

Ali Niyaf

Senior Consultant Neurosurgeon

ADK Hospital

ORCID iD: <https://orcid.org/0000-0002-3442-0092>**Kiran Niraula**

Consultant Neurosurgeon

ADK Hospital

ORCID iD: <https://orcid.org/0000-0002-1256-292X>**Shiara Zahir**

Medical Officer

ADK Hospital

ORCID iD: <https://orcid.org/0000-0003-1848-5125>**Mohamed Sajuan Mushrif**

Medical Student

Lithuanian University of Health Sciences

ORCID iD: <https://orcid.org/0000-0001-6958-4997>**Address for correspondence:**

Dr. Kiran Niraula

MBBS, MPhil (Public Health), MS (Neurosurgery)

Consultant in Neurosurgery

ADK Hospital

Male, Maldives

E-mail: nirkiran@yahoo.com

Contact number: +960 9863880

Date of submission: 22nd January 2020**Date of acceptance:** 1st March 2020**DOI:** <https://doi.org/10.3126/njn.v17i1.28344>**HOW TO CITE**

Niyaf A, Niraula K, Zahir S, Mushrif MS. Isolated unilateral temporalis muscle hypertrophy: first case in an Indo-Aryan. NJNS. 2020;17(1):48-52

To access Nepal Journal of Neuroscience Archives, scan QR code:

Isolated unilateral temporalis muscle hypertrophy: first case in an Indo-Aryan

Abstract

Unilateral hypertrophy of temporalis muscle (isolated type) is extremely rare and only 14 cases have been reported in English literature since 1990. Although no definitive etiology is identified, stress and bruxism are among the many factors that are linked to it. Reported patient ages ranged from seven years to 62 years and most of them are Caucasians. Here we report the first ever case of an Indo-Aryan ethnic patient who presented with painful swelling of the left temporal region. Temporalis muscle biopsy confirmed the diagnosis of isolated unilateral temporalis muscle hypertrophy. This case raises the importance of considering rare diagnoses such as isolated unilateral temporalis muscle hypertrophy despite inadequate statistical data on ethnicity or geographic location of a condition.

Key words: Biopsy, Indo-Aryan, IUTMH (isolated unilateral temporal muscle hypertrophy)

Introduction

Since the first case of masticatory muscle hypertrophy reported in 1880¹, there has been an increase in the amount of entries into English literature. The most common presentation of these cases being bilateral whether isolated or combined.¹ However, unilateral and isolated temporal muscle hypertrophy is still an extremely rare condition^{1,2,3,5,6} and only 14 cases have been documented thus far (Table 1). The disease occurs mainly in adults and there have been only two cases reported under fifteen years of age.^{2,3} Due to lack of data, the demographic areas that are not focused on currently are ethnicity and race. We report here the first ever case of isolated unilateral temporal muscle hypertrophy (IUTMH) in an Indo-Aryan ethnic patient from the Maldives.

Case presentation:

A thirty two-year G6PD deficient female presented with swelling and intermittent pain in her left temporal region for six months relieved by simple analgesics but

Isolated unilateral temporalis muscle hypertrophy

would always return with time. She has no history of fever, trauma, temporo-mandibular joint disease and vision abnormalities. Her past medical and surgical history is non-significant. She has a habit of unintentional clenching of her teeth while working. There is no family history of similar symptoms or diseases according to the patient.

On physical examination, she had a large mass on the left temporal region, which was soft and non-pulsatile on palpation. The temperature and color of the overlying skin was normal. No significant changes were observed when the patient clenched her teeth. Her neurological examination showed no deficits.

Author & Year	Patient Demographics		Race	Clinical presentation	Site	Onset (months)	Treatment
	Age	Sex					
Wilson & Brown et al. 1990	43	F	Caucasian	Painless swelling, bruxism, fluctuating muscle size	R	11	Supportive
Serrat et al. 1998	15	F	NR	Swelling, temporalis muscle contraction, limitation of mouth opening	L	12	Surgery
Isaac et al. 2000	35	M	Caucasian	Painless swelling	L	8	Btx-A
Lowry & Helling et al. 2003	45	M	African American	Swelling, recurrent headaches	L	12	Systemic muscle relaxant
Prantl et al. 2005	48,	F	NR ^[1]	Painless swelling	R	12	Surgery
	^a 57	F	NR	Swelling, temporalis muscle contraction, headache	R	NR	Btx-A
Rokadiya& Malden et al. 2006	33	F	Caucasian	Painless swelling, headache	L	3	Amitriptyline, splint
Vordenbäumen et al. 2009	22	F	Caucasian	Painless swelling, recurrent headaches	R	6	Acetaminophen
Wang BH et al. 2013	17	M	Caucasian	Painless swelling, recurrent headaches	R	6	Btx-A
Katsetos et al. 2014	62	M	NR	Painless swelling, bruxism	L	96	Btx-A
	22	F	NR	Painful swelling	R	NR	Symptomatic
Damian et al. 2016	^b 24	F	NR	Temporal pain	R	NR	Corticotherapy, DMARD, cyclosporine
Tuncel et al. 2017	23	M	Caucasian	Painless swelling	R	7-10	Surgery
	38	F	NR	Painless swelling	R	7-10	No treatment
N. Zwetyenga et al. 2017	8	M	Caucasian	Painless swelling, bruxism, stressful environment	L	1	Btx-A
Ranasinghe et. Al 2018	7	F	Caucasian	Painless, Swelling	R	2	Symptomatic
Present study	32	F	Indo-Aryan	Painful swelling	L	6	Undecided

