Case Report

Giant conjunctival melanoma in a paranoid schizophrenic man: A case report

Tommy Supit¹,*, Pujisriyani ², Subiyakto ³, Trilaksana Nugroho ⁴, Alifiati Fitrikasari ⁵, Najatullah ⁶

¹ Department of General Surgery, Faculty of Medicine Diponegoro University, Dr. Kariadi General Hospital, Jl. Dr. Sutomo No. 16, Randusari, Semarang Selatan, Semarang, Jawa Tengah, 50244, Indonesia
² Department of Plastic Surgery, Faculty of Medicine Diponegoro University, Dr. Kariadi General Hospital, Jl. Dr. Sutomo No. 16, Randusari, Semarang Selatan, Semarang, Jawa Tengah, 50244, Indonesia
³ Department of Oncologic Surgery, Faculty of Medicine Diponegoro University, Dr. Kariadi General Hospital, Jl. Dr. Sutomo No. 16, Randusari, Semarang Selatan, Semarang, Jawa Tengah, 50244, Indonesia
⁴ Department of Ophthalmology, Faculty of Medicine Diponegoro University, Dr. Kariadi General Hospital, Jl. Dr. Sutomo No. 16, Randusari, Semarang Selatan, Semarang, Jawa Tengah, 50244, Indonesia
⁵ Department of Psychiatry, Faculty of Medicine Diponegoro University, Dr. Kariadi General Hospital, Jl. Dr. Sutomo No. 16, Randusari, Semarang Selatan, Semarang, Jawa Tengah, 50244, Indonesia

ARTICLE INFO

Keywords:
Cancer
Orbital
Conjunctival
Melanoma
Paranoid
Schizophrenia

ABSTRACT

Introduction: and importance: Conjunctival melanoma (CM) is a rare and potentially lethal ocular tumor. As with any oncologic disease, early diagnosis and appropriate treatment of CM is paramount to limit morbidity and increase life expectancy. However, patients with severe mental disability with social isolation are usually presented in late-stage disease.

Case presentation: This report presents a case of a 55-year-old man with paranoid schizophrenic man with an extraordinarily large CM due to neglect. The patient suffered from complete left eye blindness with no clinical and radiological evidence of metastasis.

Clinical discussion: Clinicians must bear in mind the limited patient compliance and family support of mentally-ill patients that restricts treatment modalities that would have otherwise been applicable for cooperative patients. The importance multidisciplinary approach, choosing the simpler but effective surgical technique should be prioritized.

Intervention and outcome: Left exenteration and tumor wide excision was performed. The left orbital defect was reconstructed using forehead flap and split-thickness skin graft (STSG). The uncooperative nature of the patient posed early post-operative challenges that necessitates subsequent operation to drain seroma. The patient was discharged 16-days after operation with acceptable cosmetic and clinical results. However, the patient failed to return to the clinic for longer post-operative evaluation.

Conclusion: A multidisciplinary approach is mandatory to treat complex cases such as this report. Surgeons are advised to adopt simpler surgical approach that will require minimal self-care and should encourage family members to continuously support the patient.

1. Introduction

 Conjunctival melanoma (CM) is an uncommon tumor comprising 2% of all ocular tumors and 0.25% of all melanomas [1]. The risk factors for CM are not yet established however, the incidence of CM is increasing along with cutaneous melanoma, which suggests a possible association with ultraviolet light exposure [2]. The molecular pathogenesis of CM is more similar to cutaneous melanoma compared to its uveal counterpart. A study using exome sequencing detected mutations in BRAF, NRAS, NF1, EGFR, ALK, TERT, and APC oncogenes in CM [3]. Patients with CM usually complain of a painless brownish lesion on the surface of the eye, and occasionally ocular irritation or pain [4]. Differential diagnosis of
CM include primary acquired melanosis, conjunctival nevi, local
extension of uveal melanoma or melanocytoma, and distant metastasis
of cutaneous melanoma [5]. This care report is reported in line with the
surgical case report (SCARE) guideline [6].

