ABSENT MECKEL CAVE WITH CISTERNAL TRIGEMINAL NERVE ATROPHY

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CLINICAL PRESENTATION

PAROXYSMAL, ELECTRIC SHOCK-LIKE, UNILATERAL FACIAL PAIN

LASTING TO < 2 MINS IN ONE OR MORE OF THE TRIGEMINAL DIVISIONS.

PAIN TRIGGERED BY INNOCUOUS STIMULI

CHEWING, LIGHT TOUCH, BREEZE TRIGGER PAROXYSMS.

TYPICALLY AFFECT THE RIGHT SIDE

BRANCHES V2 AND V3 ARE MOST COMMONLY AFFECTED. ISOLATED V1 TRIGEMINAL NEURALGIA IS RARE (6%)



DIAGNOSIS:

TRIGEMINAL NEURALGIA

ATYPICAL PRESENTATION

COMPLETE IPSILATERAL FACIAL HYPESTHESIA

PERSISTENT NUMBNESS ACROSS THE ENTIRE AFFECTED SIDE OF THE FACE.

DEPRESSED CORNEAL REFLEX

WEAKENED AUTOMATIC BLINK WHEN THE SURFACE OF THE CORNEA IS TOUCHED, ON THE AFFECTED SIDE.

ALL BRANCHES INVOLVED

THIS PATIENT PRESENTS WITH PAIN INVOLVING ALL DIVISIONS OF THE TRIGEMINAL NERVE (V1, V2, V3).



DIAGNOSIS:

TRIGEMINAL NEURALGIA

IMAGING DISCUSSION

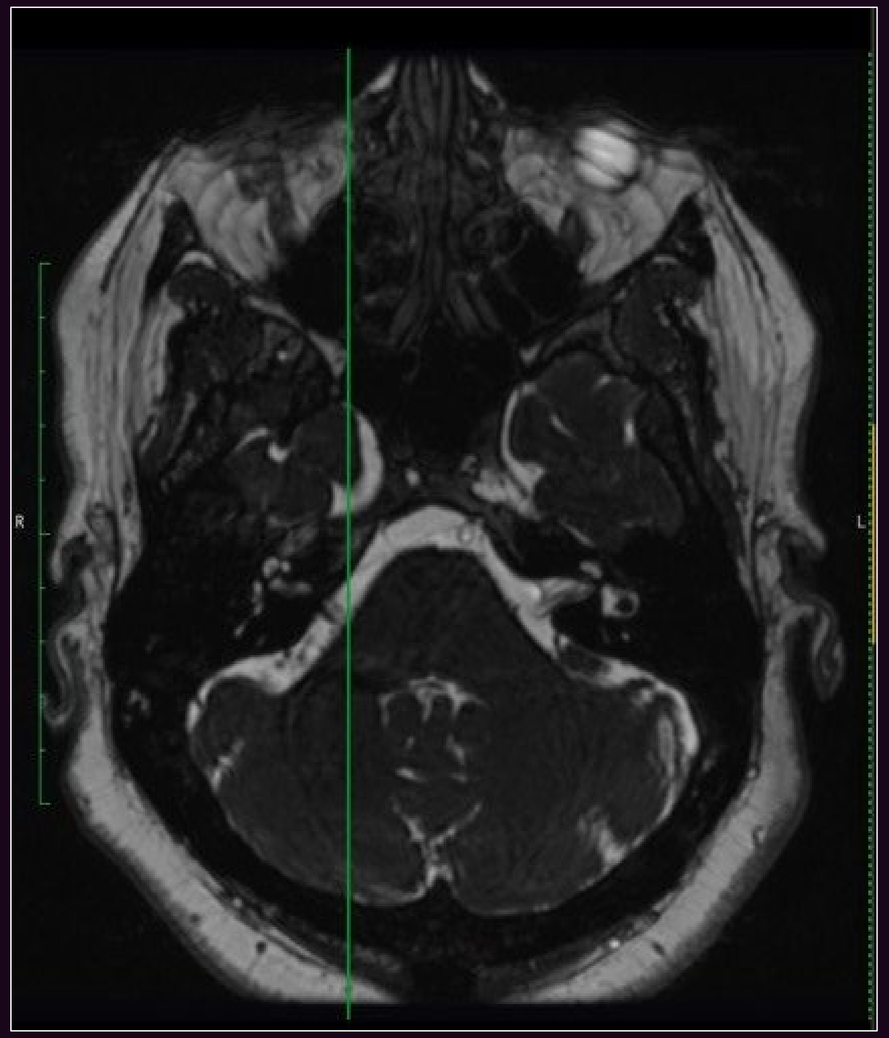


Fig 1. High resolution 3D FIESTA.

