Case Report

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Idiopathic unilateral complete oculomotor nerve palsy: a case report of diagnostic quandary

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Introduction and importance: When compared to other cranial nerve palsies idiopathic unilateral oculomotor nerve palsy with pupillary sparing is one of the least noted neurological conditions. Moreover, there lies a series of diagnostic dilemmas to come into a final diagnosis resulting in several array of clinical investigations. Hence, there is a delay in prompt management.

Case summary: An elderly female without any known comorbidities presented with the complaint of headache, dizziness and dropping of left eyelid. Several arrays of diagnostic workups was done to come to a diagnosis, but even with rigorous laboratory investigations and radiological examinations, a common working diagnosis could not be made. Hence with a diagnosis of exclusion after proper neurological and neuro-ophthalmological examination, idiopathic unilateral common oculomotor nerve palsy was identified for which improvement with steroids was noted in the patient.

Discussion: Idiopathic unilateral complete oculomotor nerve palsy is considered as a diagnosis of exclusion when all the diagnostic parameters fail to signify and positive results. The vague symptomatic presentation of the disease condition further compels the treating physician to carry out several panels of laboratory to radiological investigations. But if identified in time the treatment modality is straightforward.

Conclusion: The diagnostic quandary in timely identification of such disease conditions needs a pertinent diagnostic guideline so as to avoid the unwanted panel of investigations.

Keywords: cranial nerves, diagnostic quandary, idiopathic, oculomotor nerve palsy

Introduction

Isolated cranial nerve III palsy is a common neurosurgical presentation. It can result from either congenital or acquired causes. Intrauterine infection and trauma during delivery are common congenital causes of oculomotor nerve palsy[1]. Acquired causes can be due to trauma, intracranial neoplasm, meningitis, encephalitis, cerebral artery aneurysm, and blockage of small vasculature in hypercoagulable states[2]. Overall the incidence of acquired third nerve palsy is also quite minimal with older age group and elderly demographic groups of people under high risk. However, in a few cases, the etiology remains hidden, and literature shows that such idiopathic cases are sporadic, and only a few case reports shed light on its occurrence. The reported incidence of such idiopathic cases is 12–14% in children, but data are lacking in terms of incidence in adults[3]. Diagnostic workup is demanding in such a way that an in-depth, comprehensive laboratory and radiological workup becomes mandatory so as to accurately pinpoint the correct diagnosis. Details regarding its diagnosis and treatment are lacking in the literature, and the management is thus individualized and depends upon the expertise of the treating physician[4,5]. Moreover, a controversy always exists regarding several arrays of diagnostic workups and lines of management, due to a lack of evidence-based guidelines. We here report a case of idiopathic unilateral cranial nerve palsy along with the diagnostic workup and a review of the literature. This case report has been written in line with Surgical Case Reports (SCARE) guideline[6].

HIGHLIGHTS

- Isolated cranial nerve III palsy is a common neurosurgical presentation. It can result from either congenital or acquired causes.
- There lies a series of diagnostic dilemmas to come into a final diagnosis resulting in several array of clinical investigations.
- Although recent advances in neuroimaging have made early diagnosis easier, management of patients with isolated cranial nerve III palsies still remains challenging.
- The diagnostic quandary in timely identification of such disease conditions needs a pertinent diagnostic guideline.