Primary treatment of CM involves wide surgical excision followed by
adjuvant therapy (i.e. cryotherapy, topical alcohol application, brachytherapy). However, systemic metastasis occurs in 19% of the
patient within 3.4 years with no effective treatment for metastatic dis-
ease [5]. Disease recurrence, involvement of non-bulbar conjunctiva,
medial bulbar conjunctiva, caruncle and plica semilunaris, tumor
thickness of more than 2 mm, de novo origin, and nodular growth pattern are all risks for metastasis and mortality [5]. We present a neglected case
of CM in a paranoid schizophrenic allowing it to grow into a very large
size. We report a case of CM presented in a late-stage in a paranoid
schizophrenic man with minimal self-care and family support. The
importance of multidisciplinary approach, choosing the practical sur-
gical approach, and the challenges in treating mentally ill patient are
discussed.

2. Case report

A 55-year-old male presented in the outpatient clinic with a large and
foul-smelling tumor growing out of his left eye since a year ago. The
mass was initially a small black nodule on the temporal side of the
limbus. Other than complete blindness of the left eye, there were no
symptoms related to central nervous, cardiorespiratory, or gastrointes-
tinal system. History ocular trauma, excessive sun exposure, and family
with malignancy was denied. Physical examination revealed a baseball-
sized irregular mass with dark brown to greyish discoloration protruding
out of the left eye socket. The fungating tumor surface was partially
covered with necrotic tissue, slough, and blood clot. Computed tomog-
raphy (CT) scan with contrast demonstrated a mixed density mass with
antero-posterior diameter of 7.7 cm and latero-lateral diameter of 7.8
cm, phthisic left globe, extra cavitary stretching of optic nerve, ophthalmic vein, and extraocular muscles (Fig. 1). There was no evidence of
distant metastasis from head neck CT-scan, and pulmonary
plain radiography.

The patient was reported to display an increasingly abnormal
behavior within the last two years with reports of disorganized speech,
self-talk, and difficulty in communication. The patient mostly lived
alone or by the care of his siblings after his spouse and children left him.
There was no history of medical treatment for the tumor or mental
illness. The dominant symptoms at the time of admission and during
hospital stay were auditory hallucinations, suspicious behavior, and
delusion of persecution. The psychiatric assessment confirmed the
diagnosis of paranoid schizophrenia (International Classification of
Mental and Behavioral Disorders [ICD]-10 Code F20.0). He was pre-
scribed Olanzapine 10 mg daily, Amitriptyline 100 mg daily, Trihex-
phenidyl 1 mg daily, and Diazepam 15 mg daily along with routine
psychotherapy throughout the hospital stay.

The patient underwent left exenteration and wide excision that
include the eye globe, eyelids, retrobulbar soft tissues, and periostium
with tumor-free margin confirmed by intraoperative frozen sections
(Fig. 2). The left orbital defect was reconstructed using forehead flap and
split-thickness skin graft (STSG) harvested from the left lateral thigh to
cover the forehead wound (Fig. 3A). The surgery was performed board-
certified specialists: one ophthalmologist, one oncologic surgeon, and
two plastic surgeons. Pathologic analysis confirmed the diagnosis of
invasive conjunctival melanoma (ICD-Oncology code 8720/3) with evi-
dence of tumor invasion to the retrobulbar vasculature and fat tissue.
Frozen sections revealed clear resection margins thus, no nonexcisional
adjuvant therapy was applied. To facilitate postoperative drainage, a
single 16 French tube was inserted at the posterolateral side of the left
orbit.

However, 2 days later it was forcefully pulled out by the patient
caused by his paranoid nature. Bilateral arm restraints were applied and
additional intravenous Diazepam 10 mg was administered as necessary.
Serohemorrhagic fluid was observed to be oozing from the previous
drain tract saturating the gauze, requiring daily change. Bulging of the
distal flap covering the orbit was noted one-week post-operation
necessitating seroma drainage. The drainage was performed twice at day
10 and day 15 before the patient was finally discharged 16 days post-
operation under stable psychiatric condition. Post-operation at day 21
showed vital forehead flap and STSG with satisfactory cosmetic results
(Fig. 3B). Regrettably, this is the latest information we had regarding his
postoperative condition since the patient only managed to came once for
postoperative control 7 days after discharge (21 days post-operation).

