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Case Report

Sixth nerve palsy: Three case reports on different etiologies

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ABSTRACT

Sixth nerve palsy is the commonest isolated cranial nerve palsy. Possessing the longest course, it is susceptible to damage through a myriad of etiologies, hence termed 'false localizing sign'. Though frequently being a benign process with spontaneous recovery, it may be due to serious underlying pathology. Therefore, sixth nerve palsy requires careful clinical workup with tailored investigation. We are presenting three cases of isolated sixth nerve palsy having different etiologies.

1. Introduction

Abducens nerve (Cranial Nerve VI) palsy is the commonest ocular motor nerve palsy.[1] Being the longest cranial nerve, it is susceptible to damage from vast pathological processes; vascular, traumatic, neoplastic, infectious, inflammatory, demyelination and also idiopathic. Microvascular ischemia is the commonest in adults over age of 50 years who are suffering from vascular comorbidities; diabetes mellitus, hypertension, hyperlipidemia.[1] Neoplasms are much more common in children.[2] Aneurysms are uncommon comprising 0-3% of the cases.[3] 22-30% cases present as idiopathic.[2]

2. Case Report

We are presenting three cases of isolated sixth nerve palsy that reported to us having three different etiologies.

2.1. Case 1

An 11 year old boy presented to us with dimness of vision, headache and occasional diplopia for approximately 06 months. O/E BCVA was 6/12 OD and 6/9 OS. Normal ocular findings except limited abduction on left gaze (Figure 1). Diplopia was present throughout levoversion, levoelevation & levodepression.

MRI revealed a large lobulated soft tissue intensity mass having both solid and cystic components in the sella and suprasellar region extending up to the floor of the third ventricle. Visual field analysis showed bi-temporal hemianopia. (Figure 1)

Working diagnosis of craniopharyngioma was made and he was referred to Neurosurgery department.

2.2. Case 2

A 45 year old female presented with intermittent headache for 06 months and blurred vision in right eye for one month. On examination, BCVA was 6/24 in the right eye and 6/6 in the left eye. Limited abduction of the

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Table 1: Summaries of 09 retrospective studies on sixth nerve paresis

	Etiologies of acquired sixth nerve palsy							
	Schrader[3] 1960	**Rucker[4] 1966**	**Johnston[5] 1968**	**Robertson[6] 1970 (Children)**	**Rush[7] 1981**	**Patel[8] 2004**	**Bagheri[9] 2010**	**Jung[10] 2019**
Sample size	104	607	158	133	419	137	33	486
				Etiologies %				
Neoplasm	7	33	13	39	15	5	2	5
Trauma	3	12	32	20	17	12	18	5
Aneurysm	0	3	1	3	3	2	0	2
Ischemic	36	8	16	0	18	16	1	56
Miscellaneous*	30	24	30	29	18	19	6	4
Undetermined**	24	20	8	9	29	26	6	28

* Leukemia, migraine, pseudotumor cerebri, multiple sclerosis.
** Undetermined cause. All routine investigations normal, imaging normal.