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Case summary

An 87-year-old female presented to the outpatient department with a complaint of headache since one week, which was insidious in onset, over left frontoparietal region sharp throbbing in type, pain non-progressive without any aggravating and relieving factors associated with left periorbital pain and mild dizziness which was non-rotatory in type and associated with few episodes of vomiting, non-projectile in nature, small in amount, not blood and bile mixed. The patient also complained of associated inability to open the left eye, restricted left eye movements, and diplopia. It developed since one week and was acute in onset, but the patients relatives noticed it a few days later. Initially, it was mild difficulty in opening the left eye but later progressively worsened over the past few days until the patient has severe difficulty opening the left eye. The patient had no any features suggestive of progressive fatigability over the course of the day. The patient had no history of trauma. Past medical and surgical history was non-significant. Clinical examination revealed left upper lid complete ptosis with pupillary sparing with impaired levator function, and anomalous eye movements when attempting elevation, depression, or adduction of the left eye in motility testing. (Table 1) Ptosis was associated with pain and tearing in the left eye periodically. (Fig. 1) A detailed workup was performed: neurological and ophthalmological examination, blood pressure measurement, laboratory tests, and MRI of the brain, (Fig. 2)(Fig. 3) magnetic resonance venography (MRV), magnetic resonance angiography (MRA) of the brain(Fig. 4) of the brain was done. (see Table 4 for the diagnostic workup and the results). Basic laboratory tests consisted of complete blood count (CBC), blood chemistry, coagulation screening, inflammatory markers, urinalysis, fibrinogen degradation products (FDP), D-dimer. The patient's routine laboratory examination was normal with no evidence of diabetes mellitus (DM), hyperlipidemia, inflammation, and infection.(Table 2) MRI was also unremarkable without intracranial mass lesions and acute infarction. The authors ruled out various possible underlying causes, including microvascular ischemia, aneurysm, trauma, neoplasm, inflammation, and neurosurgical intervention. The systemic review was unremarkable for any central nervous system (CNS), cardiovascular or systemic illness. No drug history or allergies were reported by the family.

Table 1

Ophthalmological examination findings.

	Left eye	Right eye
Upper eyelid	Drooping until covering the limbus lid swelling	Normal
Sclera	Normal	Normal
Cornea	Normal	Normal
Conjunctiva	Congested	Normal
Pupil size	3mm, round	3mm,round
Direct light reflex	Normal	Normal
Consensual light reflex	Normal	Normal
Accommodation reflex	Impaired	Normal
Fundus	Normal	Normal
Visual acuity	6/6	6/6
Medial movement	Restricted	Intact
Lateral movement	Restricted	Intact
Downward movement	Restricted	Intact
Upward movement	Restricted	Intact

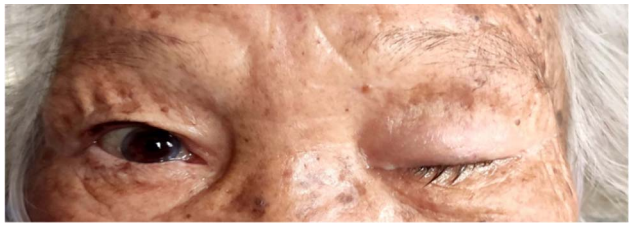

Figure 1. Picture depicting left sided complete ptosis.

An ophthalmological examination was done, and findings have been shown in Table 1.

Initial laboratory investigation including complete blood count, C reactive protein, and blood random sugar was unremarkable, as shown in Table 2.

MRI brain has been ordered for intracranial anomalies reporting no obstructive lesion in the orbit and anterior cranial fossa as given in (Figs. 2, 3)

Further tests were ordered for hypercoagulability and systemic illness and the results have been shown in Table 3.

Despite the detailed history and laboratory workup, the exact cause of the disease remained unknown, and the condition has been labeled as idiopathic unilateral cranial nerve palsy. The recommended therapy was steroid treatment. The patient, therefore, received intravenous methylprednisolone 1 gm once a day for total of 5 days then patient was discharged on oral corticosteroid (DEFLACORT) 60 mg once a day with tapering for 2 weeks. After diagnosis and symptomatic improvement patient was discharged after 10 days, during discharge there was significant improvement in diplopia, angle of squint, ocular movement and mild improvement in ptosis(Fig. 5) and the patient was instructed for regular follow-up.

Discussion

Idiopathic unilateral third nerve palsy and it's diagnosis and management has always been a matter of dilemma in terms of the right choice of investigation and line of management. Moreover several factors could be the possible causation leading to the condition, even the third nerve's innate anatomy. The third cranial nerve also known as oculomotor nerve and has two major components: -Outer parasympathetic fibers that supply the ciliary muscles and the sphincter pupillae-Inner somatic fibers that supply the levator palpebrae superioris in the eyelid (which retracts the upper eyelid) and the four extra-ocular muscles (superior, middle, inferior recti, and inferior oblique)[7]. The nerve fibers leave the midbrain through the most medial part of the cerebral peduncle and enter the interpeduncular cistern. After the oculomotor nerve emerges from the interpeduncular fossa, it enters the cavernous sinus slightly lateral and anterior to the dorsum sellae. It enters the orbit through the superior orbital fissure, after exiting the cavernous sinus, to innervate the extraocular muscles[7]. Third, the cranial nerve's long course through important anatomical landmarks can also be a potential hidden factor in many cases where the definitive cause has not been noted out despite rigorous diagnostic workup[7,8]. The age- and sex-adjusted annual incidence of acquired third nerve palsy was 4.0 per 100 000[8]. The annual incidence in patients older than 60 was greater than patients younger than 60[8]. Available studies

