

# CASE REPORT

## A NEGLECTED CASE OF EXTENSIVE RECURRENT SEBACEOUS CARCINOMA OF THE LEFT EYELIDS AND MANAGEMENT

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**ABSTRACT: INTRODUCTION:** Sebaceous gland tumor of the eyelids may arise from the meibomian glands, glands of Zeis or glands associated with the caruncle. They are included in the list of tumors of the epidermal appendages, so-called adnexal skin structures. Sebaceous gland carcinoma<sup>1</sup> (SGC) might be the second most common lid malignancy after basal cell carcinoma (BCC). Its multifocal origin and pagetoid spread give it a unique place among eyelid malignancies. Sebaceous glands are located in the periocular skin, caruncle, and eyebrow skin follicles. The tumor is a very rare, slow growing, and commonly found in elderly population with female predisposition. Mean age at diagnosis is mid-sixties; however, the tumor has been reported in children as young as 3.5 years old. It is rare in Caucasians and common in oriental Asiatics. SGC most commonly arises from the meibomian glands<sup>2</sup> anterior to the gray line, occasionally from the glands of Zeis or Moll, and from sebaceous glands in caruncle<sup>3</sup>; however, the cell of origin may not be certain in 50–60% of cases.<sup>3</sup> In contrast to basal cell carcinoma (BCC) or squamous cell carcinoma (SCC), SGC is two to three times more common in upper eyelid due to more number of meibomian glands<sup>4</sup> there. **PRESENTATION OF CASES:** In the department of ophthalmology, G.S.L medical college, Rajahmundry, we came across a 52 years old female with recurrent swelling of left eyelid from past 4 years.

**KEYWORDS:** Sebaceous carcinoma, Lid tumour, MOH's surgical procedure, Excentration, Basal cell carcinoma, Squamous cell carcinoma.

**CASE REPORT:** A female patient 52 yrs of age came with chief c/o Swelling in the left eyelid since 2 yrs and Swelling in the left pre auricular area since 1yr. [FIG. 1, 2] swelling started as a small groundnut size swelling (1\*1cm), insidious in onset, gradually progressive in nature to attain a present size. It is associated with loss of appetite and weight loss. Patient had similar complaint 4yrs back swelling around 1\*2 cm for which she underwent excision of the mass 3 yrs back, it was reported as carcinoma of the left upper eyelid with positive margins, in outside hospital. She did not undergo any further intervention. she had H/o recurrence of the swelling in the left eyelid from the same site 1yr after first surgery, for which she was given 3 cycles of chemotherapy with inj cisplatin and inj 5 flouro uracil – did not respond properly. H/o swelling in front of the left pre auricular region 1 and half yr back, underwent excision (probably excision biopsy) and sent for histo-pathological examination in a government hospital. Reported as Squamous cell carcinoma in preexisting pleomorphic adenoma with cystic changes or Mucoepidermoid carcinoma. No relavent family history.

She is a known smoker since 25 yrs 1 per day, stopped since 3 yrs. On examination Pre auricular lymph node enlargement present of size 3\*3 cms, non-tender, hard and mobile, Sub

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mandibular lymph node enlargement is present. Ulcero-proliferative growth of size 8 \* 6 cms over left orbital region involving both upper and lowerlid and extending from medial canthus and laterally towards temporalis region. Irregular and everted edges. [Fig. 1, 2] Areas of slough present with foul smelling discharge which bleeds on touch. LE is not seen separately from growth. Globe was congested with corneal opacity, Ocular movements are restricted. Vision in right eye is 3/60 and left eye is no PL.