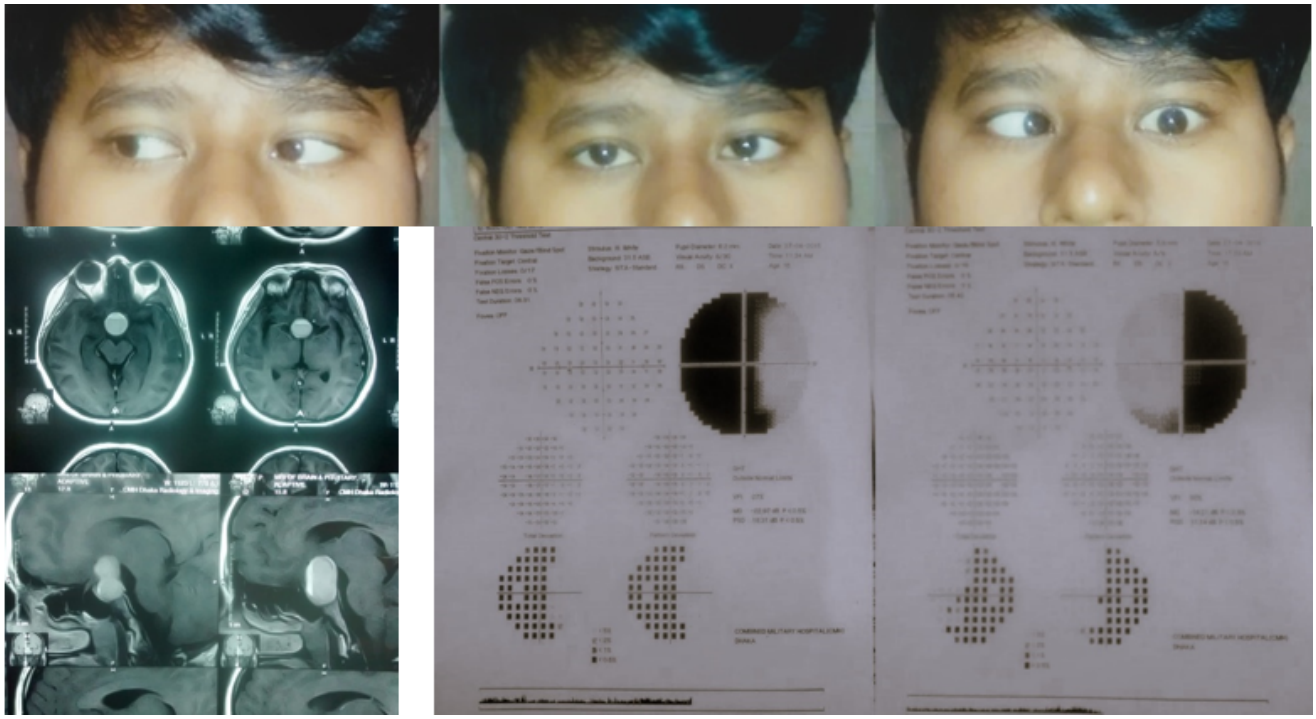


Fig. 1: A: Left gaze palsy; **B:** MRI showing solid midline tumor with cystic component, Craniopharyngioma; **C:** HVFA showing bitemporal hemianopia

left eye (Figure 2). Fundus showed right infero-temporal branch retinal vein occlusion (BRVO) with macular edema (Figure 2). Routine hemogram was normal, she was normotensive and euglycemic. MRI of brain revealed a large internal carotid artery (ICA) aneurysm on the left side (Figure 2). MRA was done (Figure 2). Temporary Frosted glass was prescribed for the left eye.

She was referred to Neurosurgery department.

2.3. Case 3

A 55 year old male reported with sudden onset diplopia on left gaze; noticed by him for two days. On ocular evaluation, BCVA was 6/6 in his both eyes. Ocular findings were normal except limited abduction on left side (Figure 3). Diplopia was present throughout levoversion, levoelevation and levodepression. Routine hemogram was normal, he was diabetic and had hyperlipidemia. He has been suffering from diabetes mellitus for 05 years and was on oral hypoglycemic agent. MRI of Brain reported no intracranial abnormality to cause left sixth cranial palsy.