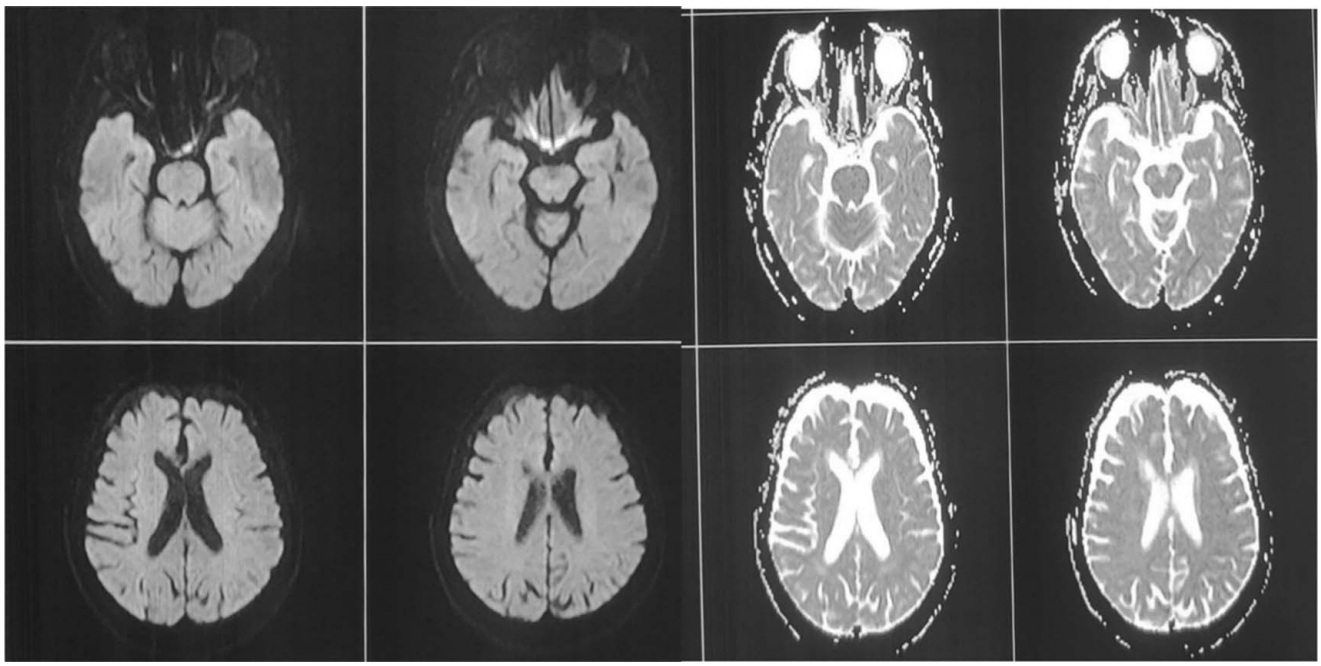

Figure 2. MRI brain ADC and DWI sequence suggestive of no any gross abnormality in the brain. ADC, apparent diffusion coefficient; DWI, diffusion weighted imaging.

have shown that the most common causes of acquired third nerve palsy were presumed microvascular (42%), trauma (12%), compression from neoplasm (11%), post-neurosurgery (10%), and compression from aneurysm (6%). Along with it Ten patients (17%) with microvascular third nerve palsies had pupil involvement, while pupil involvement was seen in 16 patients (64%) with compressive third nerve palsies[8]. The lab workup for CNIII palsy involves obtaining blood pressure recordings, complete blood counts (CBC), HbA1C, erythrocyte sedimentation rate (ESR), and imaging studies using CT angiography (CTA) or MRI of brain, particularly if an aneurysm is suspected[9]. The evolution of new imaging techniques, such as

