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Case Report

Unilateral Papillitis as the Initial Presentation of Hemifacial Atrophy: Case Report and Review of Literature

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Abstract

Papillitis is a vision threatening condition, characterized by inflammation of the optic disc which often mimicked the features of papilledema; hence these patients often referred to the Neurosurgeon to exclude any intracranial pathology. This entity is associated with a number of intracranial, as well as extracranial pathologies. Among them, Parry–Romberg syndrome, also known as progressive hemifacial atrophy (PHA) possesses multiple ophthalmologic and neurologic manifestations. Here we report the case of a 14-year-old girl, who presented with the feature of progressive dimness of vision involving the right eye. Thorough physical examination demonstrated features of PHA overlapping with papillitis on fundoscopic examination. The patient treated with steroids, following which there was visual improvement. After evaluation in our facility, she was referred to department of plastic and reconstructive surgery for aesthetic improvement.

Keywords: *Papillitis, Parry–Romberg syndrome, progressive hemifacial atrophy*

Abbreviation

3D	*3 Dimensional*
CT	*Computed Tomography*
MRI	*Magnetic Resonance Imaging*
PHA	*Progressive Hemifacial Atrophy*
PRS	*Parry-Romberg Syndrome*
T1WI	*T1 Weighted Image*
T2WI	*T2 Weighted Image*

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Introduction:

Papillitis can be the initial symptom of an isolated intraocular pathology, as well as a number of intracranial and extracranial pathologies, like- multiple sclerosis, meningitis, encephalomyelitis, sarcoidosis, systemic lupus erythematosus, and several infectious and nutritional disorders. Parry–Romberg syndrome (PRS), also known as progressive hemifacial atrophy (PHA), is a rare disorder with unknown etiology characterized by progressive atrophy of the skin, soft tissues, and bony structures involving half of the face. Among various neurologic and ophthalmologic manifestations, we focused on the presentation of papillitis because prompt diagnosis and timely intervention can save the vision.

Case Report:

A 14-year-old girl presented with the features of progressive dimness of vision in her right eye since 1 year, for which she first visited to the ophthalmology department. Through systemic evaluation showed negative results for skin rashes, back pain, joint pain, ulcers, Raynaud's phenomenon, epistaxis, hemoptysis, cough, breathlessness. There was no features of raised intracranial pressure or any focal neurological deficit.

General physical examination demonstrated enophthalmos of the right eye, minimal deviation of face to the right side, wasting of muscles involving

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right half of the face (Figure 1). Her visual acuity was restricted to counting finger at 4 feet in the right eye, normal in the left one; dilated fundus examination showed hyperemic disc with marked tortuosity and dilation of blood vessels on and around the right optic disc, obliteration of physiological cup, areas of diffuse retinal whitening and exudation (Figure 2). There was no abnormalities on slit lamp examination.

A comprehensive blood work for systemic pathologies were negative. Ocular fluroscein angiography demonstrated only telangiectatic changes (Figure 3). After that, an MRI of brain with MRA and MRV done and they referred this patient to us to exclude intracranial pathology. MRI of the brain demonstrated features of subcutaneous tissue atrophy at right fronto-parietal region. There was also atrophy of the retrobulbar fat involving right eye. Apart from this there was no identifiable abnormal contrast enhancing areas within the brain parenchyma (Figure 4). Magnetic resonance angiogram seems to be normal (Figure 5 A). However, Magnetic resonance venogram demonstrated filling defect in the right sided transverse sinus, sigmoid sinus as well as internal jugular vein (Figure 5 B).

Fig.-1: *Patient presented with the features of atrophy of the subcutaneous fat with right sided enophthalmos (A & B).*

