

CASE REPORT

GENERALIZED NON INFLAMMATORY SCLERAL THINNING IN A CASE NAEVUS OF OTA

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ABSTRACT: A 30 year old female patient came with redness, pain and watering in her left eye since five days. Past history included sudden loss of vision in right eye two years back following birth of a stillborn baby due to preeclampsia. On examination she had hyperpigmentation of the face and neck. Both the eyes showed patchy hyperpigmentation all around the limbus. Right eye showed 10 degrees of exotropia with afferent pupillary defect. Intraocular pressure was raised to 29 mm of Hg. The Right disc C/D ratio was 0.7 with pallor. The left eye showed signs & symptoms suggestive of acute iridocyclitis with normal IOP. Left visual acuity had dropped to 20/60. Further investigation confirmed the patient had generalised non inflammatory scleral thinning on UBM and associated conductive hearing loss. This is therefore an unusual case of Nevus of ota/Ito.

KEYWORDS: Nevus of Ota, Scleral thinning, POAG, Iridocyclitis, Hyperpigmentation.

INTRODUCTION: Ocular or oculodermal melanocytosis is a congenital or acquired pigmentary condition that can affect the periocular skin, sclera, uvea, orbit, palate, ear drum, and meninges. It can occur as a diffuse condition, or as a sectoral condition.^[1]

CASE HISTORY: A 30yrs old female comes with first episode of redness, pain and watering in her left eye (OS) since five days. She claims sudden loss of vision in right eye (OD) two years back following birth of a stillborn pre-eclampsia baby. At present patient is a high hypertensive controlled on medication.

On examination there is hyperpigmentation on both sides of her face, neck and forearms more on the left side (Figure 1). Conjunctiva and sclera of both eyes show patchy hyperpigmentation all around the limbus, extending upto the equator (Figure 2).

Right eye best corrected visual acuity (BCVA) is finger counting at two meters. Manifest exotropia of ten degrees (Figure 3). Anterior segment is normal. Peaked pupil in the infero-temporal quadrant, with afferent pupillary defect seen (Figure 4). +90D shows cup disc ratio of 0.7 with pallor. There is branch arteriolar occlusion in infero-temporal quadrant (Figure 6). The rest of retina appears normal (Figure 7).

Left eye has BCVA of 20/60 improving to 20/20 with -0.50 DSph. There is circumcorneal congestion with blepharospasm. Anterior segment is wnl. Mild flare with fresh keratic precipitates on endothelium seen. Pupil is 2mm immobile. Release of posterior synechae on atropinisation shows circumferential pigmented ring on anterior capsule (Figure 5). +90 D examination shows a normal retina (Figure 6) (Figure 7). The Intraocular pressure recorded were 29 mm Hg for OD and 17mm Hg for OS. (Table 1).

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Routine investigations like CBC/ ESR, blood sugar, VDRL, TORCH titre, HIV, RA factor, HLA B27 were all wnl. Xray chest and spine were found to be normal. Indirect gonioscopy showed an open angle with hyperpigmentation extending anterior to the schwalbe's line confirmed on UBM. (Figure 8) UBM depicted generalized thinning of sclera to 170 microns (Figure 9) as against the normal 350 microns. No other signs of inflammation seen. Automated 30-2 perimetry LE shows few isolated scotomas in arcuate area (Figure10). ENT reference showed no pigmentation on palate, oral mucosa or tympanic membrane. Audiometry showed mild conductive deafness. (Figure 11) MRI brain scan was normal ruling out leptomeningeal pigmentation, space occupying lesion or infarct. (Figure 12) OD was treated with eye drop Brimonidine 0.1% bringing down IOP to 23 mmHg OS was treated like any acute iridocyclitis with steroids and atropine Post treatment finding are as seen in Table I. Her anti-hypertensive treatment was continued. She was advised regular follow up every six months to rule out any onset of melanoma.

DISCUSSION: Nevus of Ota named after Professor Ota who characterized these cases of aberrant Mongolian spots located on face, also the eye, and named the disorder Naevus fusco-caeruleus ophthalmomaxillaris. In 1956, Fitzpatrick renamed the syndrome oculodermal melanocytosis (ODM).^[2] Naevus of Ota is on the forehead and face around the eye whereas Naevus of Ito is on the shoulder and upper arm area.^[3]

Ratio of female to male patients is approximately 4:1. It is more common in Asians and African descent, but very rare in Caucasians.^[4] No recognizable inheritance pattern with 60% of cases present at birth with the remainder presenting at puberty.^[5] The histological hallmark of these lesions is the presence of pigmented dendritic melanocytes.

