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A 39-Year-Old Woman with Oculodermal Melanocytosis: A Case Report

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A B S T R A C T

Introduction: Nevus of Ota, also known as oculodermal melanocytosis (ODM), is characterized by macular pigmentation which localized to the forehead and periocular area. It usually shows a dermatomal distribution from the first two branches of the trigeminal nerve. We reported a case of oculodermal melanocytosis (ODM). **Case presentation**: A 39-year-old woman, came with complaints of her left eye rolling inwards for the past 5 years accompanied by a flat black spot on her forehead that spread to her left cheek. She was done macular OCT and orbital ultrasonography of the left eye. She was diagnosed with oculodermal melanocytosis in the left eye, moderate NPDR in both eyes, juvenile cataracts in both eyes, and esotropia in the left eye due to left-eye cranial nerve VI paralysis. Follow-up at 6 months was advised to the patient. **Conclusion:** Although this is a benign condition, patients are at risk of developing complications such as increased intraocular pressure and melanoma of the skin and/or uvea, ongoing evaluation with imaging methods is highly recommended even if only skin lesions may be repaired using laser for cosmetic reasons.

1. Introduction

Nevus of Ota, also known as oculodermal melanocytosis (ODM), is characterized by macular pigmentation which localized to the forehead and periocular area. It usually shows a dermatomal distribution from the first two branches of the trigeminal nerve.[1] Approximately 50% of cases occur at birth, while the remainder occur during puberty and adulthood. The exact cause is unclear.[2] Nevus of Ota is most often found in Asian women, with a prevalence of around 0.4 to 0.8% in all dermatology patients in Japan, and is rarely found in Caucasians.[3]

Glaucoma is one of the major ocular complications, and although melanocytosis is benign, the patients are prone to developing uveal, orbital, and brain melanoma. Fundus pigmentation is often rare; thus, multimodal imaging is essential in detecting minimal changes in the retina and choroid for early diagnosis. This improves patient survival, especially because uveal melanoma in ODM has twice the risk of metastasis compared to uveal melanoma in eyes without ODM.[4] The following is reported regarding the Nevus of Ota. Therefore, we reported a case of oculodermal melanocytosis (ODM).

2. Case Presentation

A 39-year-old woman came to the eye polyclinic on December 4th, 2023, consulted by the central neurology department of Dr. Mohammad Hoesin General Hospital, Palembang, with a diagnosis of left hemicephalgia, left cranial nerve VI paralysis, left cranial nerve VII paralysis peripheral type due to suspected aneurysm or dolicoectasia. The differential diagnosis was peripheral neuropathy. She complained

that her left eye has been rolling inward, accompanied by double vision, since 6 months ago. Since approximately 5 years ago, the patient complained of the left eye rolling inward without pain and red eyes. Complaints are getting worse, and the patient feels that the left eye is difficult to move, and there is blurred vision. She denied any pain or headache.

Around 6 months ago, she complained of double vision when looking with both eyes. The patient complains that the left forehead and eyebrows are difficult to lift, numbness, and the left forehead sags to the left cheek. She often complains of headaches that come and go. Nausea and vomiting, seeing a rainbow, seeing like in a tunnel, red eyes, watery eyes, and discharge were denied. There was a history of diabetes mellitus from one month ago. No history of hypertension, using glasses, trauma, similar family history, and previous eye surgery were confirmed.

The patient looks normal. Her vital signs were normal. Visual acuity of 6/30 ph 6/7.5; normal intraocular pressure; cloudy lens; normal papil, macula, and retina on the right eye. There was a possibility of left eye cranial nerve VI paralysis. The left eye's visual acuity was 6/60 ph (-) in the nasal direction (ET 45^{o}OS). Scleral hyperpigmentation, cloudy lens, and retinal hyperpigmentation on three quadrants were found on the left eye. Neurological examination found forehead and left cheek hypesthesia, asymmetrical forehead wrinkles (the left forehead was left behind compared to the right), and left sock and glove hypesthesia.

