Rare disease

Sebaceous carcinoma of eyelid

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Summary

The authors are presenting here, a case of sebaceous carcinoma in a 50-year-old male who presented with 6-month history of a fungating mass in the right eye near medial canthus. Exenteration of eye was done, and a diagnosis of sebaceous carcinoma was made on histopathological examination.

BACKGROUND

Sebaceous carcinoma is one of the rare tumours of eyelid. Sometimes, only inflammation is seen in the tissue, and a small focus of carcinoma is seen as in our case. So careful search in multiple sections is needed to rule out small foci of carcinoma.

CASE PRESENTATION

A 50-year-old male presented with loss of vision and swelling at right upper eye lid for a duration of 6 months. General and systemic examinations were unremarkable. On ocular examination, no light perception was present. Local examination revealed a fungating mass over the right

upper eyelid measuring 3×3 cm near the medial canthus with damage to ocular tissue. Cornea was not identifiable. A clinical differential diagnosis of squamous cell carcinoma eyelid, preseptal cellulitis and lacrimal gland neoplasm was made. Exenteration of eye was done. Gross examination showed multiple greyish white soft tissue pieces, which altogether measuring 7×3×3 cm. Largest piece was 4×2×2 cm in size and smallest 1×1×1 cm. Few pieces were skin covered. Microscopic examination revealed few small foci of tumour cells with surviving lobules of tumour cells with large vesicular nucleus, prominent nucleoli and foamy multivacuolated cytoplasm (figure 1A,B). Exuberant acute on chronic inflammation with foreign body giant cell

Figure 1 (A) Section showing small foci of tumour cells (H&E ×50). (B) Section showing tumour cells with large vesicular nucleus and multivacuolated cytoplasm (H&E ×500).

Figure 2 (A and B) Section showing mononuclear cell infiltrate and giant cells (H&E ×50).

reaction and large areas of necrosis was seen (figure 2A,B). Skins, sclera with choroid and lacrimal gland tissue were free from tumour cells (figure 3A–C). Optic nerve was free from tumour cells (figure 3D). On the above finding, a diagnosis of sebaceous carcinoma was rendered. Patient is well till now (10 months since surgery).

DIFFERENTIAL DIAGNOSIS

- ▶ Basal cell carcinoma
- ▶ Squamous cell carcinoma
- ▶ Cutaneous horn
- ▶ Merkel cell carcinoma
- ▶ Pyogenic granuloma.

TREATMENT

Exenteration of eye was done.

OUTCOME AND FOLLOW-UP

Patient was discharged in good condition.

DISCUSSION

Sebaceous carcinoma is a rare and aggressive tumour, first described by Fuchs. It arises from the meibomian glands of the tarsal plate, from glands of Zeis or from sebaceous glands of the caruncle, eye brow or facial skin.¹ It can be seen anywhere on the body where sebaceous gland exist.^{2–4} Most common site of involvement is periocular area.^{5–6} It is commonly seen in sixth and seventh decade of life; however, it can be seen in any age group. It is the fourth common malignancy after basal cell carcinoma, squamous cell carcinoma and melanoma, and comprises 1–5.5% of malignant tumours of eyelid.^{7–9} Aetiology is unknown till now, but few authors made a correlation with ionising radiation for

other neoplasms, for example, cavernous haemangioma, barber's itch and retinoblastoma.^{10–12} Association with human papilloma virus and overexpression of TP53 is also reported by few authors.¹³ Clinical features include firm, painless, indurated mass or ulceration associated with loss of eyelashes. Clinical picture may mimic benign condition. Clinically, the differential diagnoses include basal cell carcinoma, squamous cell carcinoma, cutaneous horn, Merkel cell carcinoma, pyogenic granuloma and metastatic carcinoma of skin and chalazion. Cytomorphological features of sebaceous carcinoma include sheets, clusters and or singly scattered polygonal cells having centrally placed hyperchromatic nuclei and vacuolated cytoplasm.^{14–15} Microscopic examination shows lobules of cells with multivacuolated clear cytoplasm, raised nucleocytoplasmic ratio, clumped chromatin and prominent nucleoli. This is an aggressive tumour as the local recurrence and metastasis to other sites are very common. Liver, lung, bones and brain are the site of distant metastasis.^{7–16–17} Surgical excision of tumour is the treatment of choice. Many surgeons prefer Mohs technique. In case of orbital involvement, treatment of choice is orbital exenteration as in our case.¹⁸ If orbit does not appear to be involved, the treatment of choice is excision of tumour along with 5–6 mm of normal tissue.¹⁹ Chemotherapy may be given preoperatively to reduce tumour size. Role of radiotherapy is unclear. Most of the surgeons give preference to surgical removal as the observed chances of recurrences following give preference to surgical excision of tumour and the observed chances of relapse following radiotherapy is quite high.²⁰ Few authors have reported good results with concurrent administration of radiotherapy with surgical removal.²¹ Radiotherapy may be given to those patients who refuse surgery. Features of poor prognosis include orbital invasion, poor

Figure 3 (A) Section showing stratified squamous epithelium and underlying mononuclear cell infiltrate (H&E $\times 50$). (B) Section showing sclera and choroid free from tumour cell (H&E $\times 50$). (C) Section of lacrimal gland with no tumour cell infiltration (H&E $\times 50$). (D) Section of optic nerve free from tumour cells (H&E $\times 50$).

differentiation, tumour diameter of more than 10 mm and lacrimal gland involvement.^{22 23} Tumours of the lower eye lid have been associated with better prognosis than those affecting the lower lid.²⁴

Learning points

- ▶ This tumour can arise in any organ where sebaceous gland exists.
- ▶ Clinical presentation may mimic chalazion.
- ▶ It is an aggressive tumour, and any delay in diagnosis may result in early metastasis.

Competing interests None.

Patient consent Obtained.

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