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Medullated retinal nerve fibres a rare association with young ischemic stroke: A Case Report

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Abstract: Medullated (Myelinated) retinal nerve fibres(MRNF) are developmental anomaly and occur in less than 1% of the population, however, most of the reported cases of MRNF is with ocular diseases ,systemic association of MRNF is rarely described. It is usually associated with ipsilateral high myopia and amblyopia. We are reporting a case of a 28 year old young female presenting to us with sudden onset of right crural monoplegia associated with MRNF on fundoscopy. Systemic association was represented by a subacute infarct on MRI Brain which is an unusual association with MRNFs.
Keywords: Myelinated nerve fibres; ischemic stroke; crural monoplegia, subacute infarct

INTRODUCTION

The first description of myelinated retinal nerve fibres (MRNF) was given in 1856 by the German pathologist Rudolf Virchow based on eye preparations. He described that "retina was white, very thick and wrinkled. Macula was normal and near the optic disc, though more deeply situated, were thick, opaque, chalk-white spots, which spread around the disc in the shape of a star[1]. Today this developmental anomaly occurs in less than 1% of the population[2]. MRNF may be congenital or acquired in nature. It is usually asymptomatic. It presents as white or greyish spots with jagged edges, in conjunction with the optic disc, around it, or not related. It is rare case of family inheritance[3]. Strokes in young adults are relatively uncommon; the disorder usually occurs in the middle-aged and elderly. The National Survey of Stroke revealed that only 3.7% of all strokes occurred in patients aged 15-45 years[4].The etiologic and prognostic features that characterize strokes in older persons may not apply to young adults. Previous reports suggest that a cause can be found in 55-93% of young people with strokes[5].

CASE REPORT

A 28 year old married female presented to our tertiary care hospital with complaints of sudden onset of weakness of right lower limb. On proper history taking she revealed that she was apparently alright when she went to bed at night but on getting up in the morning she developed weakness and was unable to move her right lower limb which was sudden in onset and complete in nature with no precipitating cause. There was no any exertional activity preceding or accompanying the onset of weakness. There was no history of fever, headache, diarrhoea, vomiting, abnormal body movements. No history of trauma, seizure, loss of consciousness was present .No history of root pain, girdle like sensation or bladder and bowel involvement was present. No history of tingling, numbness or paraesthesia was present. No history of diplopia, loss of vision or any ocular pain. No history of intake of oral contraceptive pill or any recent vaccination. No h/o dysphagia, dysarthria or any facial weakness. There was no similar episode in the past. There was h/o blood transfusion in the past. There was no significant family history. She had history amennorhoea for last one and a half years i.e. since her last child birth. On general examination patient was average built, and severely anaemic .No icterus, clubbing, cyanosis, oedema, lymphadenopathy was present. Vitals were stable with BP -130/70 mm of Hg in right upper limb in supine position, PR -86/min regular, all peripheral pulses were palpable. Respiratory rate of 16/min, thoracoabdominal type .No bruit was audible over carotids.

On systemic examination: CNS: Patient was well oriented to time, place and person. Sensory system examination was normal. All cranial nerve examination was normal. Motor system examination revealed upper motor neuron type of paralysis of the right lower limb (pure motor monoplegia). Babinski sign was present only on right side; ankle clonus with brisk knee jerk was present. Hypertonia with clasp knife type of spasticity was present .Power was of grade 0 in right

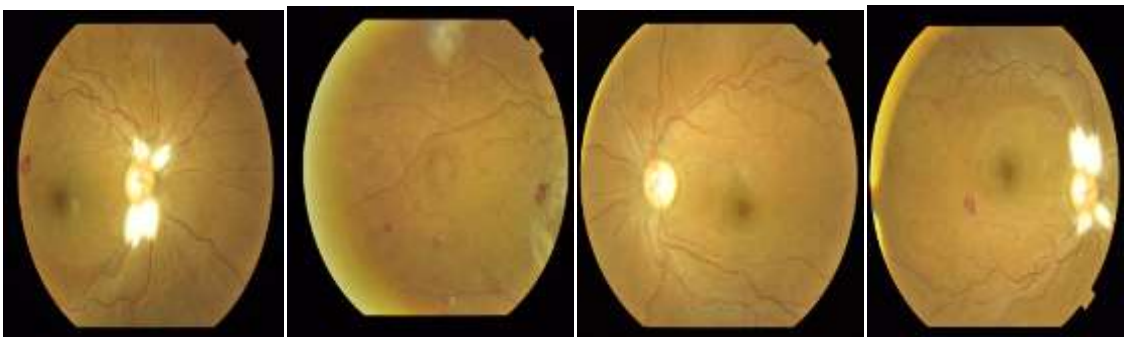
lower limb. Rest of the motor system examination was normal.

On CVS examination: S1, S2- heard, a haemic murmur was present in pulmonary area. Per abdomen examination did not reveal any organomegaly. R/S examination was normal with bilateral vesicular breath sounds.

