Cancer Association of South Africa (CANSA)



Fact Sheet on Sebaceous Gland Carcinoma

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Introduction

Sebaceous gland carcinoma (SC) is a rare skin cancer. It is considered an aggressive cancer. It is also referred to as sebaceous gland carcinoma, sebaceous gland adenocarcinoma or Meibomian gland carcinoma. If this cancer spreads, it can be deadly.

Sebaceous glands are part of epidermal appendages. Neoplasms of the sebaceous glands may be benign, such as sebaceous hyperplasia or sebaceous gland adenomas. The malignant sebaceous gland carcinoma most commonly arises in the periocular area (around the eyes). Few cases of sebaceous cell carcinoma have been reported at extraocular sites.



[Picture Credit: Sebaceous Gland Carcinoma]

The most common site of origin is the meibomian glands of the eyelids, leading to the term meibomian gland carcinoma. However, this neoplasm can occur in other sebaceous glands, such as in the caruncle (the small, red portion of the corner of the eye that contains modified sebaceous and sweat glands), the glands of Zeis (the unilobar sebaceious glands located on the margin of the eyelid which service the eyelash), and in the eyebrow.

Incidence of Sebaceous Gland Carcinoma (SC) in South Africa

The National Cancer Registry (2016) does not provide any information regarding the incidence of sebaceous gland carcinoma.

Signs and Symptoms of Sebaceous Gland Carcinoma (SC)

Many sebaceous carcinomas (SC) develop on the eyelids. When this rare skin cancer develops on an eyelid, the person may notice one or more the following:

- Slowly growing, often yellowish lump on the eyelid that feels firm, deep, and painless.
- Thickening of an eyelid, where lid meets lash.

- Yellow or reddish crust on eyelid, where lid meets lash.
- Growth on eyelid that looks like a pimple.
- Growth on eyelid that bleeds.
- Sore on eyelid that does not heal, or heals and reappears.

As the cancer progresses, it often looks like the person has pink eye. One may see growths on the upper and lower eyelid. The growths may open and ooze fluid. The eyelashes often fall out. As the cancer spreads, it can affect the eyesight.

Risks and Causes of Sebaceous Gland Carcinoma (SC)

Like most cancers, the cause is far from fully understood. These cancers may be associated with:

- Non-cancerous lumps (benign adenomas) of the sebaceous glands
- Exposure to radiation previous radiotherapy or (less likely) repeated X-rays
- A genetic condition called Muir Torré syndrome. Muir-Torre syndrome (MTS) is the combination
 of neoplasms of the skin (usually sebaceous adenoma, sebaceous epithelioma, or sebaceous
 carcinoma but also keratoacanthoma) and a visceral malignancy (usually colorectal, endometrial,
 small intestine, and urothelial).

Gall, R., Oriz-Perez, S. 2020.

"Sebaceous carcinoma (SC) is a rare and aggressive malignancy that develops from the sebaceous glands. Sebaceous carcinoma is most common in the periocular area, head, and neck, but can occur anywhere in the body where sebaceous glands are present. Sebaceous carcinomas may behave as primary neoplasms or may occur in association with Muir-Torre syndrome (a form of Lynch syndrome characterized by sebaceous tumors and other internal cancers, being the most common those affecting the gastrointestinal tract). SC behaves aggressively both locally and distantly; locally, it correlates with a multifocal origin, and causes destruction and sometimes a pattern of pagetoid spread, which is challenging to diagnose. Moreover, this tumor easily causes distant metastasis. For those reasons, the SC is considered an aggressive disease, which causes significant morbidity and mortality."

Anyone can develop SC, but some people have a greater risk:

- Middle age or older. It is very rare for SC to develop before 30 years of age. Most people are 60 years of age or older, and the risk continues to increase with age.
- Weakened immune system. People who have a weakened immune system have a much greater risk.
- Having received radiation treatments for a medical condition. People who received radiation treatments as a child have been diagnosed with this rare skin cancer in their 60s and 70s.
- Muir-Torre syndrome (MTS), a rare medical condition. People get MTS from the genes that they inherit from their parents. MTS greatly increases the risk for developing several other cancers.

SC is often the first sign that a person has MTS.

Other risk factors include:

 Previous radiation therapy to the area for a variety of benign and malignant conditions, e.g. retinoblastoma

- History of oral thiazide diuretic use
- Mutations to the tumour suppressor gene p53
- Immunosuppression

Diagnosis of Sebaceous Gland Carcinoma (SC)

When a dermatologist suspects skin cancer, the dermatologist performs a biopsy. This procedure can be safely performed during an office visit.