Table 1: Literature review of previous IUTMH cases summarized

KEY: R= Right, L= Left, M= Male, F= Female, Btx-A= Botulinum toxin type A, NR= Not reported, DMARD=disease modifying antirheumatic drugs

^a Same patient returned after 9 years with relapse

^b Same patient returned after 2 years with relapse

^c All cases before this study including relapses were used to calculate average age of patient (i.e. 31.2 years)



Figure 1: Coronal view of MRI image. Red arrow shows enlarged left temporalis muscle

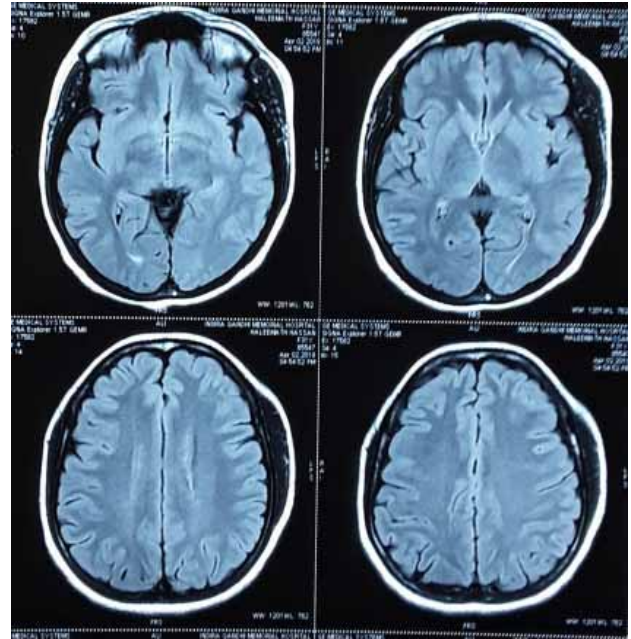
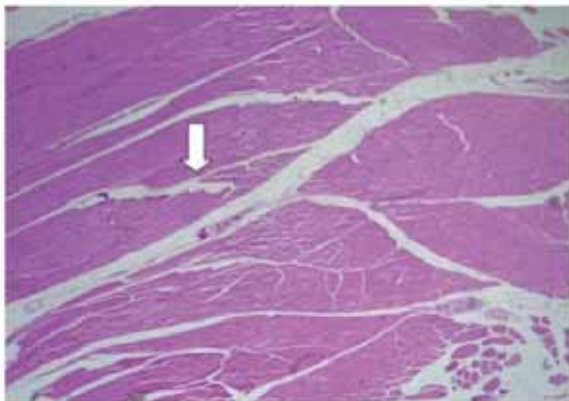
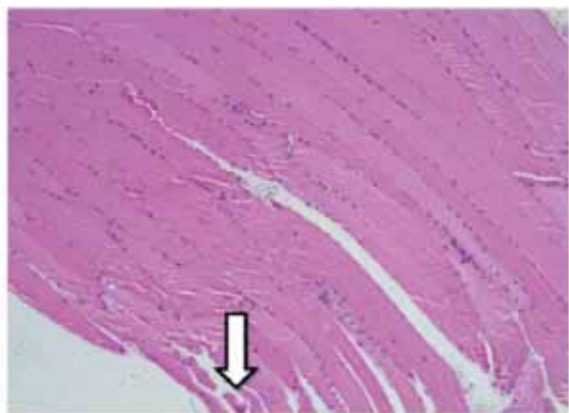


Figure 2: Axial view of MRI image. Red arrow shows enlarged left temporalis muscle