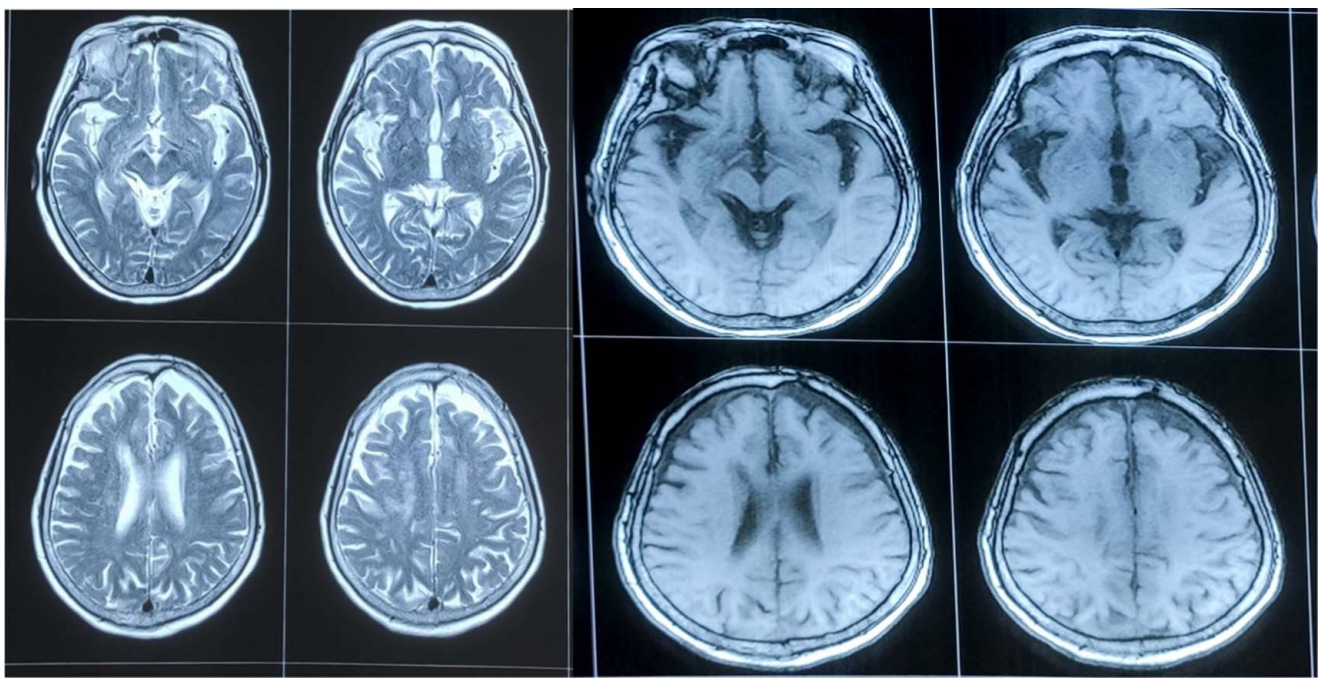

Figure 3. MRI brain T2 and FLIAR axial section suggestive of no any gross abnormality in the brain parenchyma. FLIAR, fluid attenuated inversion recovery.