Glaucoma is found in approximately 10% of patients.^[6] The aetiology being direct infiltration of the trabecular meshwork by the accumulated melanocytes or by proliferation of melanocytes within the anterior chamber of the eye. This may present as a gradual increase in IOP, acute glaucoma with uveitis, angle closure glaucoma or as congenital glaucoma. For unknown reasons 35% of adult patients have ipsilateral uveitis and transient rise in IOP.^[7]

ODM, diffuse or sector-specific as against dermal component have high risk of development of melanoma. ODM affects 0.04% of the white population and 1.4% of the uveal melanoma population.^[8] Orbital or meningeal melanoma, in ODM is seen by 35 years as against non ODM seen at 55 years.^[9] Nevus of Ota is also associated with an increased risk for melanoma of the skin, eye, and brain, particularly when it occurs in Caucasians and it may lead to tinnitus or deafness in a few individuals and carries an increased risk for malignancy.^[10] The risk for metastasis is 2.8, 2.6 and 2 times for patients with iris melanocytosis, choroidal melanocytosis, and scleral melanocytosis respectively. The only factor predictive of metastasis and death in patients with ocular melanocytosis is increasing tumor thickness

There is also a report of a case of nevus of Ota in a white woman with ipsilateral sensorineural hypoacusia emphasizing the wide extension of the pigmentation.^[10]

Patients with ODM should be routinely followed on a six-monthly basis with widely dilated fundus examination, as they carry an increased risk for uveal melanoma and subsequent doubled risk for metastasis. All patients with ODM need lifelong ophthalmic follow-up. Early detection of uveal melanoma is critical for minimizing metastatic risk and improving life prognosis.

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Our patient fitted in typical Nevus of Ota, with characteristic pigmentation on both sides of face, eyes, shoulder, forearm and back. There is associated glaucoma and anterior uveitis too. The generalized uniform scleral thinning seen on UBM has not been reported in literature as yet. If scleral thinning was patchy it could be attributed to any inflammatory cause. She had no h/o scleritis, redness, pain or watering in the past. Glaucoma of 27 mm or her 1st episode of iridocyclitis would not be contributory. We thus believe it to be idiopathic. Her loss of vision in RE is explained on the C/D ratio of 0.7 with acute perfusion loss with an inferotemporal branch arterial occlusion probably during her preeclampsia attack. We decided to report this case as generalized non inflammatory scleral thinning is not mentioned anywhere in association with Nevus of Ota.

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Findings	OD	OS	OS after Rx
BCVA on Logmar Chart	FC at 2 meters PL PR +ve	0.4	0.1
Ocular Adnexia	Hyperpigmentation on forehead and periorbital region		
Conjunctiva/Sclera	Hyperpigmentation of the conjunctiva & Sclera 360 degrees around the limbus upto the equator		
Conjunctiva/Sclera	Normal	ciliary congestion	Normal
Cornea	Size shape normal and transparent		
Anterior Chamber	Normal	Aqueous flare +1	Normal
Iris	Dark brown pigmentation with normal crypts and furrows		
Pupil	Inferotemporal peaking with afferent defect	2 mm with posterior synechiae	Dilated with anterior capsular pigment ring
Lens	Clear		
Vitreous	Clear		
Posterior Fundus	C/D ratio 0.7 with pallor. Infero temporal branch arterial occlusion.	C/D ratio 0.3. Rest wnl.	
Peripheral fundus	Normal. No pigmentation seen		
IOP on Applanation	29 mm to 23 mm of Hg on Rx	17 mm of Hg	

Table I: Ocular Examination at time of presentation & following Rx

Figure 1: Hyperpigmentation of the Face, Neck, Back and forearms with fullness of the face seen on the left side.



Figure 1

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Figure 2: Bluish grey pigmentation of the conjunctiva, episclera, and sclera in both eyes with 10 degrees Exotropia of the RE.



Figure 2

Figure 3: Bluish grey pigmentation of the conjunctiva, episclera, and sclera is almost 360 degrees surrounding the limbus.