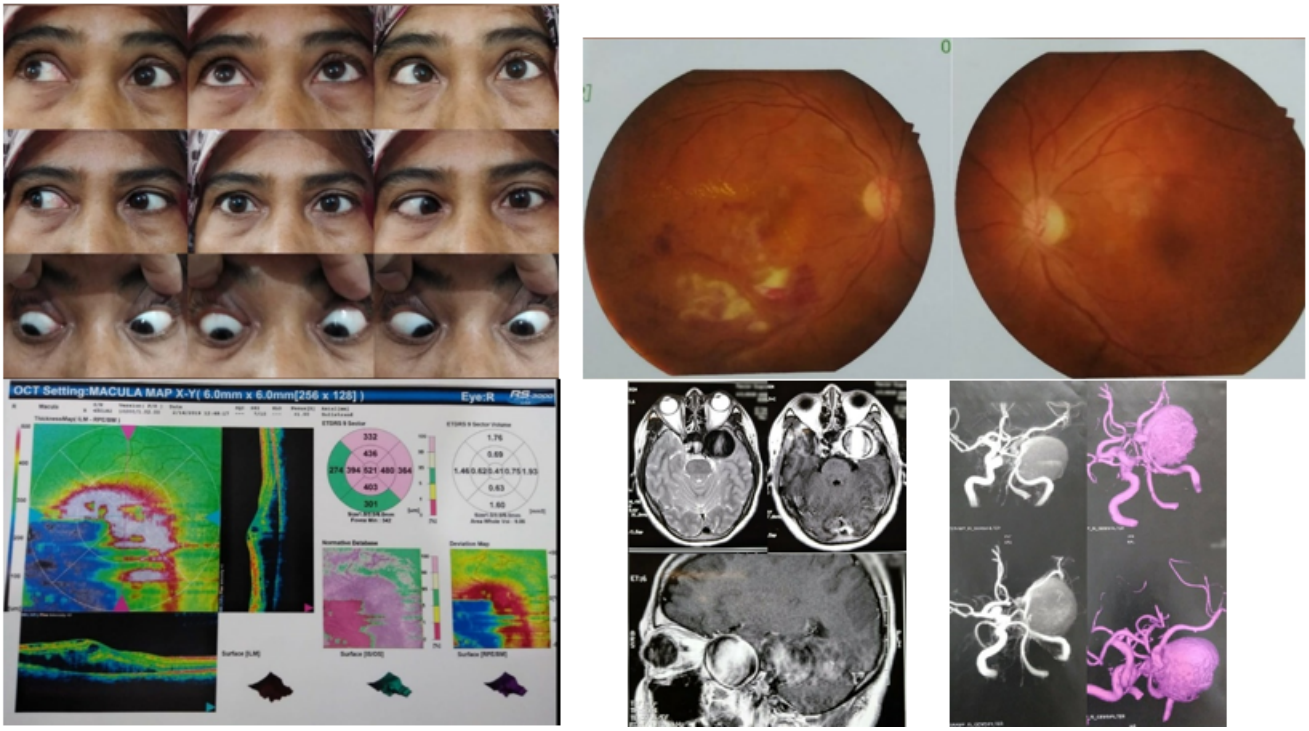


Fig. 2: A: Nine diagnostic gazes showing left gaze palsy; **B:** Color fundus photograph showing infero-temporal BRVO with macular edema in right eye; **C:** OCT Macula showing macular edema; **D:** MRI brain showing left sided Internal Carotid Artery aneurysm; **E:** MRA showing aneurysm of internal carotid artery.

He was referred to Medicine department for review and controlling of the comorbid conditions. Temporary Frosted glass was prescribed for the left eye.

During follow up after 06 months, his conditions improved with no diplopia and ocular motility showed normal in all gazes. (Figure 3)

3. Discussion

The sixth cranial (Abducens) nerve palsy is the most common oculomotor paralysis in adults and the second-most common in paediatric age group. The lateral rectus muscle is controlled by the sixth cranial nerve, which Abducts the eye. Unilateral sixth nerve palsy causes an incomitant esotropia due to the unopposed action of the antagonistic medial rectus muscle.[11]

Diagnosis of the manifestation of sixth nerve palsy is not straight forward. Studies on etiology of sixth nerve palsy reports high frequencies of microvascular disease (28-46%) and idiopathic (24-31%).[10] It is widely reported that microvascular diseases are a common cause of isolated unilateral sixth nerve palsy in patients over 50 years of age.[10] Tamhankar et al. also reported sixth nerve palsy in 80.6% of patients over 50 years of age was due to microvascular disease.[1] Sixth nerve palsy from aneurysm is low 0-6%.[10] Studies have found high frequency of neoplastic etiology in children 39-45%.[10]

The causes of acute sixth cranial nerve palsy in paediatric age group is reported on a retrospective case series study of 14 paediatric patients[12] which is shown in Graph 1. Recovery rates of sixth nerve palsy is 60-87.3%.[13] Vascular and idiopathic etiologies were associated with higher natural recovery rates than other etiologies of ocular motor nerve palsies.[10] Sanders et al reported 86% experienced resolution of sixth nerve.[14]

The clinical history is always important to find out the etiology. Sudden onset suggests a vascular cause, while compressive etiology presents slow progressive sixth cranial nerve palsy. Subacute onset is associated with a demyelinating process. All patients need a complete ophthalmologic evaluation with orthoptic assessment including visual acuity, binocular function and stereopsis, motility evaluation, strabismus examination at near, and distance. Systemic examination is essential in all types of suspected neoplasms and trauma cases especially in children, because neoplasms and trauma are the most common etiologies of sixth cranial nerve palsy.[12,15]