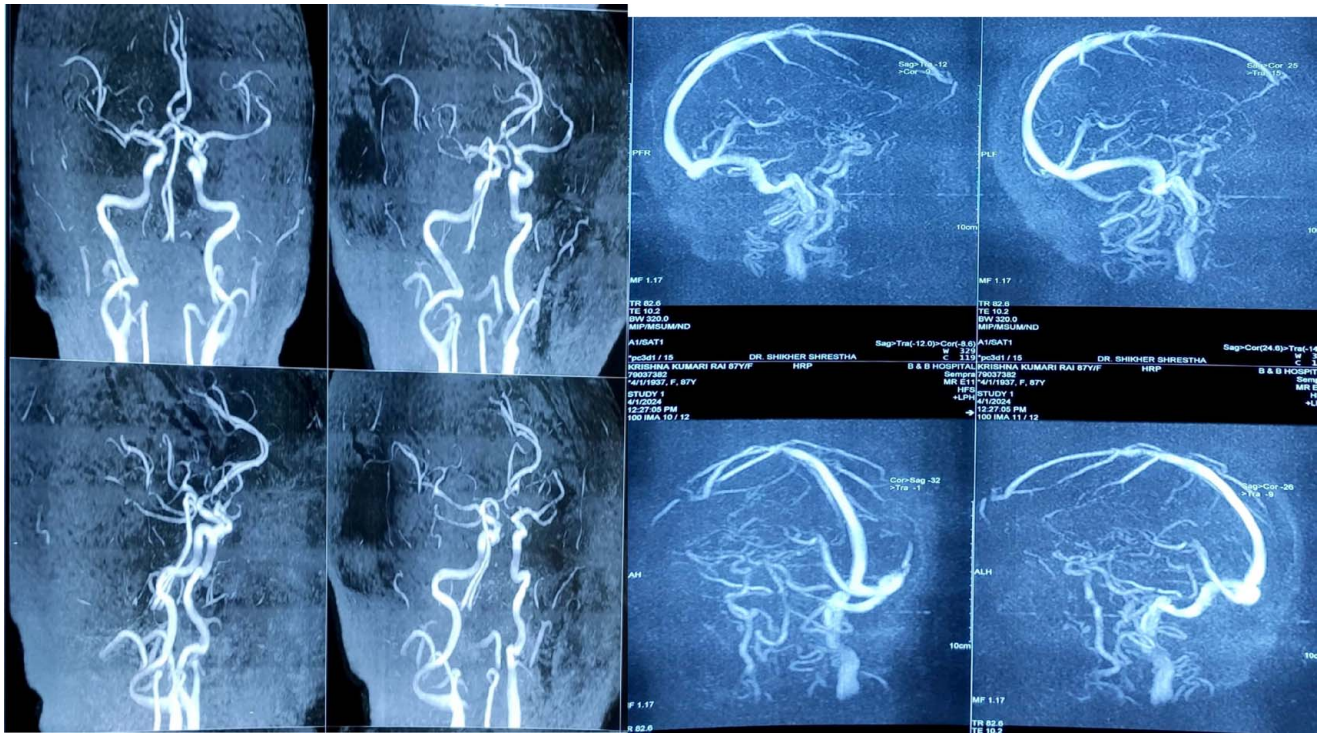

Figure 4. Magnetic resonance angiography and magnetic resonance venography of brain suggestive of normal findings hence excluding chances of aneurysm and cerebral venous sinus thrombosis in the patient.

computed tomography angiography and magnetic resonance angiography, have provided new imaging options for clinicians, which makes it easier to come to a final diagnosis[10]. While the use of MRI as an early diagnostic tool for isolated ocular nerve palsy remains controversial, a normal MRI obtained in patients who experience acute onset diplopia from isolated ocular motor palsy may help to allay anxiety and fears of brain tumor or other serious disease, which in turn may have significant social, psychological and even economic benefit in terms of productivity. This value is, however, inherently subjective and difficult to measure[11]. The diagnosis and management of third nerve dysfunction varies according to the age of the patient, characteristics of the third nerve palsy, and presence of associated symptoms and signs. Indeed, third nerve palsies may be partial or complete, congenital or acquired, isolated or accompanied by signs of more extensive neurological involvement. Depending upon the types the depth of investigations and management may vary. They can result from lesions located anywhere from the oculomotor nucleus to the termination of the third nerve in the extra-ocular muscles within the orbit[12]. Despite recent advances in modalities such as noninvasive neuroimaging, which facilitate early diagnosis; however, management of a patient presenting with an isolated third nerve palsy remains a challenge due to the lack of any obvious peculiar findings that may help to guide towards the diagnosis[12]. Oculomotor nerve palsy lacks any significant morbidity or life-threatening complications but still globally, viewed as a diagnostic concern in clinical practice due to its nature of hampering the quality of life of the patient[13]. Literature has also shown that there are also some untypically causes, such as schwannoma, meningioma, and radiation, which

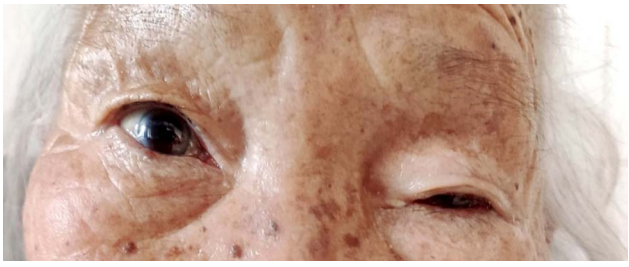

Figure 5. Picture representing mild improvement of left sided ptosis after IV steroids.