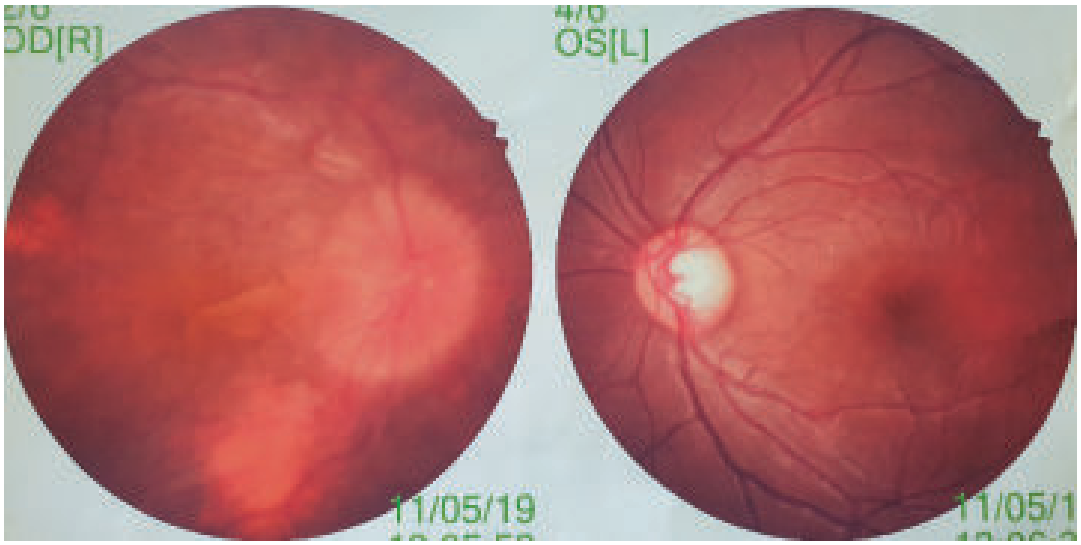

Fig.-2: *Colour fundus photograph demonstrates features of papillitis involving right eye. Left eye seems to be normal.*

Patient was initially started with 60 mg prednisolone daily. Later on, steroid become gradually tapered off. She noticed improvement of vision which restricted to 6/60 on clinical examination. However, after thorough evaluation in our department, we noticed features of lower motor neuron pathology restricted to right half of the face which is not related to any intracranial pathology. Asymmetry of the transverse sinuses is a normal variant. As contralateral fundus is normal, this asymmetry has no significance. So, we advised her to

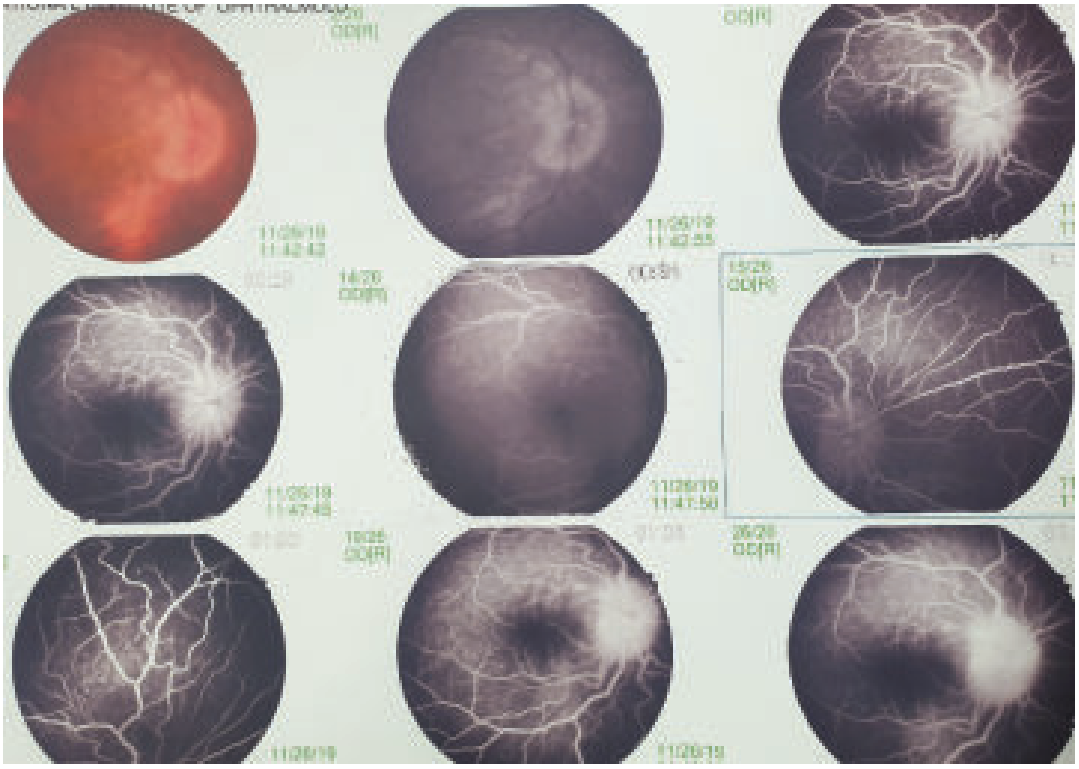

Fig.-3: *Fluorescein angiography of the right eye demonstates the area of telangiectatic changes on background retina.*