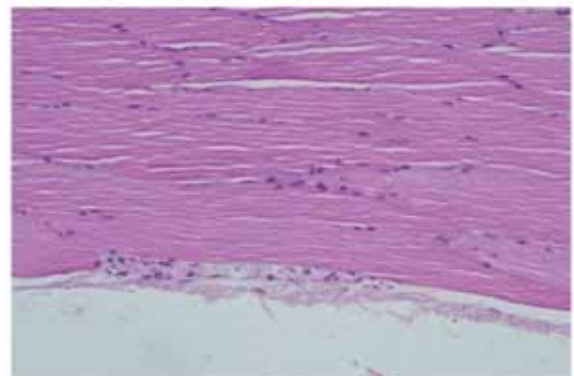


Muscle Biopsy: Endomyosial Splitting (Arrow) (H & E x 100)

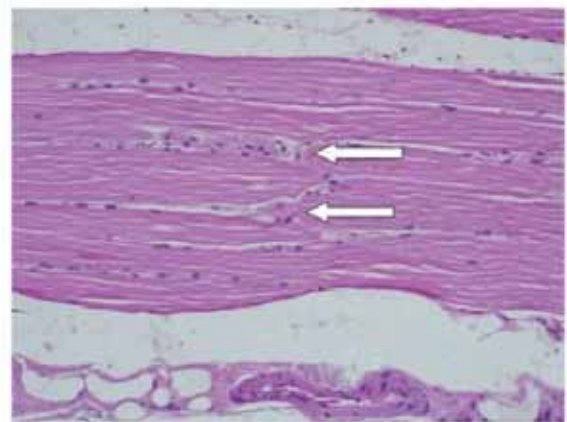


Muscle Biopsy: Internalization of Nuclei (Arrow) (H & E x 100)

Muscle Biopsy: Non-inflammatory Myopathy



Muscle Biopsy: Distorted Muscle Fibres (H & E x 400)



Muscle Biopsy: Endomyosial Fibrosis (Arrows) (H & E x 400)

Figure 3: Biopsy; Non-inflammatory myopathy

Laboratory investigations indicated normal ESR, CBC and urinalysis values. Computer tomography (CT) ruled out temporal bone pathology. Magnetic Resonance Imaging (MRI) showed left temporo-parietal superficial calvarial soft tissue edema and thickening (Figure 1 and 2) suggestive of infectious/inflammatory pathology. Left temporalis muscle biopsy was performed (Four tissue samples from different locations in the superior part of the muscle just beneath the superior temporal line) and results showed non-inflammatory myopathy (Figure 3). Therefore, according to the results of clinical and investigational examinations, the patient was diagnosed with IUTMH.

Discussion

Out of the 14 cases of IUTMH reported so far in English literature, it is interesting to note that there is a slight female predominance (Table 1). There is no distinct etiology, age of onset or side predominance.¹ To our knowledge, the only predictive causes from the reported cases are stress³ and bruxism.⁹ Other factors that have been linked to masticatory muscle hypertrophy are parafunctional jaw habits, Takayasu's disease, polymyositis⁴, temporomandibular dysfunction, myopathic diseases, rheumatic disorders, idiopathic muscle enlargement and neoplasms such as rhabdomyosarcoma or lymphomas. When it came to clinical presentation, visible swelling was seen in all cases – most of which were painless. Thirty five percent of patients complained of headaches, most of which were said to be recurring (Table 1).

The majority of the cases are adults (average being 31.2 years old) (Table 1) but our ideology must evolve along with the data to believe that this disease can affect even children under fifteen years. Fifty seven percent of the patients are Caucasian (Table 1). Here in this report we discuss a Maldivian patient from an Indo-Aryan ethnicity, which has not been documented before. Maldivians are ethnically described as Indo-Aryan¹³ and speak a language called Dhivehi, which is a member of the southern group of Indo-Aryan languages.¹⁴

In order to reach the diagnosis of IUTMH radiological tests were performed after physical and neurological examination. Cranial Magnetic Resonance Imaging (cMRI) was done in most of the cases.^{2,3,5,6} However, ultrasound scans were carried out as well. The recommended method for confirming diagnosis is muscle biopsy and histological examination.¹¹ Additional tests include stomatological/oral examinations to check for signs of bruxism or other dental anomalies^{3,4}; magnetic resonance angiography to see vessel abnormalities; and

electromyography for signs of polyphasia, dystonia, etc.⁵ As for laboratory investigations, they were either not done or not reported in a few cases^{6,12} but most had a focus on ESR and CRP^{2,4,5,6,7} as inflammatory markers and in one case serum creatine kinase was checked for possible muscle damage such as in rhabdomyolysis.³

Three main treatment methods are still practiced until now and can be intriguingly seen even in the first three cases of IUTMH reported (Table 1). The first ever case reported by Wilson & Brown in 1990 was treated supportively¹⁵, the next one in 1998 was treated surgically¹⁶ and the third one in 2000 was treated by Btx-A.¹² Supportive treatment has the widest range of applications including systemic muscle relaxants⁷, Amitriptyline (antidepressant)⁸, Acetaminophen/paracetamol⁵ and even corticosteroids and disease modifying antirheumatic drugs.⁴ Btx-A has been successfully used in four cases since 2000^{3,6,9} however Isaac¹² reported that the muscle recovers when new neuro-muscular junctions form after a period of two to four months. Hence repeated intramuscular injections of Btx-A are necessary. The best method is still debatable depending on the patient's concern, based on cost, hospitalization time or repeated treatments; hence, many differ in their opinions.