Table 2

Initial laboratory investigations.

Investigation	Result	Unit	Reference range
Hemoglobin	12.1	g/dl	13–17.5
White blood cell count	7800	Cells/mcl	4500–11 000
Red blood cell count	348 000	Cells/mcl	450 000–600 000
Platelet count	225 000	Cells/mcl	150 000–450 000
Mean corpuscular volume	76	Fl	80–100
Mean corpuscular hemoglobin concentration	32.2	g/dl	32–36
RDW	47.4	%	12–16
CRP	21		
Blood random sugar	96	mg/dl	80–126

CRP, C reactive protein: RDW, red cell distribution width.

Table 3

Hypercoagulability and systemic illness results.

Investigation	Result	unit	Reference range
ANA	1:40 (negative)		
Protein C	124.1	%	72–160
Protein S	81.7	%	60–150
Antithrombin 3	96	%	80–120
Factor V laden mutation	Negative		
Fibrinogen degradation product (FDP)	> 160 <320	Ug/ml	< 5.0
D-dimer	0.4	Mg/l	0-0.50

ANA, anti nuclear antibody.

may also lead to Oculomotor Nerve Palsy[13]; however, despite appropriate imaging studies, clinical examination and laboratory workup and no any significant radiation history there is no such evidence of atypical pathophysiology in our case.

Although recent advances in neuroimaging have made early diagnosis easier, management of patients with isolated cranial nerve III palsies still remains challenging. Recommended treatment options vary according to the etiologies. For example, the treatment of palsies with vascular causes largely centers on supportive therapy (i.e. eye patching and prism therapy). In contrast, patients with idiopathic cranial nerve III palsies have been reported to respond well to steroid treatment and to have good prognosis[14]. Unilateral oculomotor palsy is more difficult to localize particularly if it is isolated with no other signs. However, unilateral oculomotor palsy with other long tract involvement indicates a fascicular lesion. Varying manifestations can be seen based on the course of the nerve as it crosses the red nucleus,

Table 4

The results of diagnostic workup for cranial nerve III palsy.

Underlying cause	Diagnostic test	Results
Inflammatory and paraneoplastic process	Lumbar puncture	Normal
Intracranial mass lesions and acute infarction	MRI brain and orbits	Unremarkable
Diabetes	HbA1C	Normal
Inflammatory process or infection	CBC and inflammatory markers	Normal
Aneurysm	MRA	Unremarkable
trauma and neurosurgical intervention	MRI, CT and past history	Unremarkable
Alcohol	Urinalysis	Negative

CBC, complete blood count; CT, computed tomography; MRA, magnetic resonance angiography.

superior cerebellar peduncle, substantia nigra, and cerebral peduncle, thus resulting in various syndromes[15]. Moreover evidence-based guidelines should be developed so as to accurately and on time diagnosis of such idiopathic causes of third nerve palsy so as to start treatment early and improve the quality of life of the patient's.

Conclusion

Neurological conditions such as idiopathic complete oculomotor nerve palsy is a diagnosis of exclusion hence the diagnostic dilemma. Moreover, the treatment modality is not that tedious and quite straightforward along with significant improvement among the patients after prompt treatment. However, there is the utmost need of the utilization of several arrays of clinical to radiological investigations to come to a diagnosis as seen in our case (Table 4) where an in-depth diagnosis workup is mandatory.

Ethical approval

Ethical approval is exempted in case of case reports in our Institution. Whereas written informed consent have been taken from patient herself.

Consent

Written informed consent has been taken from the patient herself and can be made available if asked upon by chief editor.

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Author contribution

S.G., K.B.: conceptualization; data curation; formal analysis; methodology; project administration; supervision; writing—review and editing. K.B. : data curation; S.S., D.S., K.R., S.B.: project administration; supervision.

Conflicts of interest disclosure

The authors declare no conflicts of interest.

Research registration unique identifying number (UIN)

No clinical trials were conducted. Information was obtained from patient after written informed consent.

Guarantor

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Data availability statement

The dissemination of the article data is freely accessed.

Provenance and peer review

This entitled paper was not invited.

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