Figure 3

Figure 4: RE pupil shows correctopia, while LE pupil has been dilated with atropine. Pigment ring seen on anterior capsule of the lens due to release of posterior synechiae.



Figure 4

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Figure 5: LE shows ciliary congestion with pigment on lens depicting the anterior uveitis.



Figure 5

Figure 6: Fundus Photo showing a CD ratio of 0.7 in the RE with infero-temporal branch arterial occlusion and Normal CD ratio of 0.3 in the LE. No pigmentation seen.

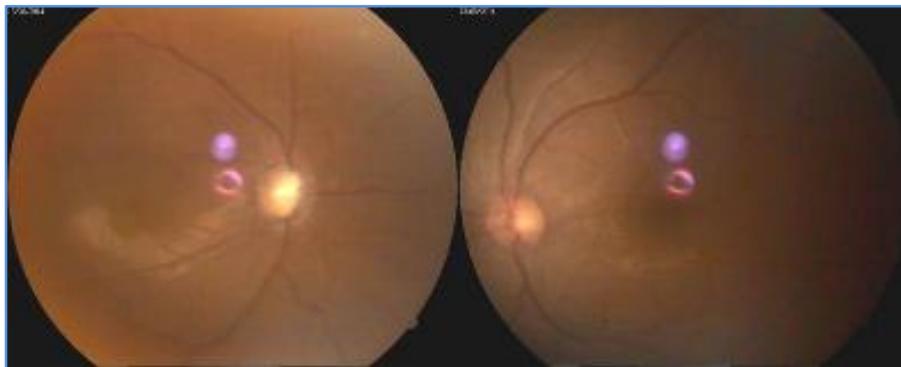


Figure 6

Figure 7: Fundus Photo showing absence of pigmentation in the periphery.

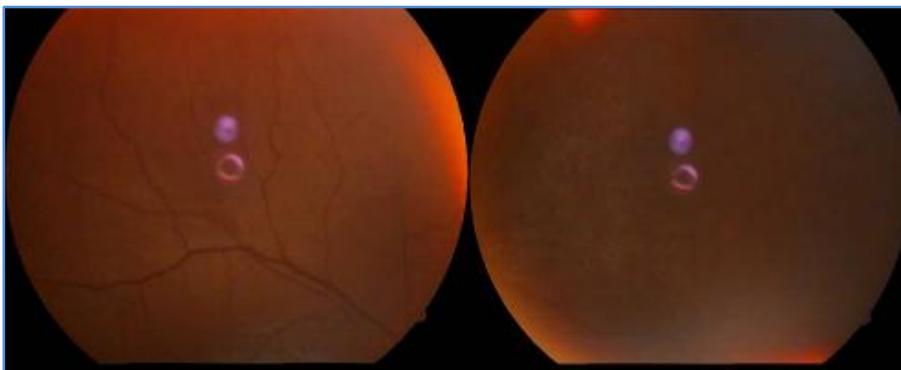


Figure 7

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Figure 8: UBM pictures showing an open angle as also seen on gonioscopy. There is intense pigmentation on the external wall of the angles of both eyes.

Figure 9: UBM shows generalized thinning of the sclera to 170 microns.

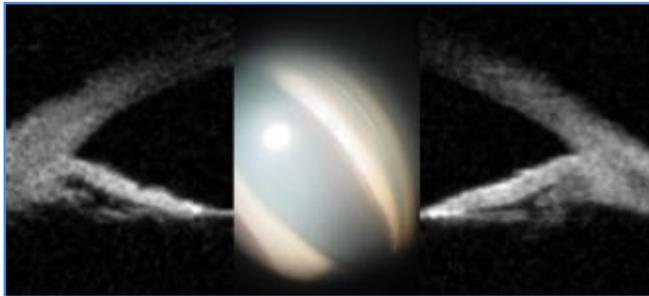


Figure 8

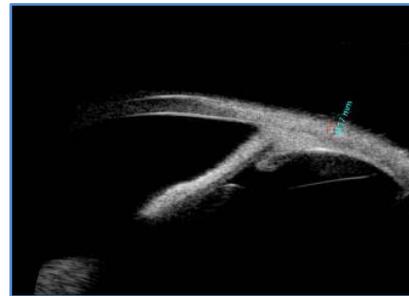


Figure 9

Figure 10: Automated Perimetry shows extremely poor vision on the RE.

