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Case Report

Sebaceous carcinoma of lacrimal gland with endophthalmitis

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ABSTRACT

Sebaceous carcinoma (SC) is a rare type of cancer that aggressive which develops from the sebaceous glands and can impair patients' visual acuity. It usually arises from the periorbital area, which commonly affects in the eyelid. This study discusses the sebaceous carcinoma of lacrimal gland with endophthalmitis. The patient was a 53-year-old Javanese man came with a chief complaint of no vision in his left eye, painlessness nowadays, no previous medical history, and slowly progressive swelling mass on the outer aspect of the left upper eyelid for a year duration. On examination, there was no light perception for the left eye and 6/7, 5 on the right eye. The tumor was rock hard and fixed on palpation, which caused partial upper eyelid ptosis, displaced the globe anteromedially, and impaired ocular motility. There was also a massive chemosis in the left eye, ptosis with corneal damage caused by endophthalmitis. The histopathological examination found that the tumor was composed of nests of cuboidal and columnar cells with proliferation round core cells, pleomorphic, hyperchromatic, vacuole cytoplasm, and prominent nuclei, arranged in a solid structure without lymphovascular invaded. The final diagnosis was sebaceous carcinoma of the lacrimal gland. The patient took tumor excision surgery followed by orbital evisceration due to endophthalmitis with post-operative radiotherapy as an alternative treatment. Sebaceous carcinoma of the lacrimal gland is extremely rare. There are no specific treatments, and it is difficult to study further for the best treatment due to its rare incidence. Sebaceous carcinoma of the lacrimal gland has so far been treated best by surgical excision and radiotherapy after.

Keywords: Sebaceous carcinoma, Tumor, Lacrimal gland

INTRODUCTION

Tumors of the lacrimal gland show about 5% to 25% of orbital tumors and can be subdivided into lymphoproliferative, benign epithelial, malignant epithelial, malignant nonepithelial lesions and malignant tumors account for 25% of all lacrimal gland neoplasms.[1,2] The most common epithelial malignancy is primary adenoid cystic carcinoma (ACC). Therefore, most studies on epithelial tumors of the lacrimal gland have been conducted predominantly on ACC. Among lacrimal gland tumors, sebaceous carcinoma of the lacrimal gland (SCLG) is immensely rare. However, it has been reported in Asian populations sebaceous carcinoma is the second most common malignant eyelid tumor and about 22%-28% of all orbital space-occupying lesions, moreover 20-50% of these tumors are malignant.[3-5]

Sebaceous carcinoma usually emerges from the periorbital area, which commonly affects in the eyelid. Sebaceous carcinoma of the orbit is commonly caused by orbital invasion of the periorbital carcinoma or by metastasis of tumors from other body parts. It is known to be aggressive, so delay in diagnosis can increase the chance of local recurrence and metastasis. Therefore, early diagnosis and accurate examination of the eyelids is essential to confirm a diagnosis and to prevent the metastasis.[3]

CASE REPORT

The patient was a 53-year-old Javanese man came with a chief complaint of no vision in his left eye, painlessness nowadays, no previous medical history, and slowly progressive swelling mass on the outer aspect of the left upper eyelid for a year duration. During the first five months of mass growth, the patient felt severe pain and mass in the upper left eye with no other specific complaints. The examination of the visual acuities found no light perception for the left eye and 6/7, 5 on the right eye. The tumor was rock hard and fixed on palpation, which caused partial upper eyelid ptosis, displaced the globe anteromedially, and impaired ocular motility. No pre-auricular or cervical lymph nodes were palpable.

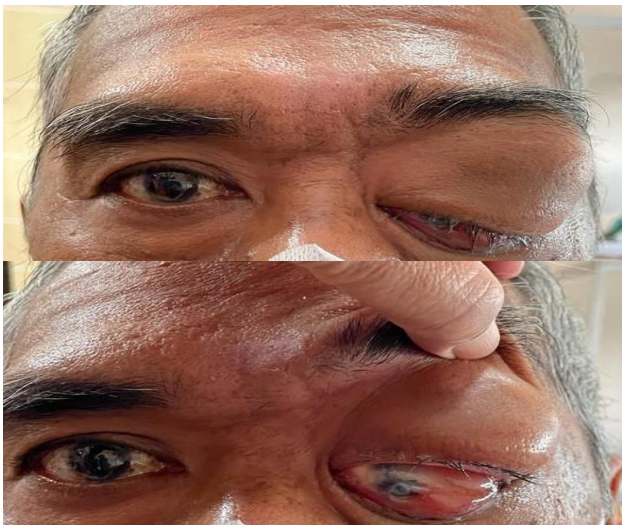

Figure 1: A 53-year-old man presented with a painless massive swelling of the left upper eyelid.