3. Discussion

A meta-analysis suggests the standardized incidence rate of mela-
noma in a patient with schizophrenia was significantly lower (0.71)
compared to the healthy population [7]. Excess dopamine, enhanced
natural killer cells, increased apoptosis rate, and modulation by anti-
psychotic drugs were some of the proposed hypotheses for this finding
[8]. There is a trend for psychiatric patients with cancer to present later
and with more advanced disease than the general population [9]. They
seek medical help when symptoms become more pronounced in the later
stages of the disease.

It is extremely rare for CM to be left to grow into very large because
of its obvious early clinical signs. To our knowledge, this is the first case
report CM with such magnitude. The necrotic tissue surrounding the
tumor can be a result of cancer cell ischemia due to the rapid expansion

![Fig. 1. A large irregularly shaped tumor protruding from the left eye socket with outstretched extraocular muscles. The phtisic left globe cannot be visualized from outside.](image1)

![Fig. 2. Left orbital defect post exenteration and wide excision revealing an empty eye socket with no bone defect.](image2)
of the tumor unsupported by angiogenesis. While the presence of noisome and air-bubble density within the tumor suggests an ongoing infection process. The patient presented with poor performance status with neglected self-care and with no supportive family members, and the condition is expected to resume throughout the duration of treatment and recovery.

This should alarm the clinicians to opt for the treatment modality that is safe and perhaps simpler for the patient to personally take care of. As in our case, efforts in contacting the patient and his family members had failed as of the creation of this manuscript. This predicament highlights the inherent challenge of treating oncoplastic patients with mental illness with limited family support.

The mainstay treatment of CM is wide local excision with "no-touch" technique followed with cryotherapy. Exenteration is justified for a large tumor with orbital and complete conjunctival invasion [4]. The postoperative defect, in this case, is what Kesting et al. described as type I defects where the resection involves the whole orbital content and periosteum without bony resection [10]. They suggested orbital coverage with STSG that can be followed by abutment placement and application of oculoplastic prosthesis [11]. We preferred forehead flap and STSG to cover the donor defect because it is a relatively quick procedure with acceptable outcomes.

Furthermore, it requires less demanding postoperative care compared to free flap and more durable for future external beam radiotherapy. The seroma formation within the hollowed orbit would have been mitigated with the use of tube drainage. However, it is evident from this case that its use is less reliable in an uncooperative patient. Temporalis muscle flap to fill in the empty eye socket would have been an excellent technique for this problem. Radial forearm flap is another alternative however, it is less ideal for this case since it is our best interest to create a wound that requires simple care.

4. Conclusion

Oncologic patients with psychiatric illness are usually presented with a more advanced stage disease. The main challenge in treating this group of patient lies in their level of compliance and self-care. Comprehensive treatment will be very hard to achieve without a multidisciplinary approach and the aid of supportive family members. Surgeons are advised to adjust their clinical decision based on the patient’s performance status and to opt for the surgical technique that will allow optimal wound care.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Consent

Written informed consent obtained from the patient for the purpose of publishing this case report. Information within the paper has been sufficiently anonymized not to cause harm to the patient or his family. A copy of signed informed consent is available for review by the Editor-in-Chief of this journal on request.

Sources of funding

Nothing to declare.

Ethical approval

Ethical approval exempted by our institution.

Author contribution

Tommy Supit and Najatullah conceptualized the paper, performed perioperative patient care, follow-up, and manuscript drafting. Pujisriyani, Subiyakto, Trilaksana Nugroho performed the operation, validation of clinical history, investigations, and operative findings. Alifiati Fitrikasari performed psychiatric assessment, perioperative care and follow-up. All authors were involved in manuscript writing and approval for submission.

Research registration

N/a.

Guarantor

Tommy Supit
Department of General Surgery, Faculty of Medicine Diponegoro University, Dr. Kariadi General Hospital, Semarang, Indonesia.

Declaration of competing interest

None declared.
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