Conclusion

IUTMH is very rare among Indo-Aryans, and requires exclusion of other more common diagnoses, as well as further investigation into aetiologies and comorbidities when considered.

References

1. Legg JW. Enlargement of the temporal and masseter muscles on both sides. **Trans PatholSoc L.** 1880; 31:361–6.
2. Ranasinghe J, Wickramasinghe C, Rodrigo G. Isolated unilateral temporalis muscle hypertrophy in a child: a case report with literature review. **BMC Pediatrics**, 2018; 18(1):71. <https://doi.org/10.1186/s12887-018-1061-7>
3. Zwetyenga N, Hallier A, Girodon M, Levasseur J, Loison-Robert L, Moris V. Isolated unilateral temporalis muscle hypertrophy: First case in an 8-year-old boy and review. **Journal of Stomatology, Oral and Maxillofacial Surgery.** 2018; 119(1):61-6. <https://doi.org/10.1016/j.jormas.2017.10.010>
4. Damian L, Botar Jid C, Rogoan L, Dinu C, Maniu A, Fodor D, Rednic S, Simon S. Unilateral temporal myositis heralding polymyositis: ultrasonographic and elastographic findings. Case report. **Medical**

- Ultrasonography.** 2016;18(1):123-6. <https://doi.org/10.11152/mu.2013.2066.181.boj>
5. Vordenbäumen S, Groiss S, Dihné M. Isolated Unilateral Temporal Muscle Hypertrophy: A Rare Cause of Hemicranial Headache. **Headache: The Journal of Head and Face Pain.** 2009; 49(5):779-82. <https://doi.org/10.1111/j.1526-4610.2009.01393.x>
 6. Prantl L, Heine N, Ulrich D, Eisenmann-Klein M. Recurrence of Isolated Unilateral Temporalis Muscle Hypertrophy: Case Report. **Aesthetic Plastic Surgery.** 2005; 29(6):574-5. <https://doi.org/10.1007/s00266-004-0122-5>
 7. Lowry TR, Helling E. Unilateral temporal muscle hypertrophy: a rare clinical entity. **Ear Nose Throat J.** 2003; 82(3):198-9. PMID: 12696240
 8. Rokadiya S, Malden NJ. Variable presentation of temporalis hypertrophy—A case report with literature review. **Br Dent J.** 2006; 201(3):153-5. <https://doi.org/10.1038/sj.bdj.4813850>
 9. Katsetos CD, Bianchi MA, Jaffery F, Koutzaki S, Zarella M, Slater R. Painful unilateral temporalis muscle enlargement: reactive masticatory muscle hypertrophy. **Head Neck Pathol.** 2014; 8:187–93. <https://doi.org/10.1007%2Fs12105-013-0480-x>
 10. Harriman DG. The histochemistry of reactive masticatory muscle hypertrophy. **Muscle Nerve.** 1996; 19:447–56. DOI: 10.1002/(SICI)1097-4598(199611)19:11<1447::AID-MUS9>3.0.CO;2-C
 11. Isaac AM, Moss CE, Whear NM. Unilateral temporalis muscle hypertrophy managed with botulinum toxin type A. **Br J Oral Maxillofac Surg.** 2000; 38(5):571–2. <https://doi.org/10.1054/bjom.2000.0298>
 12. Levinson, David (1947). **Ethnic groups worldwide: a ready reference handbook.** Oryx Publishers. ISBN 978-1-57356-019-1.
 13. Maloney, Clarence. “Maldives People”. **International Institute for Asian Studies.** Retrieved 22 June 2008.
 14. Wilson PS, Brown AMS. Unilateral temporalis muscle hypertrophy: Case report. **Int J Oral Maxillofac Surg.** 1990; 19(5):287-8. [https://doi.org/10.1016/S0901-5027\(05\)80422-9](https://doi.org/10.1016/S0901-5027(05)80422-9)
 15. Serrat A, García-Cantera JM, Redondo LM. Isolated unilateral temporalis muscle hypertrophy. **Int J Oral Maxillofac Surg.** 1998; 27:92-3. [https://doi.org/10.1016/S0901-5027\(98\)80302-0](https://doi.org/10.1016/S0901-5027(98)80302-0)