There was also a massive chemosis in the left eye, ptosis with corneal damage caused by endophthalmitis. Other ophthalmic examination results were unremarkable. The patient underwent computed tomography (CT) scan and chest radiograph examination, which revealed enhanced and infiltrative mass at the left superotemporal orbit over the superotemporal quadrant of the left orbit measuring 4×2×3 cm with no orbital erosion and chest radiograph shows both lungs are clear, no infiltrates, bone, diaphragm, soft tissues and cardiomediastinal contour is normal. Pathology of biopsy with fine needle aspiration biopsy (FNAB) showed a lacrimal gland carcinoma.

The patient then received orbital evisceration after a week of FNAB results and underwent a tumor excision. The gross examination revealed a firm white tumor measuring 5×2×3 cm. Then histopathological examination found that the tumor was composed of nests of cuboidal and columnar cells with proliferation round core cells, pleomorphic, hyperchromatic, vacuole cytoplasm, and prominent nuclei; arranged in a solid structure without lymphovascular invaded. The final diagnosis was sebaceous carcinoma of the lacrimal gland.

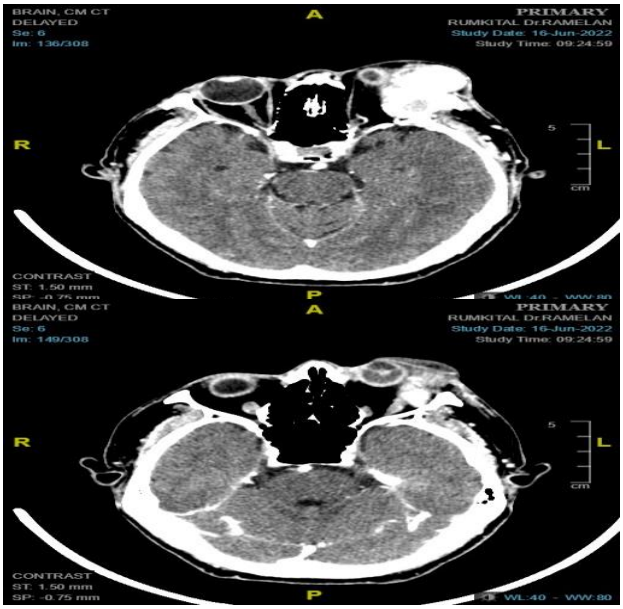

Figure 2: Computed tomography images of the orbit revealed enhanced and infiltrative mass at the left superotemporal orbit.

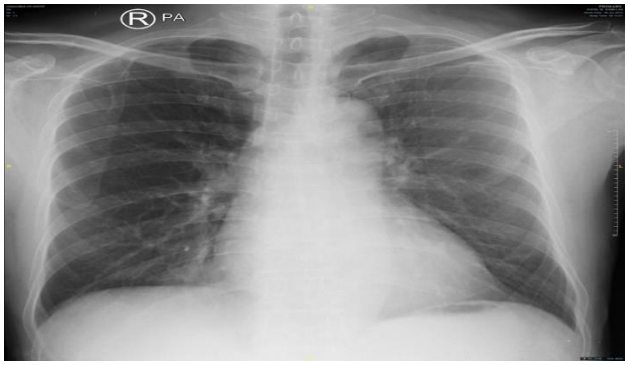

Figure 3: Chest radiograph shows both lungs are clear, no infiltrates, bone, diaphragm, soft tissues and cardiomediastinal contour is normal.

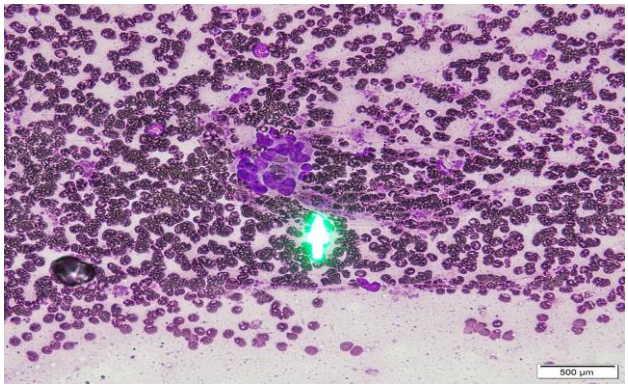

Figure 4: Microscopic findings with FNAB showed the tumor was composed of nests of cuboidal and columnar cells with proliferation round core cells, pleomorphic, hyperchromatic, vacuole cytoplasm, and prominent nuclei.