Fundus right eye dry ARMD changes present, left eye no view. FNAC of the pre auricular node and biopsy from the eyelid mass was done and sent to HP, showed ample cellularity comprising of oval to polygonal cells arranged in small and large sheets, clusters and papillaroid structures, The nuclei are round to oval showing moderate pleomorphism, cytoplasm is scant to ample dense eosinophilic, in some cells appears vacuolated, Background shows keratinous material and scattered large cells with foamy cytoplasm, few lymphoid cells, features suggestive of mucoepidermoid carcinoma local recurrence – left parotid gland. [Fig. 3] Biopsy showed scattered malignant epithelial cells, these are large round to oval to polygonal having fair amount of eosinophilic / vacuolated cytoplasm exhibiting irregular hyperchromatic nuclei with high N: C ratio, some showing prominent nucleoli. [Fig. 4] Stroma shows focal area of necrosis with neutrophilic infiltration. Suggestive of squamous cell carcinoma/ sebaceous carcinoma<sup>5</sup>. [Fig. 5, 6] FNAC from left pre-auricular swelling showed loose clusters, nests, monolayered sheets and discretely scattered round to oval to polygonal epithelial cell showing scanty to fair amount of foamy vacuolated / orangeophilic cytoplasm exhibiting large pleomorphic nuclei some showing prominent nucleoli, Backguard shows many clumps of eosinophilic keratinous and necrotic material with few neutrophils and lymphocytes suggestive of cytomorphology favours metastatic squamous cell carcinomatous deposit in left pre auricular lymph node.[Fig. 7] 2d Echo and Colour Doppler Showed Good Lv Systolic Function.Ct Showed Lobulated Enhancing Left Peri Orbital Mass With Extraconal Component, Heterogenous lesion in the superficial portion of the left parotid gland.[Fig. 8, 9] planed for Wide local excision with orbital exenteration with parotidectomy with neck node dissection was done and three specimens were sent to HP. [Fig. 10, 11, 13, 14] Gross examination showed Jar (1) labelled as left orbit tumor contains skin attached exophytic growth measuring 7.0x6.0x4.0 cm, cut section grey white solid Jar (2) labelled as carcinoma parotid node, contains single bit of fibrofatty tissue measuring 8.0x5.5x2.0 cm, which on cutting shows grey white solid area measuring 2.5x2.0 cm. Jar (3) labelled as left SOHND node contains single bit of fibro fatty tissue measuring 4.0x3.0x1.0 cm., cut section shows 5 lymph nodes of sizes varying from 0.5cm to 2.0x1.0x1.0 cm (G/S). microscopic examination jar 1 showed: tumor with large round to polygonal cells having fair amount of granular eosinophilic / vacuolated / clear cytoplasm exhibiting moderately pleomorphic vesicular / hyperchromatic nuclei with clumped chromatin many showing prominent nucleoli with Few bi and multinucleated tumor cells and bizarre cells also seen. There is presence of comedo necrosis with Surface epithelium as well as stroma shows and dense neutrophilic infiltration and some of the areas show sebaceous differentiation.jar 2 showed: parotid gland with secondary deposit in lymph node, 3. Showed nodes with reactive hyperplasia and no secondary deposit seen. Final diagnosis is ORBITAL tumor – moderately differentiated sebaceous carcinoma with squamous differentiation with parotid node – positive.

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**DISCUSSION:** Sebaceous gland carcinoma is a malignant neoplasia that originates from cells that comprise sebaceous glands. There is an unusual abundance of sebaceous glands in the ocular region,<sup>6</sup> particularly in the tarsus (meibomian glands), cilia (Zeis glands) and caruncle.

Historically, sebaceous gland carcinoma of the eyelid is notorious for masquerading as a more common benign condition, often resulting in a long delay before the correct diagnosis is made. Such a delay in diagnosis can increase the chance of local recurrence, metastasis and death. Its occurrence in the western literature is reported to be less than 1% of all eyelid tumors and accounts for 1-5% of all malignant eyelid tumors [7-9]. Recent studies from India and China have shown that sebaceous carcinoma accounts for 33-60% of malignant eyelid tumors.<sup>7,8,9</sup>

Though there have been numerous publications that stress the clinical features of sebaceous carcinoma and its tendency to masquerade as chalazion or chronic blepharoconjunctivitis, studies show that delay in diagnosis is still common. Biopsy has been reported as the preferred mode for establishing correct diagnosis of such lid nodules. FNAC has been employed only rarely in the diagnosis of periocular sebaceous carcinoma<sup>10</sup> possibly because of the limited tissue availability. However, FNAC can be helpful in differentiating benign from malignant condition by a careful study of the smear. The important components of any cytology smear are the nucleus, cytoplasm, cellular arrangement and the slide background. Nuclear characteristics are important criteria in diagnostic cytopathology. As a rule, benign cells have smooth contours and are similar to one another. Malignant cells show alterations in size and shape with irregular nuclear folding, indentation and outward irregularities, all in haphazard arrangement. Malignant cells usually have large nucleoli with prominent polymorphism and greater variation in the number of nucleoli per nucleus. In contrast, smears of chalazion display inflammatory cells without any malignant cells. Microscopy shows a lipogranulomatous reaction caused by liberated globules of fat, surrounded by epithelial cells and multinucleated giant cells intermixed with neutrophils, lymphocytes and plasma cells.

**DIFFERENTIAL DIAGNOSIS:** Occasionally, SGC may diffusely infiltrate conjunctival epithelium. This may resemble lesions such as chronic blepharoconjunctivitis, superior limbal keratoconjunctivitis or cicatricial pemphigoid, and BCC. The tumor may mimic chronic inflammation and even grow bacteria on repeated cultures but does not improve with antibiotics. Conjunctival scraping may reveal the underlying SGC pathology. Due to such mimicking, SGC is one of the forms of masquerade syndromes. Diffuse intraepithelial sebaceous carcinoma<sup>11,12,13</sup>

Closely resembles SGC in its rarity and indolent course; however, the former is not an obvious primary tumor of the eyelid and may not require exenteration. Papilloma simulates SGC and sometimes may be differentiated only by histology.