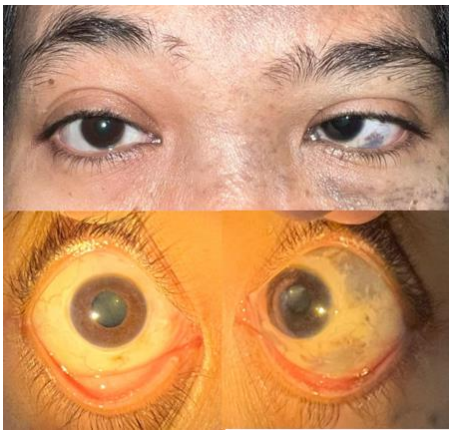

Figure 1. Clinical picture of the patient.

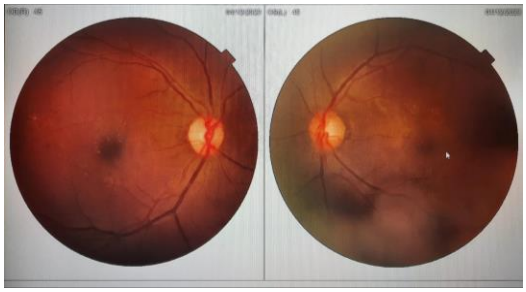

Figure 2. Fundoscopy examination.

Brain MRA was normal. The RPE appears hyperreflective and thickened, which concluded to RPE hyperpigmentation on macula OCT. The overall laboratory examination was normal, except the HbA1c levels were elevated (6.9%). She underwent left eye orbital ultrasonography and found an axial length of 20.88 mm, clear vitreous, intact retina, and normal choroid thickness. It concluded that the ultrasonography was normal.

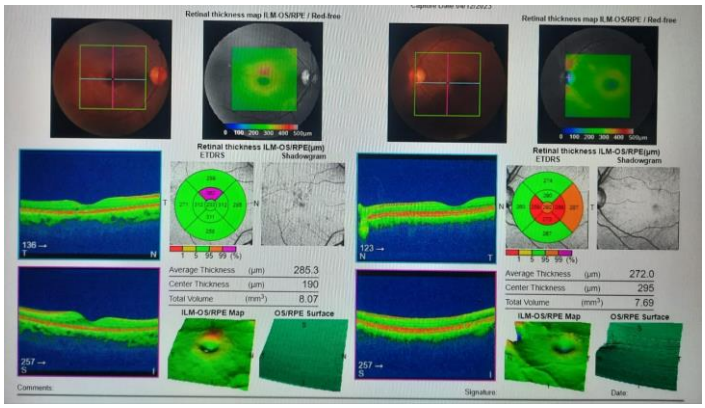

Figure 3. Macular OCT.

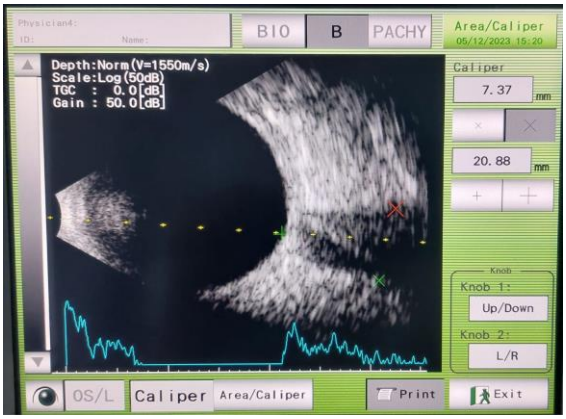

Figure 4. Orbital ultrasonography of the left eye.

She was diagnosed with oculodermal melanocytosis in the left eye, moderate NPDR in both eyes, juvenile cataracts in both eyes and esotropia in the left eye due to left-eye cranial nerve VI paralysis. The differential diagnosis was ptosis neurogenic left eye and hypertrophy of RPE. Education, glucose control by an internist, follow-up at 6 months, and consultation with neuro-ophthalmology subdivision and strabismus subdivision were done. The overall prognosis of this patient was good.