On investigation: Hb- 1.9 gm/dl, TLC-6530/mm^3 (N-88% ,L- 10%, M-01% ,E 01%), Platelet count 71000/mm^3, MCV -153.5 [Fl] peripheral picture was s/o Macrocytic anaemia. Blood sugar- 125 mg%, urea-29 mg% , S. bilirubin – 2.7 mg%, S. creatinine – 0.81 mg% , SGOT- 153 IU/L, SGPT- 63 IU/L, ALP-158 U/L, S. Protein- 6.4 gm/dl.
ECG of the patient was within normal limit except sinus tachycardia. Chest X-ray was s/o mild cardiomegaly but 2D-echocardiography was within normal limit except mild thickening of papillary muscle.

Ophthalmoscopic examination:

In both eye distant vision was 6/9 with no improvement with pinhole and near vision was N6. Right eye fundus examination revealed myelinated retinal nerve fibers, superior and inferior to disc with superficial white centred haemorrhagic spots present supero temporal to fovea. Left eye fundus shows multiple dot and blot haemorrhages. Cup: disc ratio was 0.6. In both eye foveolar reflex was dull and bilateral retinal haemorrhages were present. Bilateral generalised pallor of fundus was present. Bilateral retinal vascular tortuosity was present over disc and general fundus.



(A) (B) (C) (D)

Figure: 1 Fundus photograph of patient showing myelinated retinal fibres in the superior and inferior quadrant of retina and white centred haemorrhage in the right eye [A,B]. Left eye showing multiple dot and blot haemorrhages [C,D].

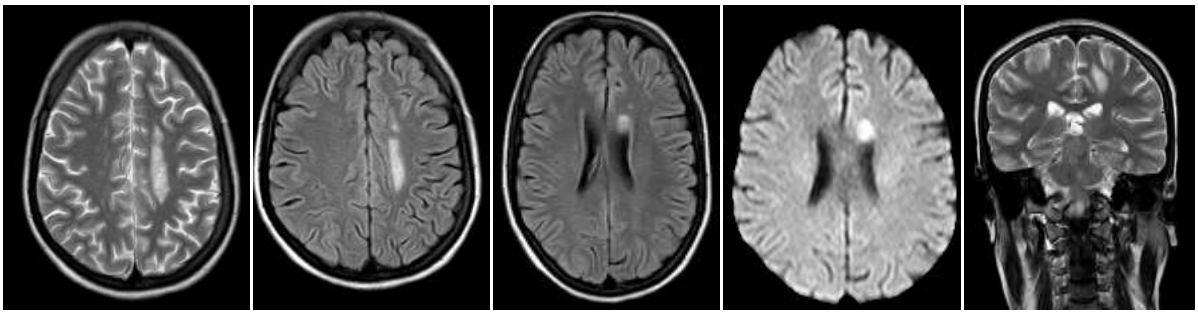


Figure: 2 MRI scan of brain showing multifocal round to oval areas of altered signal in bilateral Centrum semiovale, left periventricular deep white matter and left anterior pericallosal area.

MRI study of brain revealed multifocal round to oval shaped areas of altered signal in bilateral centum semiovale, left periventricular deep white matter, left anterior pericallosal white matter without any abnormal enhancement, suggest possibility of subacute infarcts.MRI spine of the patient was within normal study.

DISCUSSION

MRNFs are more frequently found in females as compared to males and are more commonly seen at the superior sector of the nerve head and in the infero temporal retina than in other areas. Myelinated RNFLs are bilateral in 7.7% of cases[6]. MRNFs are formed because of an error during the process of myelintion of fibres of retinal ganglion cells. This process of myelination is carried out by oligodendrocytes. Physiologically, it starts around the 5th month of fetal

life from the corpus geniculatum laterale, following the visual pathway and ending on the lamina cribrosa at childbirth or shortly thereafter[7-8]. Lamina cribrosa is a protective barrier against the penetration of fibre myelin in the area of the retina. The development of myelinated nerve fibers associated with the optic disc depends on the degree of hypoplasia of the disc itself and that is why this type of fiber is often accompanied by ambylopia, axial myopia, anisometropia and strabismus[8-9]. In areas occupied by the myelinated nerve fibres there may appear vascular anomalies, such as neo vasularization, bleeding and thrombus and also retinal abnormalties. It happens that these fibers are found in patients with diseases such as neurofibromatosis type I or Gorlin syndrome,Down's syndrome and developmental disorders , or various forms of craniofacial dysostosis[3]. Myelinated nerve fibres are occasionally progressive[10]. MRNFs had been associated with other disorders such as Turner syndrome, epilepsy [11], Dolichocephly[11] , Von Recklinhausen;s disease, Arnold –Chiari malformation and hydrocephalus. There has been a recently published literature on MRNF and its association with ACS in a young male[12].

CONCLUSION

MRNF has been recently associated with coronary artery disease in a young adult in a recently published literature. In our case the association is seen in a young female with ischemic stroke. Thus the presence of systemic association in some patients with MRNF's cannot be ruled out and this correlation needs to be further explored .A possible explanation for this could be that the developmental anomaly in blood vessel is not only restricted to ocular blood vessels but also to systemic ones predisposing the patient for high risk of thrombosis which can further lead to coronary artery disease or ischaemic stroke. Thus whenever an ophthalmologist or physician comes across MRNFs in a young patient , it should not be seen only as incidental finding per se but possibility of its systemic associations should also be explored.

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