

ADVANCED UVEAL MELANOMA WITH SUBDURAL METASTASIS MIMICKING MENINGEOMA - A CASE REPORT

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Uveal melanoma is a rare malignancy. Its clinical course is highly aggressive. At the time of diagnosis, extraocular extension is present in most of the cases.

We present a case of 69-year-old white man admitted for sharp orbital pain. Advanced uveal melanoma was diagnosed. We found black-colored tumor protruding from the left eye and multiple cutaneous metastases on the scalp. CT scan revealed intracranial tumor mimicking meningeoma in the left parietal region. Lymphogenous metastases were not found and other hematogenous metastases were excluded. After biopsy of the eye tumor and excisional biopsy of one skin tumor, the uveal melanoma was diagnosed and the left orbital exenteration and extirpation of intracranial tumor were performed. The reconstruction was performed using galeacutaneous flap harvested from craniotomy flap. Postoperative course was uneventful and the patient was released from pain. He refused the additional oncological treatment. After four months, he died of liver metastatic disease.

The uveal melanoma is highly aggressive malignancy and isolated subdural metastasis is quite rare. The reconstruction with transposed galeacutaneous flap is versatile and secure technique after orbital exenteration. *Acta Medica Medianae* 2011;50(1): 47-50.

Key words: uveal melanoma, orbit, orbital exenteration, reconstruction

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Introduction

Melanoma is highly aggressive tumor. More than 95% of all ocular melanomas occur intraocularly, arising from melanocytes in the uvea. The uvea is a vascular tissue consisting of: choroid, ciliary body and the iris. About 90% of uveal melanomas involve choroid. It is a rare tumor and the incidence increases with age reaching peak at about sixty years of age (1,2). In the early stage uveal melanoma can cause impairment of vision, but in advanced stage the eye is blind and painful as a result of retinal detachment, glaucoma and inflammation.

Case report

A 69-year-old white man was admitted suffering sharp pain in the left orbit. Patient had perforative wound in the left eye at the age of 19, resulting in permanent loss of vision at that side. He observed a tumor in his left eye two years before admittance but not referred to

ophthalmologist. Six months before admittance, he found black nodes on the scalp. He had high blood pressure for 13 years. The patient was moderate smoker (20 cigarettes daily) and denied alcohol abuse.

Clinical examination revealed dark brown to black tumor protruding from the left eye (Figure 1) and multiple (18) nodular black tumors sized 3mm to 1cm in the scalp skin. These scalp skin tumors were not fixed to deeper layers. (Figure 2). Moderate exophthalmus and limitation of eye movements was found.

An incisional biopsy of the left eye tumor and excisional biopsy of one skin tumor in frontoparietal region were performed under local anesthesia. Histopathological examination revealed uveal melanoma and skin metastasis of melanoma. CT scan of orbits and brain was performed and presented advanced tumor destroying the left eye and infiltrating muscles, intraconal and extraconal fat. CT scan of the brain revealed solitary tumor in the subdural space in the left frontoparietal region sized 5x3x1,5cm without infiltration of the brain tissue mimicking meningeoma according to its CT features (Figure 3). The orbital exenteration and simultaneous pterion craniotomy for tumor extirpation were performed. The postoperative orbital defect was covered using frontal flap based on anterior branch of superficial temporal artery (Figure 4). Secondary defect was covered by galeacutaneous flap

intraoperatively expanded by incision of galeal layer of rotational flap. Those incisions were paralel and made from the basis of flap to incision line. Skin nodes on the scalp out of the created flap were excised (Figure 5).

The skin incision in the left frontoparietal region and pterion craniotomy were performed. The tumor was removed from subdural space in frontoparietal region and dura was closed. The neurosurgical procedure was completed with the drainage of epidural space, fixation of calvarian bone and suture of periost.

The skin was incised in the line of orbital rim and orbital exenteration was performed. After adequate hemostasis defect was covered using frontal galeacutaneous flap based on the anterior branch of superficial temporal artery and the drain was placed in the orbit.

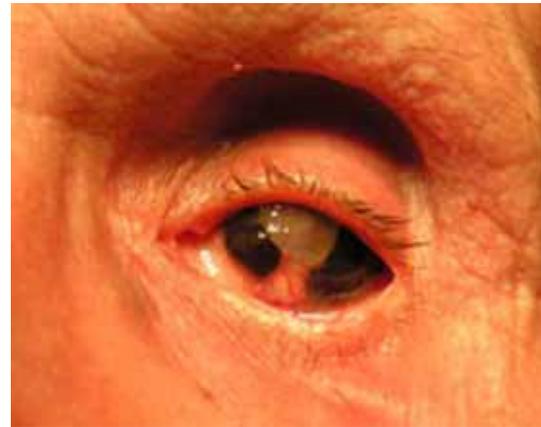


Figure 1. Uveal melanoma of left eye



Figure 2. Metastases in head skin



Figure 4. Schematic view on flaps: 1 galeacutaneous flap for orbital coverage, 2 galeacutaneous flap for scalp reconstruction, 3 anterior branch of superficial temporal artery, 4 posterior branch of superficial temporal artery, 5 retroauricular artery, 6 longitudinal galeal incisions.

