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Case Report

Pathology Section

Metastatic Sebaceous Carcinoma Presenting as Cheek Nodule: A Rare Case

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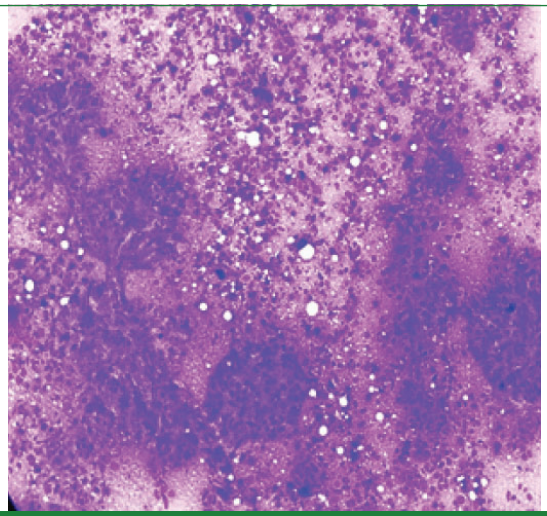
ABSTRACT

Sebaceous Carcinoma is a rare locally aggressive tumour of adnexal epithelium with a potential to metastasize to regional lymph node and distant organs. It constitutes between 1.5 to 5% of all eyelid malignant tumours. After basal cell and squamous cell carcinomas, it ranks third in incidence. Sebaceous carcinoma is seen more commonly in elderly females with average age at diagnosis in mid-sixties. Few cases have been reported in paediatric age group. Sebaceous Gland Carcinoma (SGC) is a slowly progressive tumor that arises usually from the meibomian gland in the eyelid, and has a locally aggressive nature with a tendency for pagetoid spread. The neoplasm is known to masquerade other benign and less malignant lesions. The non-specific clinical symptoms may lead to a delay in diagnosis of sebaceous carcinoma of the eyelid. Less than 120 cases of extra-occular sebaceous cell carcinoma have been reported so far. Here, the authors present a case of 60 years old female patient, who presented with cheek nodule and cytological diagnosis of metastatic sebaceous carcinoma was made based on cytomorphological features.

Keywords: Distant organs, Fine needle aspiration cytology, Ocular sebaceous carcinoma

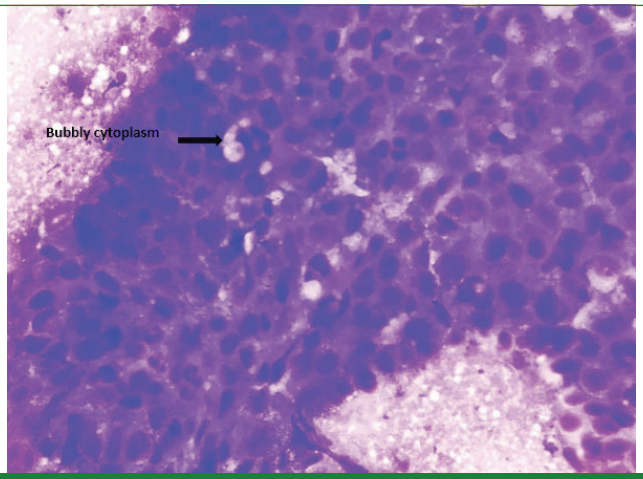
CASE REPORT

A 60 years old female patient, asymptomatic till four years ago, presented with ulcerated nodule in left eyelid. Biopsy from the same confirmed the diagnosis of sebaceous carcinoma. Three months later, patient presented with multiple firm non tender superficial nodules largest measuring 1×1×1 cm in left cheek [Table/Fig-1]. There was no other significant medical history neither was there any history of lymphadenopathy. On Computed Tomography(CT) scan, ill-defined homogeneously enhancing soft tissue mass lesion measuring 40×28×26 mm was seen in left preorbital region involving left eyeball. It was abutting anterior wall of left eye ball extending into anterior aspect of the orbit, also involving left lacrimal gland and anterior aspect of left medial rectus muscle.

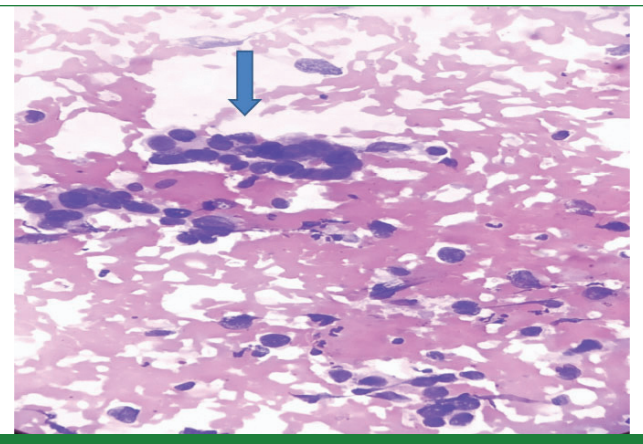


[Table/Fig-1]: Cellular smears showing clusters of malignant epithelial cells in a background of fat vacuoles and necrosis (Giemsa stain 100x).