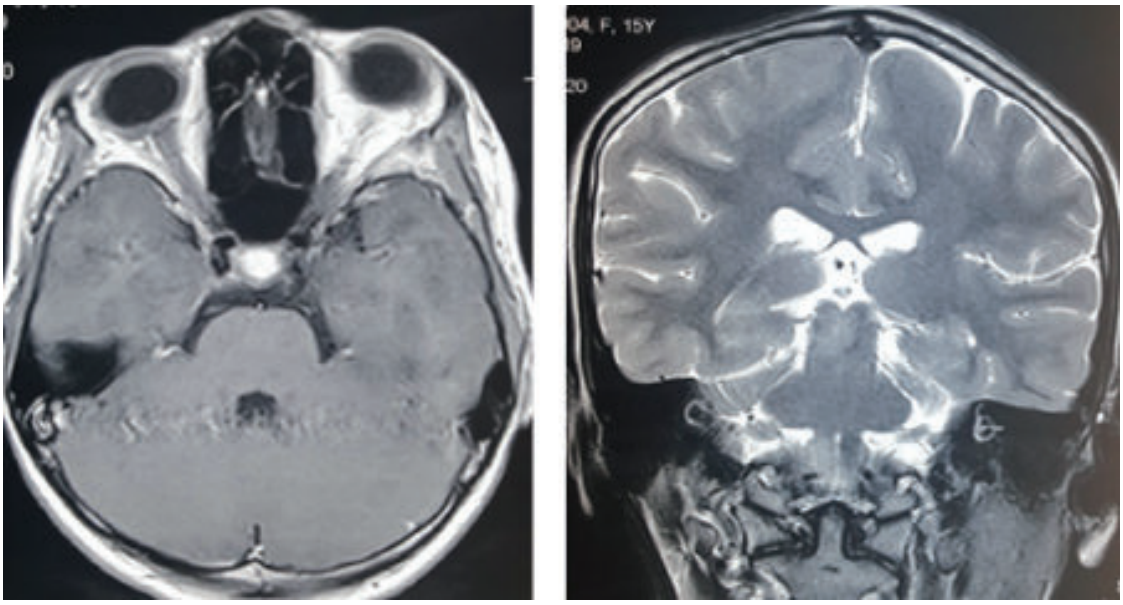

Fig.-4: *(A) MRI of brain, post contrast axial section showing atrophy of the retrobulbar fat (marked by arrow head) , (B) T2WI coronal section showing the atrophy of subcutaneous fat (marked by arrow head) .*