If the biopsy confirms that the patient has SC, the dermatologist may also:

- Perform a full-body skin examination to check for other skin cancers
- Enquire regarding a family history of Muir-Torre Syndrome (MTS)

Owen, J.L., Kibbi, N., Worley, B., Kelm, R.C., Wang, J.V., Barker, C.A., Behshad, R., Bichakjian, C.K., Bolotin, D., Bordeaux, J.S., Bradshaw, S.H., Cartee, T.V., Chandra, S., Cho, N.L., Choi, J.N., Council, M.L., Demirci, H., Eisen, D.B., Esmaeli, B., Golda, N., Huang, C.C., Ibrahim, S.F., Jiang, S.B., Kim, J., Kuzel, T.M., Lai, S.Y., Lawrence, N., Lee, E.H., Leitenberger, J.J., Maher, I.A., Mann, M.W., Minkis, K., Mittal, B.B., Nehal, K.S., Neuhaus, I.M., Ozog, D.M., Petersen, B., Rotemberg, V., Samant, S., Samie, F.H., Servaes, S., Shields, C.L., Shin, T.M., Sobanko, J.F., Somani, A.K., Stebbins, W.G., Thomas, J.R., Thomas, V.D., Tse, D.T., Waldman, A.H., Wong, M.K., Xu, Y.G., Yu, S.S., Zeitouni, N.C., Ramsay, T., Reynolds, K.A., Poon, E. & Alam. M. 2020.

"Sebaceous carcinoma usually occurs in adults older than 60 years, on the eyelid, head and neck, and trunk. In this Review, we present clinical care recommendations for sebaceous carcinoma, which were developed as a result of an expert panel evaluation of the findings of a systematic review. Key conclusions were drawn and recommendations made for diagnosis, first-line treatment, radiotherapy, and post-treatment care. For diagnosis, we concluded that deep biopsy is often required; furthermore, differential diagnoses that mimic the condition can be excluded with special histological stains. For treatment, the recommended first-line therapy is surgical removal, followed by margin assessment of the peripheral and deep tissue edges; conjunctival mapping biopsies can facilitate surgical planning. Radiotherapy can be considered for cases with nerve or lymph node involvement, and as the primary treatment in patients who are ineligible for surgery. Post-treatment clinical examination should occur every 6 months for at least 3 years. No specific systemic therapies for advanced disease can be recommended, but targeted therapies and immunotherapies are being developed."

Treatment of Sebaceous Glad Carcinoma (SC)

Treatment for SC may include:

- Surgery (often Mohs surgery)
- Surgery to remove lymph nodes
- Radiation treatment
- Cryotherapy this treatment involves removing diseased skin by freezing it
- Clinical trial some patients are encouraged to join a clinical trial

McGrath L.A. Currie, Z.I. Mudhar, H.S., Tan, J.H.Y. & Salvi, S.M. 2020.

Objective: To evaluate the incidence and management of recurrent periocular sebaceous gland carcinoma at a tertiary ocular oncology service in the United Kingdom.

Methods: This was a retrospective cohort study of 62 patients with sebaceous gland carcinoma treated between 2004 and 2017. A total of 10 eyes were treated for local recurrence. The following variables were recorded: age and sex of patient; tumour location, histological subtype; recurrence type; treatment and outcome.

Results: Of the 62 cases with eyelid SGC, 10 (16%) had recurrences during the study period and satisfied inclusion criteria. There were six (60%) females and four males in the recurrent group. The mean time interval between initial excision and tumour recurrence was 37 months (median 23 months; range 4 to 84 months). Four patients received cryotherapy to the lids and conjunctiva to control recurrent disease and two patients were treated with topical or intralesional chemotherapy. Four patients (40%) underwent orbital exenteration during the study period. Metastasis occurred in 20% over a mean follow-up of 113 months (median 106; range 47-184 months).

Conclusions: The risk factors for local recurrence of SGC after wide excision with paraffin section control were reported, and an approach to these recurrent lesions was proposed. The results of this study will help guide surgeons dealing with the medical and surgical conundrum of recurrent disease. The risk of recurrence is highest in the first 2 years after initial excision.