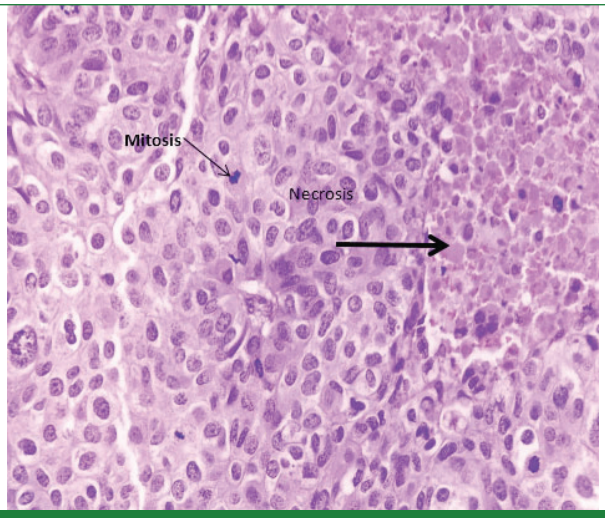
Fine Needle Aspiration (FNA) was performed from cheek nodule and yielded blood mixed aspirate. Cytological examination revealed cohesive clusters of malignant epithelial cells with moderate to marked nuclear pleomorphism hyperchromasia, increase nucleus: cytoplasm (N:C) ratio, occasional prominent nucleoli and moderate amount of eosinophilic cytoplasm. Few of the cells exhibited cytoplasmic vacuolation with bubbly cytoplasm [Table/Fig-2,3]. There was presence of focal acinar pattern [Table/Fig-4] and mitotic activity. Background showed scattered neoplastic cells with few inflammatory cells including neutrophils and lymphocytes. Fat vacuoles were also present in the background. Later, nodules were excised and the biopsy of the same revealed features of sebaceous carcinoma.



[Table/Fig-2]: Cohesive clusters and sheets of cells with dense to vacuolated cytoplasm and moderate nuclear atypia (Giemsa stain 200x).



[Table/Fig-3]: Glandular pattern arrangement of malignant epithelial cells (200x).



[Table/Fig-4]: The tumor cells are arranged in nests and lobules with central comedo necrosis. The cells have foamy cytoplasm, round to oval nuclei with moderate pleomorphism and increase mitosis (Haematoxylin & Eosin stain 200x).

Histopathology slides of the previous specimen were retrieved and examined, showed typical features of sebaceous carcinoma with lobules of epithelial cells at places containing central comedo type of necrosis. The epithelial cells showed round to oval nuclei, with moderate pleomophism, mitotic activity and eosinophilic to foamy cytoplasm. Subsequently, a diagnosis of metastatic sebaceous carcinoma was given on FNA cytology. Based on radiological and histopathological findings, it was diagnosed as a case of sebaceous carcinoma eyelid for which exenteration was done [Table/Fig-5].

[Table/Fig-5]: Clinical image of the patient after exenteration.

Patient's cheek nodules were excised and she underwent multiple cycles of chemotherapy.

DISCUSSION

Malignant eyelid neoplasms contribute for 5-10% of all skin tumours [1]. SGC are rare tumors that account for 3% of all cancers worldwide [2]. After Basal Cell Carcinoma (BCC) and Squamous Cell Carcinoma (SCC), sebaceous gland carcinoma (SGC) ranks third most common malignant tumor of eyelid. The SGC is a slowly growing tumor with a high mortality, and a potential to metastasize to the regional lymph nodes and distant organs. It is seen more commonly in the elderly female population. Majority of the tumours are detected in the head and neck region (face/ear/scalp/neck/lip, 42.8%), followed by the eyelid (34.5%), trunk (14.8%), and extremities (6.5%). SGC arises mostly from the meibomian gland and occasionally from the glands of Zeis or Moll [2]. Locally aggressive nature and pagetoid spread makes this tumour unique among all eyelid malignancies [3]. The tumour is a very rare, slow growing and usually found in elderly female population. Mean age of diagnosis is mid-sixties; however, it has been reported in young children [4]. In a study by Tripathi R et al., regional lymph node metastasis was seen in 20% patients and systemic metastasis in 14% of all cases [5].