3. Discussion

A 39-year-old woman came with complaints of her left eye rolling inwards for the past 5 years, accompanied by a flat black spot on her forehead that spread to her left cheek. This spot has also appeared on the white part of her left eye since birth, but there have been no black spots on the roof of the mouth; scleral involvement occurs in two-thirds of cases. The spot feels numb and painless. As many as 50% of hyperpigmentation due to an increase in the number of melanocytes occurs at birth with macular pigmentation, which is usually localized in the forehead and periocalar region of the distribution region of the ophthalmic and mandibular branches of the trigeminal nerve.[2,5] Oculodermal melanocytosis (ODM) affects 0.014% - 0.034% of the Asian population and is more common in Women up to 85%, generally occurs unilaterally as in these patients.[2]

Several pathological theories have been put forward, such as that melanocytes have not completely migrated from the neural crest to the epidermis at the embryonic stage. Migration of hair bulb melanocytes and reactivation of pre-existing latent dermal melanocytes, triggered by dermal inflammation, ultraviolet radiation cannot reach the deep dermal melanocytes or hormonal changes during pregnancy.[4]

According to the Mishima classification, she was classified to type II, namely, moderate gray intensity with involvement of the eyelids, zygomatic area, and bridge of the nose. Skin biopsy is necessary only if clinical changes are suspected in the form of malignant transformation of the skin, eyes, and mucosal tissue.[2]

The patient complains that the left eye is rolling inward, resulting in esotropia 45^{o} of the eyeball, indicating the possibility of a muscle disorder or nerve disorder, and this will be proven by further examination. Due to esotropia, she also complained of double vision due to unattainable alignment, so that vision becomes non-binocular.

In the ophthalmological examination carried out, there was a restriction in the movement of the left eyeball in the superotemporal, temporal, and inferotemporal directions. No restrictions were found in the extraocular muscles during the forced duction test (FDT). In this patient, the possibility of left eye cranial nerve VI paralysis occurred, with sharp vision in the right eye 6/ 30 ph 6/7.5 and the left eye 6/60 ph (-) in the nasal direction. There was no increase in intraocular pressure in the patient; in some cases, glaucoma complications often occur because melanocyte invasion can block fluid drainage, thereby causing an increase in intraocular pressure.[5,6] The sclera of the left eye appears hyperpigmented in 3 quadrants, supporting the diagnosis of ODM in the patient with both cloudy lenses.

We can think of juvenile cataracts in this patient. With the patient's age being 39 years, the patient's cataract management can be considered cataract extraction using the phacoemulsification technique and IOL implantation. The posterior segment of the left eye shows a darker color (hyperpigmentation) in the 3 quadrants of the retina compared to the color around the retina. The cause is excessive choroidal pigmentation, which sometimes causes mottling and dot pigmentation.[4]

We can classify it into the Nevus of Ota classification by Vishnevskia-Dai, where the patient is included in the B 3+ classification, which shows hyperpigmentation in two quadrants. 3+ indicates surface involvement with the choroid.[5] Supported by OCT examination, the left eye showed thickening of the RPE layer when compared to the right eye. In choroidal nevus, OCT shows a hyporeflective mass that does not significantly alter the choroidal vascularization and EPR-Bruch membrane complex. Fundus autofluorescence examination may be recommended to help in the differential diagnosis between choroidal nevus and melanoma. The latest imaging techniques are very helpful in the diagnosis and management of ODM complications.[4]

Generally, nevus of ota is benign and does not require further treatment; only therapy must be carried out if complications occur, such as if there are complications of glaucoma in the patient, administration of antiglaucoma drugs can be considered and periodic evaluation of intraocular pressure in the patient. Apart from that, evaluation must still be carried out looking at the development of the nevus and any development towards malignancy, although it has been proven that twice as many uveal melanomas develop as malignant when compared with melanocytosis.[4] Laser treatment has been widely used for many years on skin lesions for cosmetic purposes. However, some side effects can occur as a result of laser treatment, such as purpura, crusting, post-inflammatory hyperpigmentation, and scarring. Meanwhile, there have been no reports of laser use on the surface of the eyeball or intra-ocular areas.[2]

4. Conclusion

A case report of a 39-year-old woman diagnosed with oculodermal melanocytosis (ODM). Although this is a benign condition, patients are at risk of developing complications such as increased intraocular pressure and melanoma of the skin and/or uvea. Ongoing evaluation with imaging methods is highly recommended, even if only skin lesions may be repaired using a laser for cosmetic reasons.

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