures. Left parotid gland was enlarged for about 3 cm x 3 cm which was firm and mobile. Right level II hard mobile cervical lymphadenopathy was present.

Patient hospitalized and on investigation hemoglobin–12mg/dl, total count–8600cells/cu.mm, blood sugar–121mg/dl, blood urea–30mg/dl, serum creatinine–0.8mg/dl, HIV–non reactive. Ultrasound of abdomen and pelvis and x-ray chest was normal. CT brain showed soft tissue mass in forehead without calvarial erosion. CT neck showed metastasis involving both parotid gland and bilateral superficial parotid lymphadenopathy with right level II cervical lymphadenopathy.

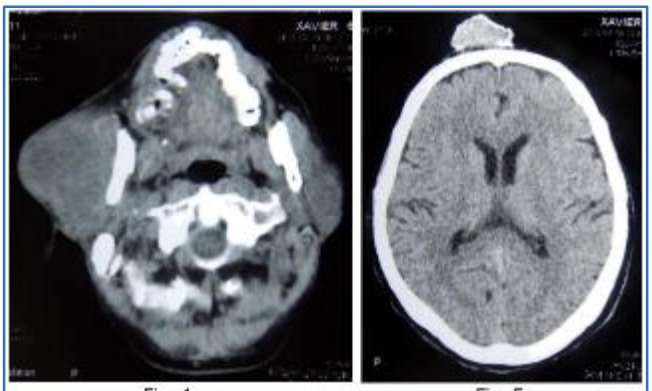


Fig. 4 Fig. 5

Edge wedge biopsy from forehead growth showed features of squamous cell carcinoma. FNAC from right parotid and right cervical lymph node showed features of metastatic deposits. FNAC from left parotid showed features of sialadenitis.

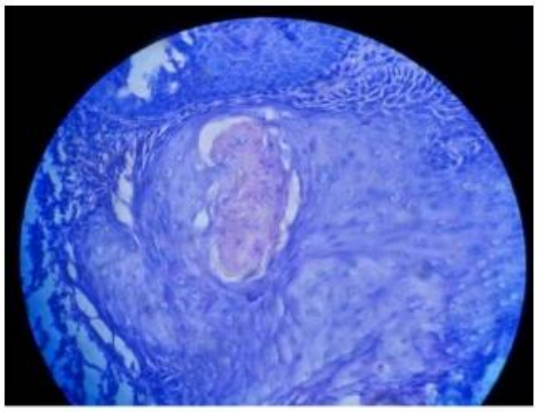
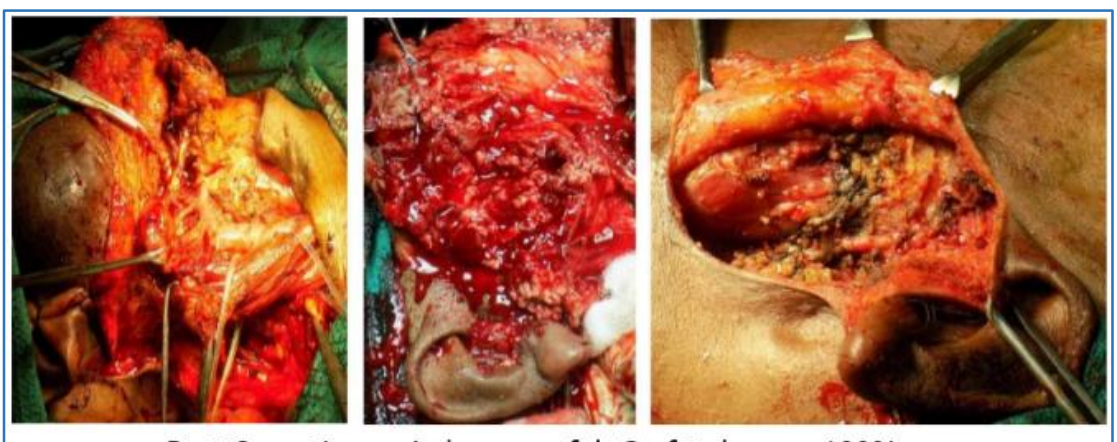


Fig. 6: Biopsy from forehead lesion

We planned for wide local excision of forehead lesion with a margin of 5mm with split skin grafting, right radical parotidectomy, left superficial parotidectomy and right modified radical neck dissection as suggested by surgical oncologist

INTRA OPERATIVE FINDINGS: Growth of size 5 cm x 5 cm over forehead not involving pericranium, multiple neck nodes in right side in level II, III, IV, Right parotid enlargement of size 8 cm×6 cm with masseter adhesion with intra parenchymal necrosis and invasion of facial nerve. Left parotid enlargement involving superficial lobe of size 4 cm×3 cm not involving masseter muscle.