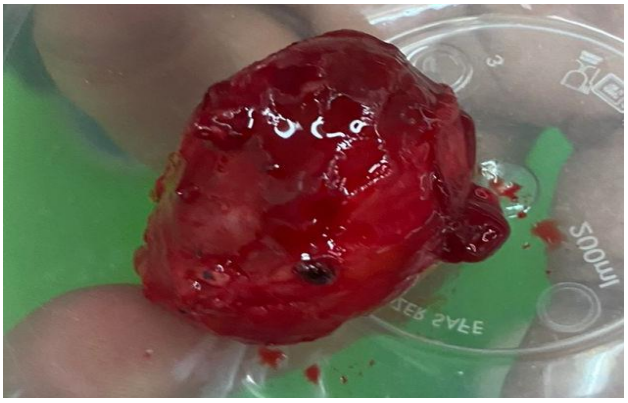

Figure 5: Tumor excision.

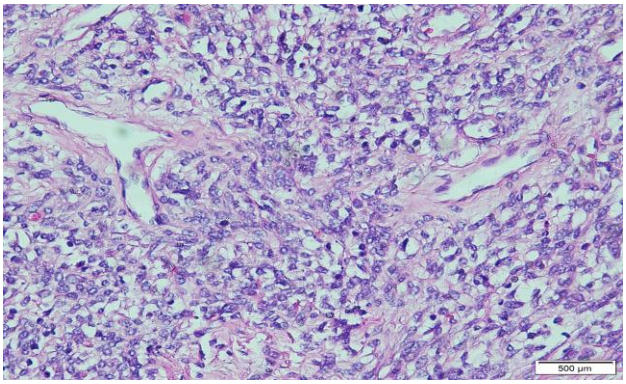

Figure 6: A microscopic view with histopathological examination.

The patient also took an orbital evisceration, then the patient was observed for two weeks after the surgery. In the first week after surgery, the hematoma and oedema were still seen on the upper eyelid. However, there were no chemosis and swelling of the lymph node. After two weeks of operations, the hematoma was gone, and still no swelling of the lymph node. Radiotherapy was suggested for further treatment.

DISCUSSION

Sebaceous carcinoma is one of the rarest tumors of lacrimal gland, in some cases this type of tumors begins in the skin. SC occurs commonly 80% within the head and neck, and 40% within the ocular region, commonly in the upper lid due to more sebaceous glands there and frequently considered as either ocular or extraocular.[3,4] Although sex reports have varied, most authors found that SC is more frequent in older women.[6,7] SC commonly arises in patients over the age of 40 or presents within the elderly (average of 70 years of age), and the peak incidence arises in the seventh and eighth decades of life.[6,8,9]

The mortality rate has been approximated to be over 20%, and the survival rate for sebaceous carcinoma has been reported to reach 91.9% at five years and 79.2% at ten years.[10]

Sebaceous carcinoma originately comes from sebaceous cells in the periocular region. SC usually emerges from Meibomian glands, which are modified sebaceous glands, but moreover from the sebaceous glands of Zeis and the sebaceous glands in the ocular caruncle. Sebaceous carcinoma emerges from de novo mutations and may also develop from benign neoplasmas.[7,11,12] The cause of sebaceous carcinoma in the lacrimal gland remains unknown, but it is presumed several hypotheses that the tumor may likely arise from pluripotent cells that have differentiated into sebaceous cells.[10,13] Also several things reported to become the risk factors include elderly ages, radiation, immunosuppression, and genetic predisposition for Muir–Torre syndrome.[4,9,14]

Sebaceous carcinomas' histopathology fall four histopathologic categories: papillary, lobular, comedocarcinoma, and mixed. Malignant cells show significant pleomorphism, hyperchromatism, mitotic activity, and nuclear atypia.[14] Park et al also found malignant cells mixed with sarcomatous, adenocarcinomatous, and pleomorphic sarcomatoid components in their patient's histopathology.[3] Our patient's histopathology showed identical results: the tumor was composed of nests of cuboidal and columnar cells with proliferation round core cells, pleomorphic, hyperchromatic, vacuole cytoplasm, and prominent nuclei.