The cardinal sign of the sixth cranial nerve palsy is esotropia, diplopia and restricted abduction in the affected eye. The esotropia of the affected eye is caused by the unopposed action of the ipsilateral medial rectus muscle. The esotropia is incomitant and the esotropia is greater on attempted abduction and on distant fixation.[16] The patient

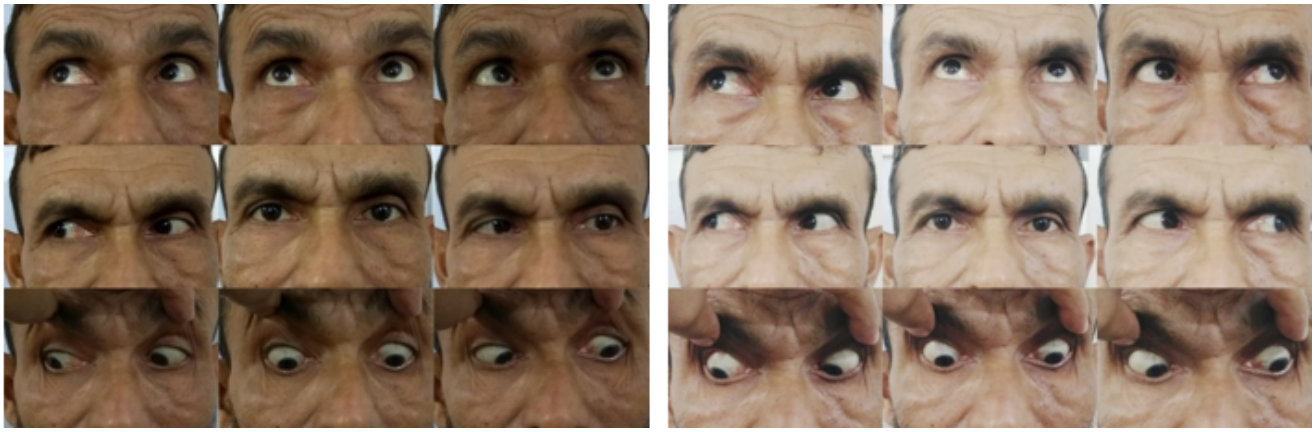
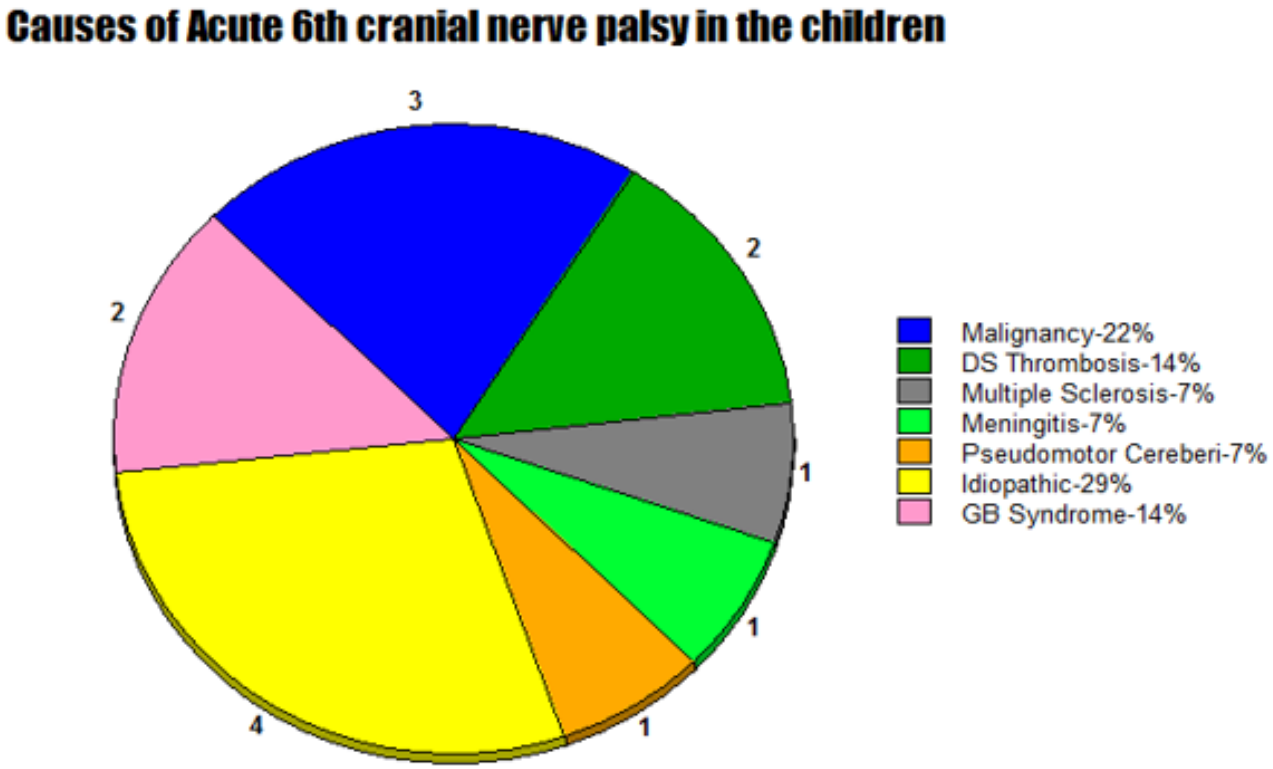


