PAGETOID SEBACEOUS GLAND CARCINOMA

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Abstract
A High index of clinical suspicion for any recurrent chalazion of unusual consistency demands a full thickness resection, histological examination with frozen section control, rather than repeated curettage which enhances orbital invasion, delays treatment and heralds lymph node metastasis prior to orbital invasion yielding a poorer prognosis & a 15% ten year mortality rate

Key Words : Recurrent, Pagetoid, Meibomian, Metastasis, Carcinoma

Introduction :
Pagetoid Spreading Sebaceous Carcinoma or Meibomian Gland Carcinoma is a very rare disease, frequently affecting elderly patients, commonly in the upper lid. It originates from the numerous Meibomian Glands and the Glands of Zeis or the sebaceous glands in the caruncle and brow, and makes up 10% of all eyelid tumours.
There are various types- Nodular Sebaceous Gland Carcinoma.
Spreading Sebaceous Gland Carcinoma
Sebaceous Gland of Zeis carcinoma.

Case Report :
A 70 years old debilitated patient had a slowly growing swelling noticed on the left eye upper lid since 2 years with redness of the left eye, the swelling being non-tender, hard in consistency and fixed to underlying structures. It was fixed immovably to thickened skin over the swelling with hair loss over it. The conjunctiva was intensely chemotic.


Photo 1

It was treated with a wide margin excision surgery with a glabellar flap reconstruction of the upper eyelid followed by 50 Gy.radiation in 33 Fractions, This was supplemented with Chemotherapy for micrometastasis. Within a year, the patient developed a similar recurrence (photo 1) with findings of: thickened conjunctival epithelium, severe pain, photophobia, non-axial proptosis with extraocular movements being completely restricted, complete lagophthalmos and corneal infiltrates. The vision was perception and projection of light and the pupil fixed semi-dilated and not reacting to light. There was an advanced senile mature cataract yielding a poor fundal glow, and a huge swelling in the left temporal region : 6 cms by 6 cms by 6 cms with skin metastasis, infiltration & fungation A chemotherapy cycle with cisplatin and 5-Flurouracil was started and the patient advised Exenteration for a better survival rate. Routine investigationsat this point were within normal limits except for a normocytic hypochromic anemia. An X-Ray Skull A-P/Lat confirmed metastasis to the orbit and the skull: (Photo 2)
Signs of bad prognosis were: origin from meibomian glands, infiltrative pagetoid growth, upper lid involvement, tumour diameter > 10 mm and symptoms for > 6 months.


Photo 2

Discussion:
This is a difficult diagnosis in the early stages as it resembles less aggressive lesion with subtle signs of malignancy and may masquerade as a chalazion with normal overlying skin. A high index of clinical suspicion for any recurrent chalazion of unusual consistency demands a full thickness resection, histological examination with frozen section control, rather than repeated curettage which enhances orbital invasion,& lymph-node metastasis prior to orbital invasion. Repeated curettage also delays treatment and thus yields a poor prognostic value .

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