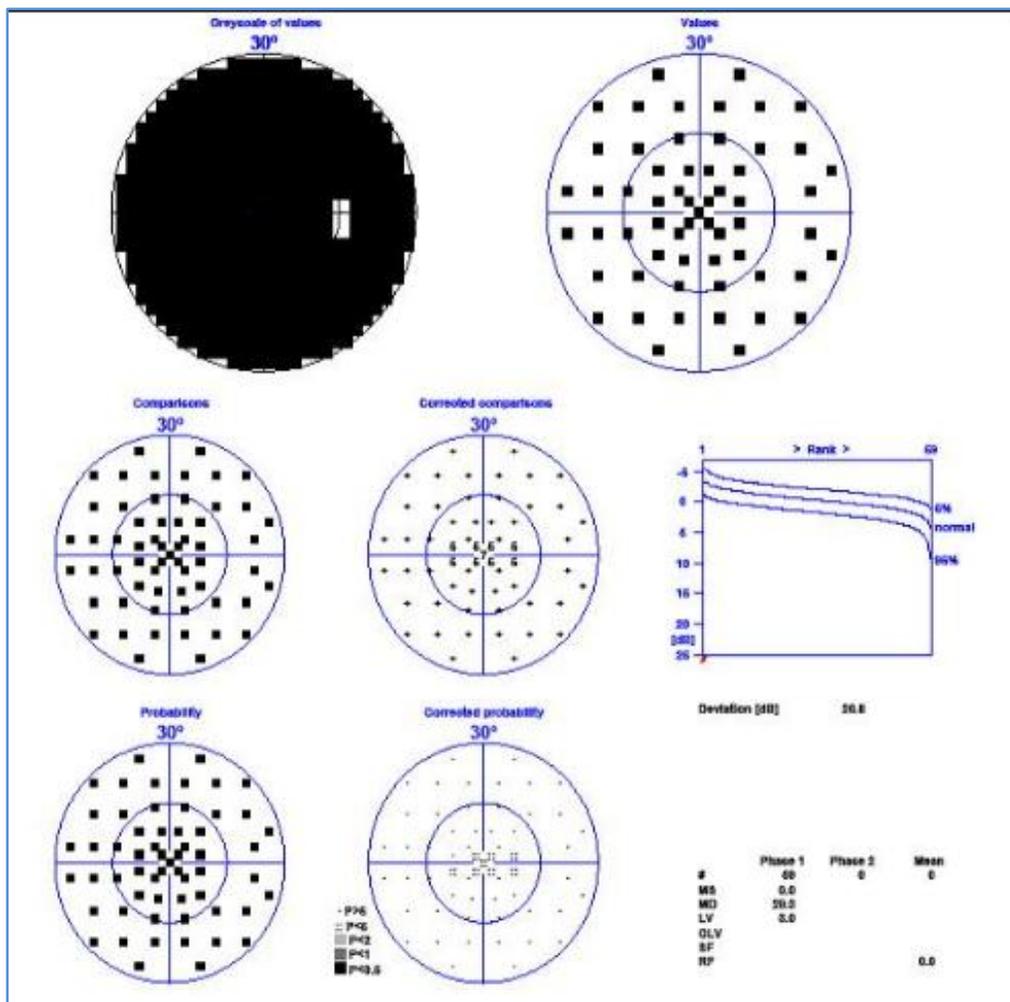


Figure 10

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Figure 11: Automated Perimetry with few isolated scotomas in the arcuate nerve fibre layer in the LE.