**TREATMENT:** SGCs are often inadequately treated at first intervention. Different treatment modalities include local excision, orbital exenteration, radical neck dissection, radiation, or chemotherapy depending on the stage of the tumor at the time of presentation. Wide excision at an early stage is important. Prior to surgical excision, it is important to examine the patient carefully for evidence of pagetoid spread or multicentric origin by double eversion of the eyelids, and any conjunctival alteration such as telangiectasia, papillary change, or a mass. In such an

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instance, conjunctival punch biopsies should be taken in addition to surgical resection of the lid lesion.

Surgical treatment may range from a local excision to orbital exenteration. Radical surgical excision with frozen section control by either a standard method or Moh's micrographic surgery<sup>14</sup> is the most common and effective method of treatment. The wound edges should be approximated as far as possible. Approximately, 30% of SGCs recur after resection. Two weeks after surgical resection of the tumor and reconstruction of the eyelid. The advantages of Moh's micrographic surgery, in comparison to a conventional excision of non-melanotic skin cancer are; (1) definitive margin excision and (2) minimal loss of surrounding normal tissue. Few patients with regional nodal metastasis remain alive for long time, and radical neck dissection for isolated cervical node disease is often indicated. Local nodal disease without distant metastasis<sup>15</sup> is treated by radical neck dissection. Few patients with regional nodal metastasis remain alive for long time, and radical neck dissection for isolated cervical node disease is often indicated. Local nodal disease without distant metastasis is treated by radical neck dissection.

**PROGNOSIS:** Prognosis of sebaceous gland carcinoma (SGC) based on clinicopathological<sup>16</sup> tumor morphology. The overall mortality rate is 5–10% because of frequent difficulties, mistakes in diagnosis, and delay in the treatment. The mortality from metastasis may go up to 25%. The adverse prognostic features include involvement of upper or both eyelid and tumor size of 10 mm or more. Others include duration of symptoms more than 6 months (mortality 38%), poorly differentiated tumors, infiltration into blood vessels and/or lymphatics, orbital extension, multicentric origin, and finally pagetoid spread. Tumors less than 6 mm have excellent prognosis. Prognosis of SGC arising from glands of Zeis is more favorable. It goes beyond doubt that SGC is a great mimicker. On one hand, it mimics as simple a clinical condition as blepharitis, on the other hand it may turn out to be a fatal metastatic tumor.

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TABLE 1:

PRIMARY TUMOR (T)	
<input type="checkbox"/> TX	Primary tumor cannot be assessed
<input type="checkbox"/> T0	No evidence of primary tumor
<input type="checkbox"/> Tis	Carcinoma <i>in situ</i>
<input type="checkbox"/> T1	Tumor 5 mm or less in greatest dimension. Not invading the tarsal plate or eyelid margin.
<input type="checkbox"/> T2a	Tumor more than 5 mm, but not more than 10 mm in greatest dimension. Or, any tumor that invades the tarsal plate or eyelid margin.
<input type="checkbox"/> T2b	Tumor more than 10mm, but not more than 20 mm in greatest dimension. Or, involves full thickness eyelid.
<input type="checkbox"/> T3a	Tumor more than 20 mm in greatest dimension. Or, any tumor that invades adjacent ocular, or orbital structures. Any T with perineural tumor invasion.
<input type="checkbox"/> T3b	Tumor complete resection requires enucleation, exenteration or bone resection.
<input type="checkbox"/> T4	Tumor is not resectable due to extensive invasion of ocular, orbital, craniofacial structures or brain.
REGIONAL LYMPH NODES (N)	
<input type="checkbox"/> NX	Regional lymph nodes cannot be assessed.
<input type="checkbox"/> N0	No regional lymph node metastasis, based upon clinical evaluation or imaging.
<input type="checkbox"/> N1	No regional lymph node metastasis, based upon lymph node biopsy. Regional lymph node metastasis.
DISTANT METASTASIS (M)	
<input type="checkbox"/> M0	No distant metastasis (no pathologic M0; use clinical M to complete stage group)
<input type="checkbox"/> M1	Distant metastasis