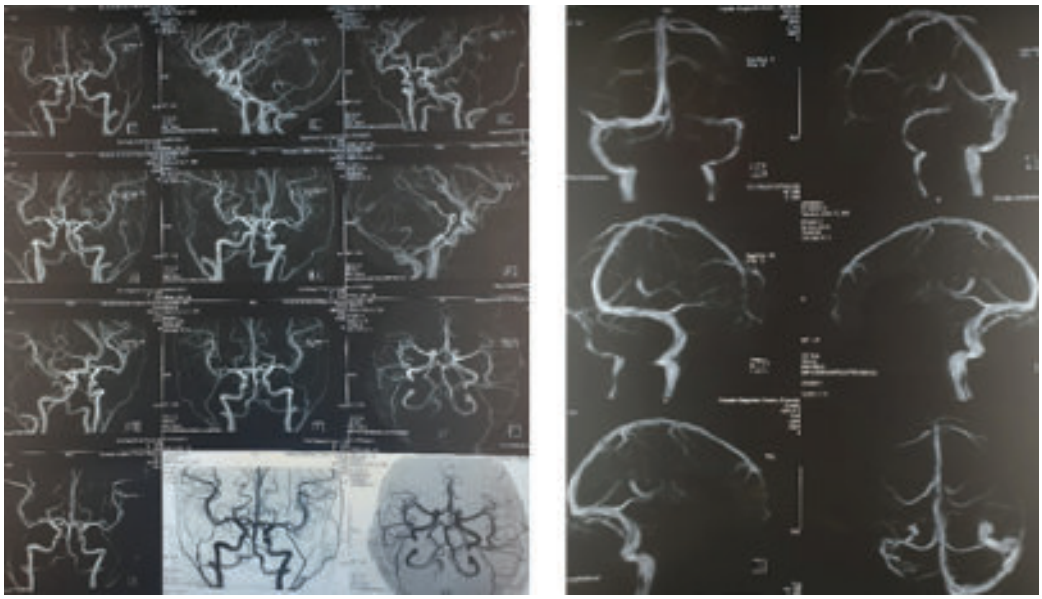

Fig.-5: *magnetic resonance angiogram (A) demonstrates no pathology along the major arterial territory. Magnetic resonance venogram (B) showing the stenosis and filling defect involving right sided transverse and sigmoid sinus, internal jugular vein.*

go for a CT scan of brain with 3D reconstruction which will be helpful during cosmetic correction and referred this patient to the Plastic and reconstructive surgery for aesthetic improvement with autologous fat transfer.

Discussion

Papillitis is a vision threatening ocular inflammatory disorder, associated with various isolated ocular and systemic diseases. The fundoscopic features often simulates papilledema which creates a diagnostic dilemma. Our reported case presented with a rare clinical entity known as Progressive hemifacial atrophy which is characterized by progressive atrophy of one side of the face involving the skin and underlying soft tissue. This disease possesses numerous neurologic as well as ophthalmologic manifestation, among them papillitis is one of the earliest presentation[1].

The etiology of PHA is not well known. Possible association include traumatic, infectious, autoimmune, inflammatory process, sympathetic nervous system disorders, trigeminal neuralgia, and hereditary disorders[2]. This entity has been associated with multiple ophthalmologic and neurologic manifestations occurring in up to 46% and 60% of cases, respectively[3]. Notably, there could be a considerable delay between the diagnosis of PHA and onset of these complications[4]. The most common neurological abnormalities described in association with PHA include seizures, headaches, movement disorders, neuropsychological symptoms, and focal neurological deficit[4]. However, our patient presented with the features of dimness of vision only.

The reported ophthalmologic manifestations are enophthalmos, eyelid atrophy, ptosis, corneal changes, heterochromia of the iris, strabismus, uveitis, retinal vasculitis, ipsilateral and contralateral third nerve paresis, glaucoma, neuroretinitis, and macular edema[5]. Comparing this, our patient had features of right sided enophthalmos with papillitis. Though this has been subjected in several case reports which supports an underlying immune-mediated inflammatory process as the pathogenesis of this syndrome. However, frequent association of PHA with other autoimmune diseases support this hypothesis[7-10].

Considering the treatment options, various immunosuppressant and biological therapies have been tried[11]. Still now, autologous free fat transfer is the accepted modality for cosmetic improvement[12-14]. In case of pediatric patient, Mishra et al. proposed Botulinum toxin A for pain reduction in case of PHA [15]. However, further studies are required to analyse the best management options for this rare disorder.

Conclusion:

Papillitis can manifest as initial presentation of Progressive hemifacial atrophy or can occur several

years after the initial diagnosis of this syndrome. However, multidisciplinary approach, including Ophthalmologists, Neurosurgeon, Neurologist, Rheumatologists, and Plastic surgeons are required for optimum management of this rare entity.

Declarations:

Authors' contributions:

Conception, diagnosis and design, Radiological diagnosis
Dr. Akhlaque Hossain Khan

Manuscript preparation, Technical revision, Manuscript editing and revision
Dr. Nazmin Ahmed, Dr. Vijay Kumar Raut

Literature search
Dr. Nazmin Ahmed

Final approval of manuscript
Dr. Akhlaque Hossain Khan

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Patient Consent: An informed written consent was obtained from the patient.

Ethics Approval: There is no ethics issue in this paper.

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