About Clinical Trials

Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The <u>South African National Clinical Trials Register</u> provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

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Sources and References consulted or Utilised

American Academy of Dermatology

 $https://www.aad.org/dermatology-a-to-z/diseases-and-treatments/q---t/sebaceous-carcinoma \\ https://www.aad.org/dermatology-a-to-z/diseases-and-treatments/q---t/sebaceous-carcinoma/signs-and-symptoms \\ https://www.aad.org/dermatology-a-to-z/diseases-and-treatments/q---t/sebaceous-carcinoma/diagnosis-treatment-and-outcome \\$

DermNet NZ

http://www.dermnetnz.org/lesions/sebaceous-gland-carcinoma.html

eMedicine.Medscape

http://emedicine.medscape.com/article/1213781-overview http://emedicine.medscape.com/article/1213781-treatment#a1135

EveWeb.Org

http://www.eyeweb.org/evisceration.htm

Gall, R., Oriz-Perez, S. 2020. Sebaceous Gland Carcinoma. *In*: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2020 Jan. 2020 Sep 1.

Lally, S.E., Rao, R., Shields, J.A. & Shields, C.L. 2018. Comparison of posterior lamellar resection versus lumpectomy for initial management of localized tarsal conjunctival sebaceous carcinoma in 54 cases. *Indian J Ophthalmol.* 2018 Sep;66(9):1295-1300. doi: 10.4103/ijo.IJO_239_18. PMID: 30127144

McGrath L.A. Currie, Z.I. Mudhar, H.S., Tan, J.H.Y. & Salvi, S.M. 2020. Management of recurrent sebaceous gland carcinoma. *Eye* (Lond). 2020 Sep;34(9):1685-1692.

MedScape

http://emedicine.medscape.com/article/1093640-overview

National Cancer Institute

http://www.cancer.gov/about-cancer/treatment/clinical-trials

Owen, J.L., Kibbi, N., Worley, B., Kelm, R.C., Wang, J.V., Barker, C.A., Behshad, R., Bichakjian, C.K., Bolotin, D., Bordeaux, J.S., Bradshaw, S.H., Cartee, T.V., Chandra, S., Cho, N.L., Choi, J.N., Council, M.L., Demirci, H., Eisen, D.B., Esmaeli, B., Golda, N., Huang, C.C., Ibrahim, S.F., Jiang, S.B., Kim, J., Kuzel, T.M., Lai, S.Y., Lawrence, N., Lee, E.H., Leitenberger, J.J., Maher, I.A., Mann, M.W., Minkis, K., Mittal, B.B., Nehal, K.S., Neuhaus, I.M., Ozog, D.M., Petersen, B., Rotemberg, V., Samant, S., Samie, F.H., Servaes, S., Shields, C.L., Shin, T.M., Sobanko, J.F., Somani, A.K., Stebbins, W.G., Thomas, J.R., Thomas, V.D., Tse, D.T., Waldman, A.H., Wong, M.K., Xu, Y.G., Yu, S.S., Zeitouni, N.C., Ramsay, T., Reynolds, K.A., Poon, E. & Alam. M. 2020. Sebaceous carcinoma: evidence-based clinical practice guidelines. *Lancet Oncol*. 2019 Dec;20(12):e699-e714.

Schmitz, E.J., Herwig-Carl, M.C., Holz, F.G. & Loefler, K.U. 2017. Sebaceous gland carcinoma of the ocular adnexa – variability in clinical and histological appearance with analysis of immunohistochemical staining patterns. *Graefes Arch clin Exp Opthalmol*. 2017 Nov;255(11):2277-2285. doi: 10.1007/s00417-017-3738-2. Epub 2017 Jul 24.

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http://mrdavidcheung.com/functional/eyelidcancerpictures.html

Shields, J.A., Saktanasate, J., Lally, S.E., Carrasco, J.R. & Shields, D.L. 2015. Sebaceous Carcinoma of the Ocular Region: The 2014 Professor Winifred Mao Lecture. *Asia Pac J Ophthalmol (Philla)* 2015 Jul-Aug;4(4):221-7. doi: 10.1097/APO.00000000000105.

SkinCancerNet

http://www.skincarephysicians.com/skincancernet/whatis_sebaceous_carcinoma.html

Takahashi, Y., Takahashi, E., Nakakura, S., Kitaguchi, Y., Mupas-Uy, J. & Kakizaki, H. 2016. Risk Factors for Local Recurrence or Metastasis of Eyelid Sebaceous Gland CarcinomaAfter Wide Excision With Paraffin Section Control. *Am J Ophthalmol.* 2016 Nov;171:67-74. doi: 10.1016/j.ajo.2016.08.028. Epub 2016 Aug 31.