Fig. 3: Showing nine diagnostic gazes; **A:** Left gaze palsy during presentation; **B:** Follow up after 06 months showing almost complete recovery following good glycemic control.



Graph 1: Based on a retrospective study of 14 patients between Jan 01, 2002 and December 31, 2012.[12]

is usually present with a head turn toward the affected eye, to avoid abduction and also to minimize diplopia. It is important to differentiate isolated sixth nerve palsy from a gaze palsy or Internuclear Ophthalmoplegia (INO).[17]

After clinical evaluation, the most common diagnostic procedure is MRI of the brain and orbit. MRI is recommended for all patients especially under the age of 50 years, Patients younger than 50 years, associated pain or other neurologic abnormality, history of cancer, patients with bilateral sixth nerve palsy, Optic disc oedema to rule out any intracranial pathology/neoplasms.[18] Laboratory test like complete blood count, blood sugar assessment, Glycosylated haemoglobin (HbA1C), C-reactive protein, Fluorescent treponemal antibody-absorption test, VDRL or RPR, Antinuclear antibody test, Rheumatoid Factor test can be done depending on clinical evaluation and differential diagnosis. Lumbar puncture may be done if MRI report is unremarkable.[15,19]

The management of the sixth cranial nerve depends on the underlying etiology. In general, underlying or systemic conditions are treated primarily. Most patients with a microvascular sixth nerve palsy are simply observed and spontaneously recover within 3 to 6 months. The diplopia can be treated with base out Fresnel prisms, patching, botulinum toxin type-A injection to the ipsilateral medial rectus muscle, or surgery.[11–13,18–20]

The prognosis for sixth nerve palsy depends on the etiology. The recovery rate of 49.6% in 419 non-selected cases of sixth nerve palsy, and a higher recovery rate of 71% in 419 patients with microvascular causes such as diabetes mellitus, hypertension, or atherosclerosis.[20]

4. Conclusion

Sixth nerve palsy being a false localizing sign, warrants examination and tailored investigation. It may not be a benign process. Hence the clinician must consider the potential of a serious neurological process. Early diagnosis is critical in some conditions with sixth nerve palsy.

5. Conflict of Interest Statement

There are no potential conflicts of interest.

6. Declaration of Patient Consent

The authors certify that they have obtained all appropriate patients consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients assured that their names and initials will not be published.

7. Author Contributions

SMRI, SMK, SA, RA- designed the Study, procured the samples and performed the experiments, SMRI, SMK, SA, GR- provided critical input; SMRI, SA, RA -wrote the first draft of the manuscript with inputs from all co-authors; SMRI, SMK, SMBI- critical appraisal of the manuscript; All authors reviewed and approved the final version of the manuscript prior to submission.

8. Conflict of Interest

The authors declare that there are no conflicts of interest in this paper.

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None.

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