Table 1



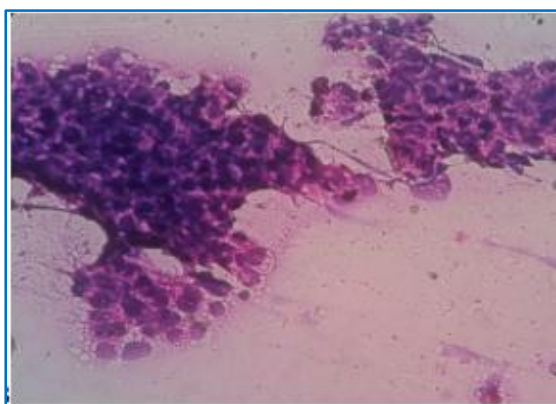
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GROUP	CLINICAL		
	T	N	M
<input type="checkbox"/> 0	Tis	N0	M0
<input type="checkbox"/> I A	T1	N0	M0
<input type="checkbox"/> I B	T2a	N0	M0
<input type="checkbox"/> I C	T2b	N0	M0
<input type="checkbox"/> II	T3a	N0	M0
<input type="checkbox"/> III A	T3b	N0	M0
<input type="checkbox"/> III B	Any T	N1	M0
<input type="checkbox"/> III C	T4	Any N	M0
<input type="checkbox"/> IV	Any T	Any N	M1

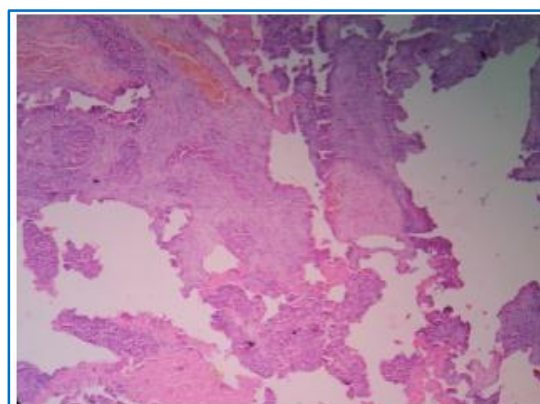
**Table 2**



**FIG. 1 & 2**

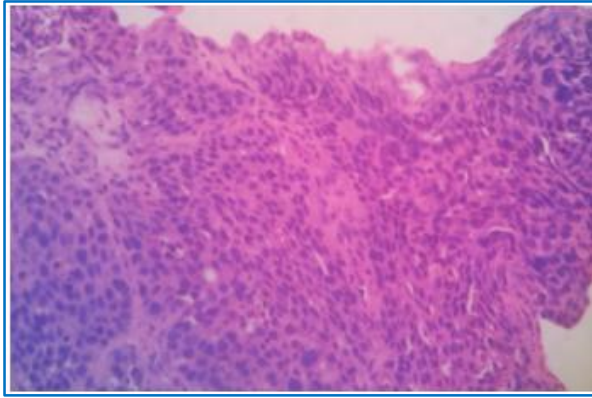


**FIG. 3**

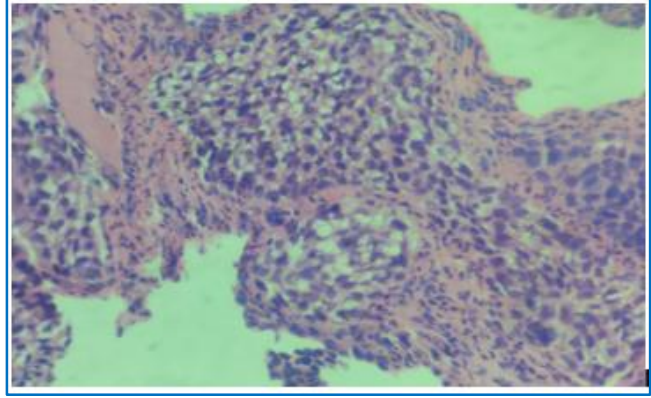


**FIG. 4**

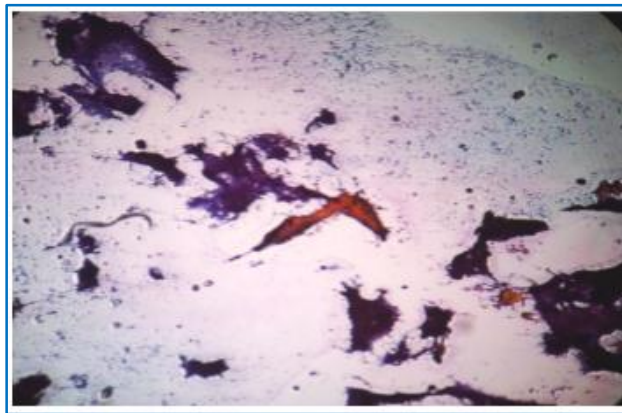
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**FIG. 5**



**FIG. 6**



**FIG. 7**



**FIG. 8**



**FIG. 9**

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**FIG. 10**



**FIG. 11**



**FIG. 12**



**FIG. 13**



**FIG. 14: Post OP**



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