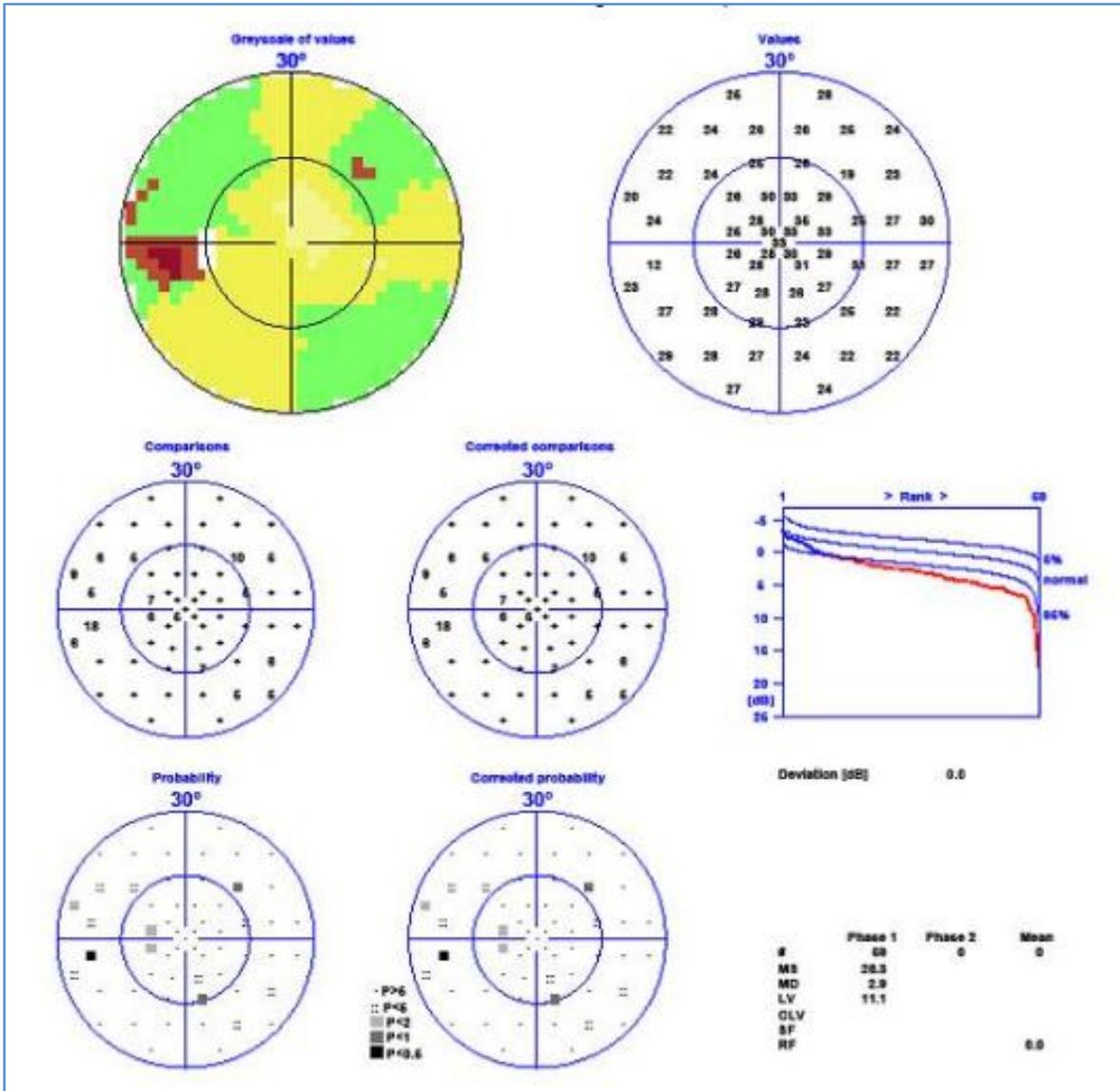


Figure 11

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Figure 12: Audiometry showing mild conductive deafness both sides.