Post Operative period uneventful. Graft take was 100%

Fig. 7: ssg take

Post operative histopathological report was sebaceous carcinoma with metastatic deposits in right parotid gland and right cervical lymph node and left parotid sialadenitis.

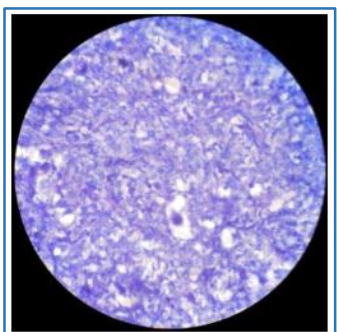


Fig. 8: HPE sebaceous ca

Epithelial membrane antigen was positive in the tumor tissue which is a marker for sebaceous carcinoma.

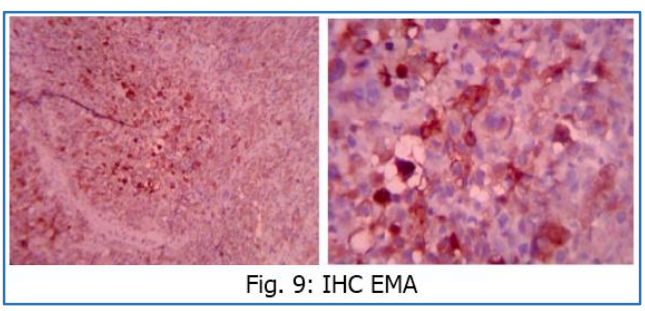


Fig. 9: IHC EMA

Subsequently CECT abdomen and pelvis, colonoscopy was done to rule out visceral malignancies which were normal. He was referred to medical oncologist who planned for chemotherapy with 5-fluorouracil, cisplatin regimen. He was followed up for recurrence.

DISCUSSION: Sebaceous carcinoma is uncommon tumor arising from sebaceous glands of skin and thus it can arise anywhere in the body.[1,2,3]But 75% of the tumor arises in the periocular region which is more aggressive.[4] Only 25% of the tumor arise in extra ocular sites.[5]The head and neck are where most extra ocular sebaceous gland carcinoma occur, with the parotid gland alone representing 30% of cases.[5] Women tend to be more affected than men.[6,7] Most patients present in their 6th or 7th decade of life.[7] It is shown to be related to germ line mutations in the MSH2 and MLH1 genes found on chromosomes 3p and 2p, respectively.[8] Reported risk factors for sebaceous carcinoma include advanced age, Asian or South Asian race, women, previous irradiation to the head and neck and a genetic predisposition for Muir-Torre syndrome or possibly familial retinoblastoma.[9]

It may occur in association with Muir-Torre syndrome which is a combination of sebaceous carcinoma, epithelioma or adenoma with internal visceral malignancies and keratoacanthoma.[10]

Sebaceous carcinoma often has more nonspecific and varied clinical presentation which often results in delayed diagnosis. Most often it presents as a painless gradually enlarging nodule and may mimic more common benign ophthalmic condition.[6,7] Definitive diagnosis is only by histopathological examination. Possibility of MTS must be considered in every case of sebaceous carcinoma and lifelong surveillance is required.

The morphological hall mark of sebaceous carcinoma is the detection of sebaceous cells and demonstration of fat in vacuolated tumor cells.[4] Poorly differentiated sebaceous carcinomas lack lipid vesicles and consist of predominantly basaloid tumor cells that imitate carcinomas such as basal cell carcinoma and squamous cell carcinoma.[8] The histological criteria for sebaceous carcinoma are high mitotic activity, nuclear pleomorphism, lobular architecture and foamy vacuolization of the cytoplasm.[11]

Using immunohistochemical battery, including epithelial membrane antigen (EMA), breast carcinoma antigen 255 (BCA255), CAM 5.2 can be helpful in distinguishing sebaceous carcinoma from basal cell carcinoma and sebaceous cell carcinoma. Human milk fat globules subclass 1 and 2 (HMFG1 and HMFG2) are positive in basal cell carcinoma which helps it to distinguish it from sebaceous cell carcinoma in which it is usually negative.[12] Histologically, the poor prognostic indicators are poor differentiation, presence of lymphatic or vascular permeation, presence of pagetoid cells shown in histology and immuno-histochemical staining.[11]