Primary SGC can metastasis upto five year [6]. Typically, beginning as a painless papule on the skin, diagnosis and therapy of sebaceous carcinoma tend to be delayed because of its innumerable presentation. In sebaceous carcinoma, the sebaceous lobules show cellular atypia and dysplastic features. There may be local infiltration into tarsal and adnexal tissues. Microscopically, SGC typically shows cells arranged in lobules or nests with pleomorphic, hyperchromatic nuclei and vacuolated (foamy or frothy) cytoplasm by virtue of high lipid content. On histological examination, SGC shows resemblance with the SCC. Although in SGC, cytolplasm appears to be more basophilic in comparision to the eosinophilic appearance of SCC [6].

Biopsy examination shows characteristic histopathological features and therefore, considered diagnostic test for this tumour. However, FNAC is a minimally invasive procedure and can help clinch an early diagnosis. Sebaceous carcinoma shows typical cytomorphological features with cells arranged in loose groups, clusters as well as dispersed singly, these cells show moderately, pleomorphic, hyperchromatic nuclei and moderate to scant amount of clear vacuolated cytoplasm and conspicuous nucleoli. The periorbital primaries tends to metastasize early with an aggressive behaviour culminating into mortality in most cases. Rarity of this tumour delays the exact diagnosis of a sebaceous since it mimics a number of other eye conditions including chalazion, chronic blepharoconjunctivitis, squamous cell carcinoma, pilomatricoma, and basal cell carcinoma, the diagnosis is often delayed. There is ductal obstruction leading to release of fatty contents from sebaceous glands, and granulomatous pattern of inflammation may be seen with presence of neutrophils, lymphocytes, plasma cells and few multinucleated giant cells, which may be similar to a chalazion. Sometimes, there may be non-granulomatous inflammation, often containing neutrophils, similar to blepharoconjunctivitis [7]. Pilomatrixoma on cytological evaluation shows sheets of basaloid cells, "ghost" cells and nucleated basophilic cells with minimal atypia. On the contrary, Basal Cell Carcinoma shows tightly cohesive small clusters of monomorphic basaloid cells devoid of vacuolation [8]. In Squamous Cell carcinoma, cells are in groups or dispersed singly with hyperchromatic nuclei and dense basophilic cytoplasm. Pagetoid spread into the conjunctival epithelium or skin epidermis and presence of vacuolated cytoplasm is a characteristic feature of sebaceous carcinoma [6]. Histologically, it shows solid sheets or lobules of atypical epithelial cells with large, pale or clear cells with vacuolated cytoplasm and scalloped nuclei. On IHC, sebaceous cell carcinoma stain positive for cytokeratin, ER, PR and Oil Red O [7].

Sebaceous carcinomas are aggressive tumours that spread locally. They have potential to metastasize. Metastasis can occur by lymphatic spread or hematogenous spread. The tumour may involve adjacent tissues such as orbit, the periocular area and the parotid gland or spread to the regional cervical nodes. Rarely, metastases may occur to distant sites, such as lung, pleura, liver, brain, pericardium, lips, ethmoid sinus and skull. Mortality rate has been reported as 5-10%, and it may be higher in cases where the symptoms have persisted for a longer period such of time [9].

SGCs have a propensity to recur. Recurrence can occur at the site of previous tumour or at a different site, and can be difficult to treat. The multicentric origin of tumour makes it difficult to determine the nature of the new lesion, which may be a new tumour or a recurrence of the previous one. Tumour differentiation upon microscopy has prognostic importance, with poorly differentiated tumours showing a worse outcome. Clinicopathologic features at the time of presentation are important prognostic factors. Size of tumor more than 10 mm, multicentric origin and duration of symptoms for more

than six months are poor prognostic factors as are presence of local extension to both upper and lower eyelids and/or orbital invasion. Lymphovascular invasion, presence of necrosis, pagetoid invasion of the overlying epithelia of the eyelids and infiltrative pattern on microscopic examination indicate worse prognosis [10]. In the present case, histopathology slides demonstrated pagetoid spread of tumor cells and comedo necrosis predicting a poor prognosis.

The treatment of sebaceous carcinoma without orbital involvement is wide local excision of the tumor [7]. If there is involvement of the orbit, exenteration is required as was done in the present case scenario.

CONCLUSION(S)

Sebaceous carcinoma exhibits a variable clinical presentation so it is often misdiagnosed, delaying proper prognosis and care. Fine Needle Aspiration Cytology is a minimally invasive, cost-effective technique that can be useful to achieve early diagnosis, particularly in such cases in which the histopathological diagnosis is already established. This can help clinician in deciding management plan of the patient without any further delay.

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