Sebaceous carcinomas' masses typically develop as a painless, firm, solitary nodule, round, enlarging nodule, or diffuse pseudo inflammation. The more common painless solitary nodule presents as a firm subcutaneous lesion that arises in and is fixed to the tarsus or appears in the eyelid margin when it emerges from the Zeis gland. This tumor may involve the fornical and bulbar conjunctiva that may cause blepharitis or conjunctivitis and increased eyelid skin laxity which may disguise as a chalazion and typically mimicking other conditions which can lead to a misdiagnosed.[4,9]

In this case, the patient's symptoms and clinical manifestations were mainly representatives of a lacrimal gland tumor. However, it was necessary to confirm which lacrimal gland tumor the patient had. To see how far and massive the tumor is, the patient took computerized tomography imaging and chest radiograph, which revealed an infiltrative orbital mass over the superotemporal quadrant of the left orbit measuring 4×2×3 cm with no orbital erosion and a normal chest radiograph. However, it still raises suspicion about whether the tumor was malignant or not. Therefore, an incisional biopsy might be necessary to confirm the diagnosis. Excisional biopsy is considered the gold-standard treatment and in confirming the diagnosis.Thus, the tumor must be confirmed again by histopathological examination, revealing similar results to the theories.[7,14] The patient's microscopic view with histopathological examination showed that the tumor was composed of nests of cuboidal and columnar cells with proliferation round core cells, pleomorphic, hyperchromatic, vacuole cytoplasm, and prominent nuclei;

arranged in a solid structure without lymphovascular invaded. This result suggested that the patient's diagnosis was a sebaceous carcinoma of the lacrimal gland.

Local spread is numerous and have been reported as 26-51%. Local spread may comprise broad areas of the conjunctiva and moreover the cornea. Advanced infiltration may cause invasion of the orbit causing an ocular irritation, diffuse eyelid thickening and adjacent periorbital structures.[14,15] According to this, the patient was suspected of having a massive inflammation caused by endophthalmitis. The patient was not aware of his eye and did not receive any treatment for the eye inflammation. This could be attributed to delayed diagnosis and treatment that may cause a massive inflammation leading to endophthalmitis.

Although the optimal treatment of SC is challenging to study due to its rare incidence, treatment of lacrimal gland carcinoma has been viewed by an local excision as the most common standard approach. Our patient underwent a tumor excision based on the consideration with strongly suspected malignancy of sebaceous carcinoma based on histopathology results and the most common treatment so far for SC.[7,16] The patient also underwent an evisceration due to no light perception in his left eye, leading to a poor prognosis of eye preservation caused by endophthalmitis.[17]

Other alternative teartment for SC may considered to take a radiotherapy. Radiotherapy recommended because some studies reported local recurrence after the surgical excision.[11,14,16] It has been reported that SC tend to be aggressive with potential nodal and distant metastasis. Regional lymph nodes are the most common sites of spread; lungs, brain, liver, small intestine, and urinary tract metastasis have also been reported.[14,18,19] However, another previous literature study by Park et al reported only one patient among 29 patients with extraocular sebaceous carcinoma showed regional lymph node involvement and no distant metastasis or death during the follow-up period was observed.[20] Although there are different results on whether there is lymph metastasis or not, radiotherapy achieves the best treatment effect, and radiation can be administered with superior tumor control and relative safety when the cancer is adjacent to a vital organ. Also, several institutes use particle radiotherapy for improved tumor control and toxicities in treating tumors of the lacrimal gland.[4,11,19] The patient was suggested to take radiotherapy for further treatment after the surgery. Then we followed the patient for two weeks after the surgery. In the first week after surgery, the hematoma and oedema were still seen on the upper eyelid, but no chemosis and swelling was observed of the lymph node. Wound debridement was also performed one day after the surgery and once a week. We also educated the patient and his family to take care of the wound dependently for homecare treatment. In the second week, the hematoma was gone, and there was still no swelling of the lymph node. Radiotherapy was then recommended for further treatment.

CONCLUSION

Sebaceous carcinoma is a rare type of aggressive cancer which develops from the sebaceous glands and can impair patients' visual acuity. There are no specific examination and treatment and this tumor may mimic other diagnosis, hence proper examination and treatment are needed. We applied the suggested studies to take local exicision for the tumor and found that our patient was in stable condition and had no evidence of metastases for two weeks. Also, our patient considered to take a radiotherapy due to the recurrence risk and underwent an orbital evisceration. However, accurate and early diagnosis is essential for appropriate treatment and a moderate mortality rate. Therefore, further studies or treatments including radiotherapy are needed.

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