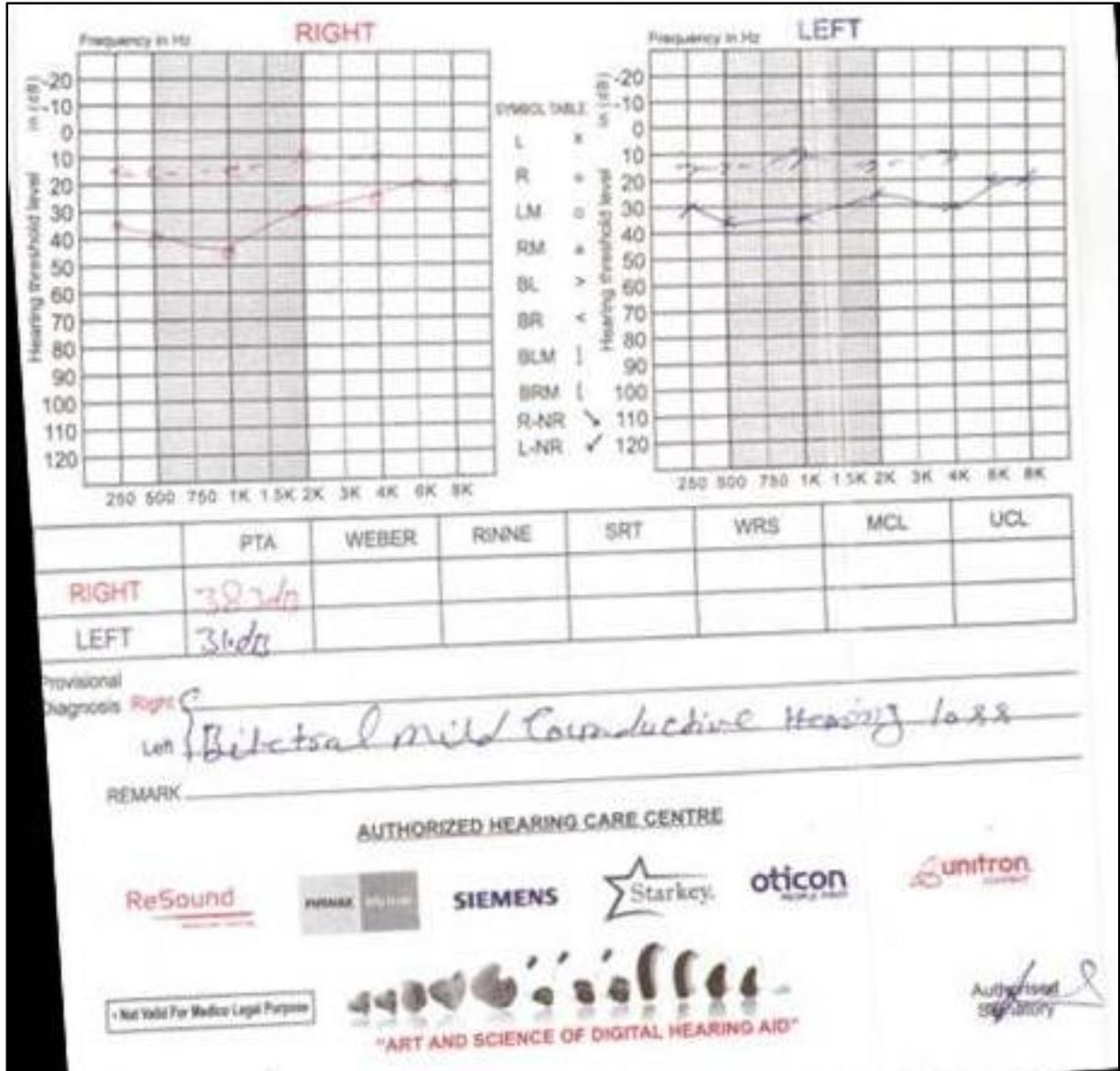


Figure 12

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Figure 13: MRI Brain Scan is completely normal ruling out leptomenigeal pigmentation, space occupying lesion or infarcts.

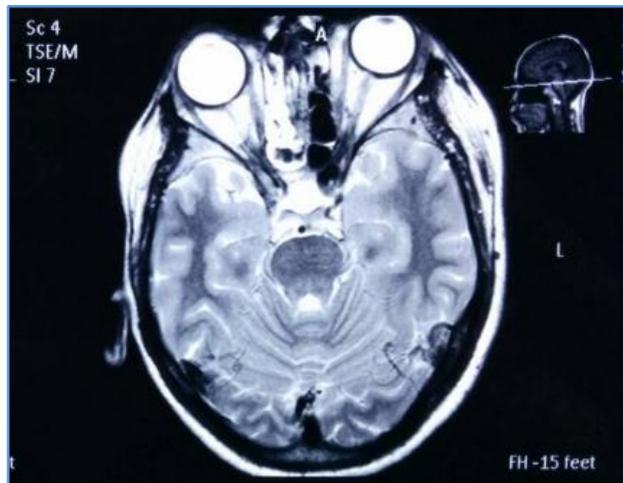


Figure 13

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