Local recurrence rate 5 years after surgical excision range from 9–36%.[13] Metastasis occur in 14–25% of patients.[14] It first spreads to draining lymph nodes and then to distant sites which include liver, lung, bone and brain.[15]

Surgery remains the primary modality of treatment for sebaceous carcinoma which involves wide excision of lesion with a margin of 5mm–6mm.[8,6] Radiation therapy is considered as palliative treatment. Systemic chemotherapy with 5-fluorouracil, cisplatin, docetaxel and capecitabine may be an option for tumor refractory to surgical excision and radiotherapy.

CONCLUSION: In conclusion, sebaceous carcinoma is a rare, aggressive tumor which provides a diagnostic dilemma for many physicians and surgeons. Accurate diagnosis is crucial as diagnosis is mainly by histopathological examination. Besides that, this tumor

should alert the clinician to the possibility of Muir-Torre syndrome and lifelong surveillance is needed. Patients must be followed up for possibility of recurrence.

REFERENCES:

1. Kass LG, Hornblass A. sebaceous carcinoma of the ocular adnexa.Surv opthalmol. 1989 May-Jun. 33(6): 477-90.
2. Gomes CC, Lacerda JC, Pimenta FJ, do Carmo MA, Gomez RS. Intraoral sebaceous carcinoma. Eur Arch Otorhinolarngol. 2007 Jul. 264(7): 829-32.
3. Tan O, Ergen D, Arslan R. Sebaceous carcinoma on the scalp. Dermatol Surg. 2006 Oct. 32(10): 1290-3.
4. Karthika Natarajan, Reena Rai, and Suma B. Pillai. Extra ocular sebaceous carcinoma: A rare case report. Indian Dermatology Online Journal, 2011 Jul-Dec; 2(2): 91–93.
5. Wick MR, Goellner JR, Wolfe JT 3rd, Su WP. Adnexal carcinomas of skin. Extra ocular sebaceous carcinomas.cancer. 1985 sep 1.56(5): 1163-72.
6. Doxanas MT, Green WR. Sebaceous gland carcinoma. Review of 40 cases. Arch Ophthalmol. 1984 Feb. 102(2): 245-9.
7. Pang P, Rodriguez-Sains RS. Ophthalmologic oncology: sebaceous carcinomas of the eyelids. *J Dermatol Surg Oncol*. 1985 Mar. 11(3): 260-4.
8. JoonHo Lee, Hea-Kyeong Shin, and Tae Jung Jang. A Case of Rapidly GrowingExtraocular Sebaceous Carcinoma 2014 Apr 15(1): 32-35.
9. Dasgupta T, Wilson L.D. and Yu, J.B. A retrospective review of 1349 cases of sebaceous carcinoma. Cancer 2009; 115: 158-65.
10. Schwartz RA, Torre DP. The Muir-Torre syndrome: a 25-year retrospect. *J Am Acad Dermatol*. 1995 Jul. 33(1): 90-104.
11. Mathur S K, Singh Sunita, Yadav Rajni, Duhan Amrita, Sen Rajeev. Extraocular Sebaceous Carcinoma-a Rare Tumour at a Rare Site. Egyptian Dermatology Online Journal 2010; 6(2): 14.
12. Bhatia SK, Atri S, Anjum A, Sardha M, Ali SA, Zaheer S, et al. Postauricular sebaceous cell carcinoma. International Journal of Case Reports and Images 2012; 3(9): 29–32.
13. Nelson BR, Hamlet KR, Gillard M, Railan D, Johnson TM. Sebaceous carcinoma. *J Am Acad Dermatol*. 1995 Jul. 33(1): 1-15; quiz 16-8.
14. Rao NA, Hidayat AA, McLean IW, Zimmerman LE. Sebaceous carcinomas of the ocular adnexa: A clinicopathologic study of 104 cases, with five-year follow-up data. Hum Pathol. 1982 Feb. 13(2): 113-22.
15. Husain A, Blumenschein G, Esmaeli B. Treatment and outcomes for metastatic sebaceous cell carcinoma of the eyelid. Int J Dermatol. 2008 Mar. 47(3): 276-9.
16. Spencer JM, Nossa R, Tse DT, Sequeira M. Sebaceous carcinoma of the eyelid treated with Mohs micrographic surgery. J Am Acad Dermatol. 2001 Jun. 44(6): 1004-9.