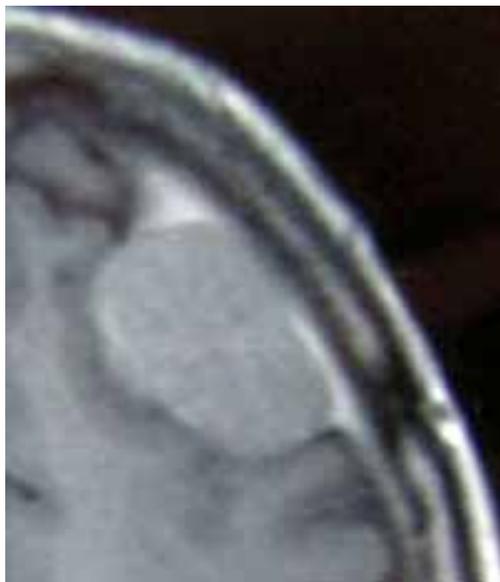


Figure 3. Computer tomography coronal view with subdural tumor



Figure 5. Final result after reconstruction

There were no wound complications. Drains were removed on the 4th postoperative day and the patient was discharged on 8th postoperative day. He refused further surgical treatment for skin metastases and refused additional oncological treatment. Four months after operation patient died with liver metastases.

Discussion

Uveal melanoma is very rare and not covered in literature with large clinical studies data. In one third of cases, the tumor is asymptomatic being discovered during routine ocular examination. The disease can mimic vascular anomalies or be diagnosed with other pigmented lesions (3,4). It may be diagnosed in phase of symptomatic disease, as in our case (5). Diagnostic standard is CT scan of orbit for staging of primary tumor. Uveal melanomas metastasize by hematogenous and/or lymphatic spread. Because the metastatic spread is often found at the first visit an adequate clinical examination including thorax X-ray and liver ultrasound must be performed. Hematogenous spread of the disease is more frequent than lymphatic. Lymphogenous metastases may be found in parotid and cervical nodes (6). Metastatic disease involves the liver in more than 90% of patients. Other relatively common sites being skin, lung, and bone. Most patients with metastases die within a year of the onset of symptoms (1,2).

The present TNM (Tumor, Node and Metastasis) staging system is based on basal tumor diameter, tumor thickness, ciliary body involvement and extraocular spread, but it is not widely used (7). The therapy for small melanomas in most centers is brachytherapy with iodine or ruthenium plaque (8). Primary enucleation now tends to be performed only if the tumor involves optic nerve or if it is very large, exceeding 17mm in basal diameter. The conjunctival melanoma, the other group of orbital melanoma, could also be the indication for orbital exenteration (9).

Treatment of choice is surgery for large primary tumor, for metastatic disease in lymph nodes and for solitary hematogenous metastases in visceral organs (eg. liver metastasectomy). Lymphogenous metastases in parotid gland should be treated with total parotidectomy and positive neck lymph nodes with radical neck dissection. Treatment of hematogenous metastases depends on their site and size. Multiple hematogenous cutaneous metastases could also be treated by surgery. Postoperative treatment can include immunotherapy, chemotherapy or irradiation. In terminal phase of the disease, therapy is only supportive and symptomatic (9).

Screening for metastases is performed for detecting resectable hepatic metastases (1,2). Methods include liver imaging, such as ultrasonography, and liver function tests in blood, which are performed every six months (10).

The treatment of choice is orbital exenteration and reconstruction using local flaps or temporal muscle flap and skin grafts. The reconstruction in this case with galeacutaneous transposition flap for orbital coverage could be useful as safe surgical option. For lymphatic metastases parotidectomy and neck dissection are preferred. Solitary metastases in viscera can be surgically removed. Solitary metastasis in subdural space can be misdiagnosed as meningioma. The additional irradiation, immunotherapy and chemotherapy can be used in the treatment. Prognosis is poor and survival rate is measured in months.

Conclusion

The uveal melanoma is highly aggressive malignancy and subdural metastasis is very rare. Despite poor prognosis, the sharp pain unresponsive on medication and intracranial metastasis in this case indicated surgery. The reconstruction after orbital exenteration with transposed galeacutaneous flap harvested from craniotomy flap site is versatile and secure technique.

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UZNAPREDOVALI MELANOM SUDOVNJAČE SA METASTAZOM U SUBDURALNOM PROSTORU KOJA LIČI NA MENINGEOM - PRIKAZ BOLESNIKA

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Melanom uveje je redak maligni tumor. Ovaj tumor ima jako agresivan klinički tok, a ekstraokularno širenje je u trenutku postavljanja dijagnoze prisutno kod većine bolesnika.

Bolesnik starosti 69 godina primljen je u bolnicu zbog jakog bola u levom oku. Postavljena je dijagnoza melanoma uveje. Tumor je bio crne boje i prominirao iz leve očne jabučice. Verifikovane su multiple metastaze na koži poglavine. CT pregledom je dijagnostikovao intrakranijalni tumor koji je po CT karakteristikama ukazivao na meningeom. Nisu nađene limfogene metastaze kao ni druge hematogene metastaze. Nakon biopsije tumora oka i ekscizione biopsije jedne kožne promene na poglavini potvrđena je dijagnoza melanoma uvee. Urađena je simultana operacija, egzenteracija leve orbite i eskirpacija intrakranijalnog tumora. Rekonstrukcija defekta orbite urađena je pomoću poglavinско-kožnog režnja koji je podignut iz režnja podignutog u toku kraniotomije. Postoperativni tok bio je bez komplikacija i bolesnik nije imao jake bolove. Bolesnik je odbio terapiju interferonom. Smrtni ishod je nastupio posle 4 meseca zbog metastaza u jetri.

Melanom uvee je visoko agresivan tumor a izolovana subduralna metastaza je veoma retka. Rekonstrukcija defekta posle egzenteracije transpozicionim režnjem poglavine je jednostavna i sigurna tehnika kod ovih bolesnika. *Acta Medica Medianae* 2011;50(1):47-50.

Ključne reči: melanom sudovnjače, očna duplja, egzenteracija, rekonstrukcija