Brief Report

MRI findings of contralateral oculomotor nerve palsy in Parry-Romberg syndrome☆

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ABSTRACT

Purpose: To study a case of right Parry Romberg syndrome associated with contralateral oculomotor nerve palsy using high-resolution cerebral and orbital magnetic resonance imaging (MRI).

Observations: There were no brain MRI abnormalities. However, there was marked enophthalmos on the right with reduction of orbital fat. Extraocular muscles contracted normally in the right eye, but in the left eye, there was reduced contractility of the medial, inferior, and superior rectus muscles. The intraorbital motor nerves were unremarkable bilaterally, and the right optic nerve was tortuous.

Conclusions and importance: Parry Romberg syndrome is a disease of unknown etiology with various ophthalmologic manifestations. This case study contributes extensive MRI data to the limited literature on ophthalmologic anatomic findings in a patient who had Parry Romberg syndrome with contralateral paralytic strabismus.

1. Introduction

Parry-Romberg Syndrome, also known as progressive facial hemiatrophy, is characterized by idiopathic, gradual unilateral atrophy of facial muscles, bones, and skin. It is a disease of unknown etiology with various ophthalmologic manifestations. This case study contributes extensive MRI data to the limited literature on ophthalmologic anatomic findings in a patient who had Parry Romberg syndrome with contralateral paralytic strabismus.

2. Case report

This 37-year old woman was diagnosed with right-sided Parry Romberg syndrome as a teenager. She had previously undergone multiple filling procedures in the right cheek and temporal orbit, and right eyelid surgery. She presented with a one-year history of worsening horizontal, binocular diplopia, and progressive left exotropia. She was otherwise healthy with no family history of Parry Romberg syndrome. Conventional brain MRI with contrast was unremarkable. High-resolution cerebral and orbital magnetic resonance imaging (MRI) revealed marked enophthalmos on the right with reduction of orbital fat. Extraocular muscles contracted normally in the right eye, but in the left eye, there was reduced contractility of the medial, inferior, and superior rectus muscles. The intraorbital motor nerves were unremarkable bilaterally, and the right optic nerve was tortuous. Conclusive MRI findings were normal.

Conventional brain MRI with contrast was unremarkable. High-resolution cerebral and orbital magnetic resonance imaging (MRI) revealed marked enophthalmos on the right with reduction of orbital fat. Extraocular muscles contracted normally in the right eye, but in the left eye, there was reduced contractility of the medial, inferior, and superior rectus muscles. The intraorbital motor nerves were unremarkable bilaterally, and the right optic nerve was tortuous.
resolution, T2 weighted, surface coil MRI of the orbits was performed with target fixation in multiple gaze positions to ascertain extraocular muscle contractility. There was marked enophthalmos with reduction of right orbital fat, yet normal sized extraocular muscles that contracted normally (Fig. 2). Orbital fat volume and extraocular muscle sizes were normal on the left, but there was reduced contractility of medial rectus, inferior rectus, and superior rectus muscles. IR – inferior rectus muscle. LR – lateral rectus muscle. MR – medial rectus muscle. SO – superior oblique muscle. SR – superior rectus muscle.

This case report was approved by the institutional review board at the University of California, Los Angeles. Written informed consent was obtained for study procedures and the patient consented to publication of photos in this case report.

3. Discussion

Parry Romberg Syndrome is a sporadic condition characterized by unilateral facial degeneration of fat, muscles, and bone. Although recognized for more than 2000 years, its etiology remains mysterious. To our knowledge, this is the first case report demonstrating extensive, high-resolution MRI findings in a patient with Parry Romberg syndrome having contralateral extraocular muscle manifestations.

Other than characteristic features of Parry Romberg syndrome including unilateral facial deformation, scar-like cutaneous changes, subcutaneous fat atrophy clearly respecting the hemifacial midline, and enophthalmos, our patient also had contralateral oculomotor palsy and bilaterally fixed, dilated pupils. The contralateral extraocular muscle manifestations in Parry Romberg syndrome were previously described.
by Miller et al., who reported in 1995 a 21-year old female with partial paralysis of the medial rectus and left inferior oblique muscles, and mild ptosis consistent with partial oculomotor nerve palsy, similar to our patient. However, Miller’s case also had seizures, ipsilateral central retinal vein occlusion, uveitis with secondary cataracts and glaucoma. Other case reports also described additional contralateral ocular manifestations of granulomatous uveitis and peripheral retinal vasculitis with optic neuritis. However, our patient did not have any of the foregoing disorders. Extensive MRI was not performed on the previous reported cases.

The neurodegenerative effects of Parry Romberg syndrome are still unclear. Moko et al. described brain MRI of 10 patients with Parry Romberg syndrome, half of whom being normal with the remainder exhibiting ipsilateral abnormalities including focal occipital and parietal region atrophy, and ipsilateral parietal and bilateral frontal white matter hyperintensities. Interestingly, conventional brain MRI in the current patient was normal, although this cannot exclude a small subarachnoid lesion of the oculomotor nerve that might be detectable using higher resolution, heavily T2-weighted technique. However, high resolution, multi-positional orbital imaging showed reduction in contractility of the left medical rectus, left superior rectus, and left inferior rectus muscles without evidence of infiltrative enlargement or motor nerve abnormality. Although Moko et al. found intracranial changes that may explain the oculomotor palsy, no such brain MRI abnormalities were evident in our patient. This suggests another abnormalities below the detection threshold of routine brainstem MRI. The orbital imaging technique employed here typically demonstrates atrophy of the orbital branches of the oculomotor nerve in cases of compressive oculomotor palsy; absence of this finding argues against the presence of a tumor or aneurysm.

The findings in the left side are atypical of Parry Romberg syndrome and probably do not represent an early form. However, it seems improbable that the co-existing abnormalities of oculomotor nerve function on the left could be simply coincidental, given the extreme rarity of Parry Romberg syndrome, and the relative rarity of idiopathic, partial oculomotor paresis.

4. Conclusions

This present case of Parry Romberg syndrome provides detailed MRI of the orbits, detailing enophthalmos, asymmetry of bony orbits, and fat changes, and excludes evidence of gross central nervous system disease. However, although our patient has normal motor nerves bilaterally, she presented with progressive reduced contractility in the contralateral oculomotor nerve-innervated extraocular muscles. Since our patient’s progressive strabismus is also common in Parry Romberg syndrome, serial imaging in this and other cases of Parry Romberg syndrome may be warranted to understand progression.

**Patient consent**

Consent to publish the report was obtained from the patient in writing. This study was approved by the Institutional Review Board at the University of California, Los Angeles.

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**Conflicts of interest**

The following authors have no financial disclosures: EKT, ML, JLD.

**Authorship**

All authors attest that they meet the current ICMJE criteria for Authorship.

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