01 MRI IS MANDATORY IN ALL CASES OF TN TO ASSESS NEUROVASCULAR CONTACT AND EXCLUDE SECONDARY CAUSES

02 DEDICATED MULTI- SEQUENCE PROTOCOL AT 3.0 T

- 3D HEAVILY T2 - WEIGHTED CISTERNOGRAPHY (CISS / FIESTA / DRIVE)
- 3D TOF- MRA
- PRE/POST- GADOLINIUM 3D T1
- THIN SLICES (≤1 MM); NEURORADIOLOGIST BLINDED TO SYMPTOMATIC SIDE

IMAGING: POSITIVE FINDINGS

COMPLETE ABSENCE OF THE IPSILATERAL MECKEL CAVE

- NO CSF- CONTAINING DURAL RECESS
- NO PORUS TRIGEMINUS

MARKED ATROPHY OF THE CISTERNAL SEGMENT OF THE IPSILATERAL TRIGEMINAL NERVE

NORMAL CONTRALATERAL CAVE

NERVE ATROPHY CORROBORATES THE SYMPTOMATIC SIDE

THE AFFECTED TRIGEMINAL NERVE IS CONSISTENTLY SMALLER IN VOLUME/LENGTH THAN THE CONTRALATERAL NERVE, A VALIDATED MRI MARKER IN TN

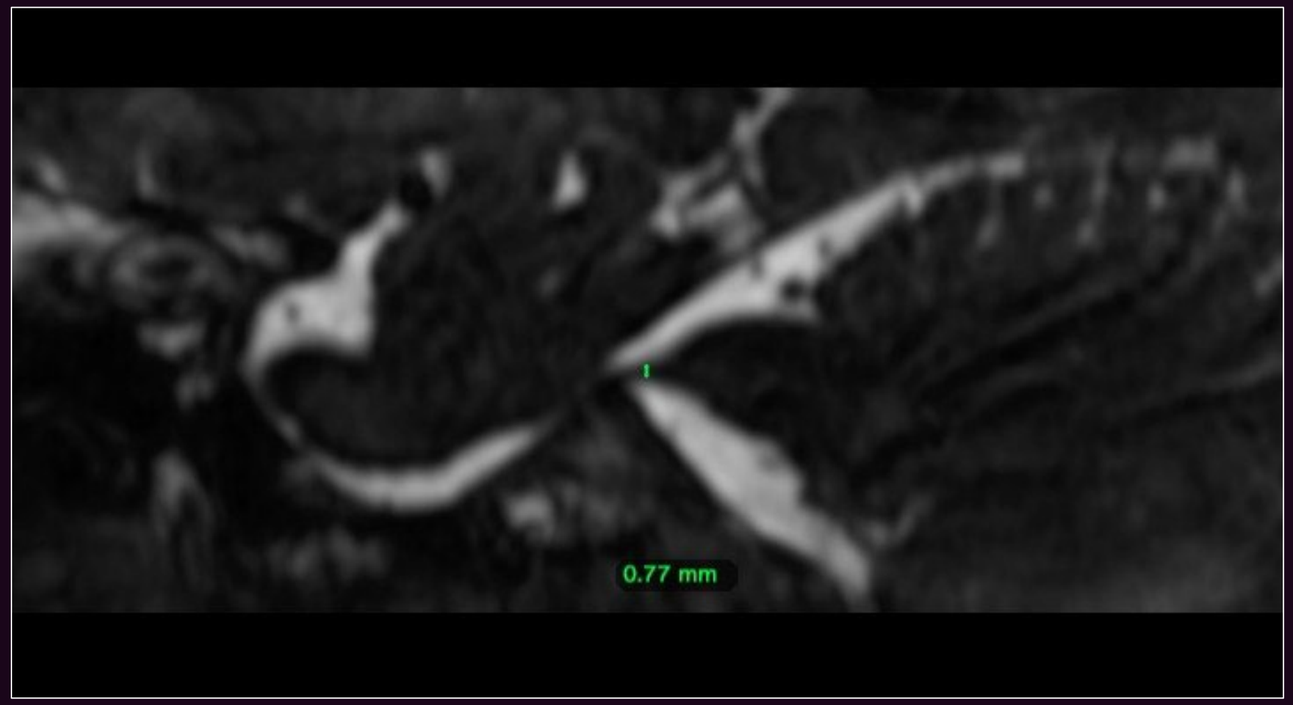


Fig 2. High resolution 3D FIESTA demonstrates complete absence of the right Mecke's cave with atrophy of the cisternal segment of the right trigeminal nerve, measuring 0.77 mm, marked decreased in size .

IMAGING: NEGATIVE FINDINGS

CRITICAL NEGATIVES TO DOCUMENT

NO NEUROVASCULAR COMPRESSION AT THE ROOT ENTRY ZONE, NO MASS/VASCULAR MALFORMATION, NO DEMYELINATING PLAQUE, AND NO ABNORMAL POST-CONTRAST ENHANCEMENT.

IMAGING IS SUPPORTIVE, NOT DIAGNOSTIC

MUST BE INTERPRETED WITH THE SITE OF SYMPTOMS ABSENT CAVE ANATOMY RECLASSIFIES THE CASE AWAY FROM IDIOPATHIC AND AWAY FROM AN MVD TARGET

MECKEL CAVE NORMALLY SHOWS MORPHOLOGIC ASYMMETRY

TRUE ABSENCE MUST BE DISTINGUISHED FROM NORMAL VARIANT HYPOPLASIA ON THIN - SECTION T2 UNDERSCORING THE VALUE OF HIGH - RESOLUTION CISTERNOGRAPHY.

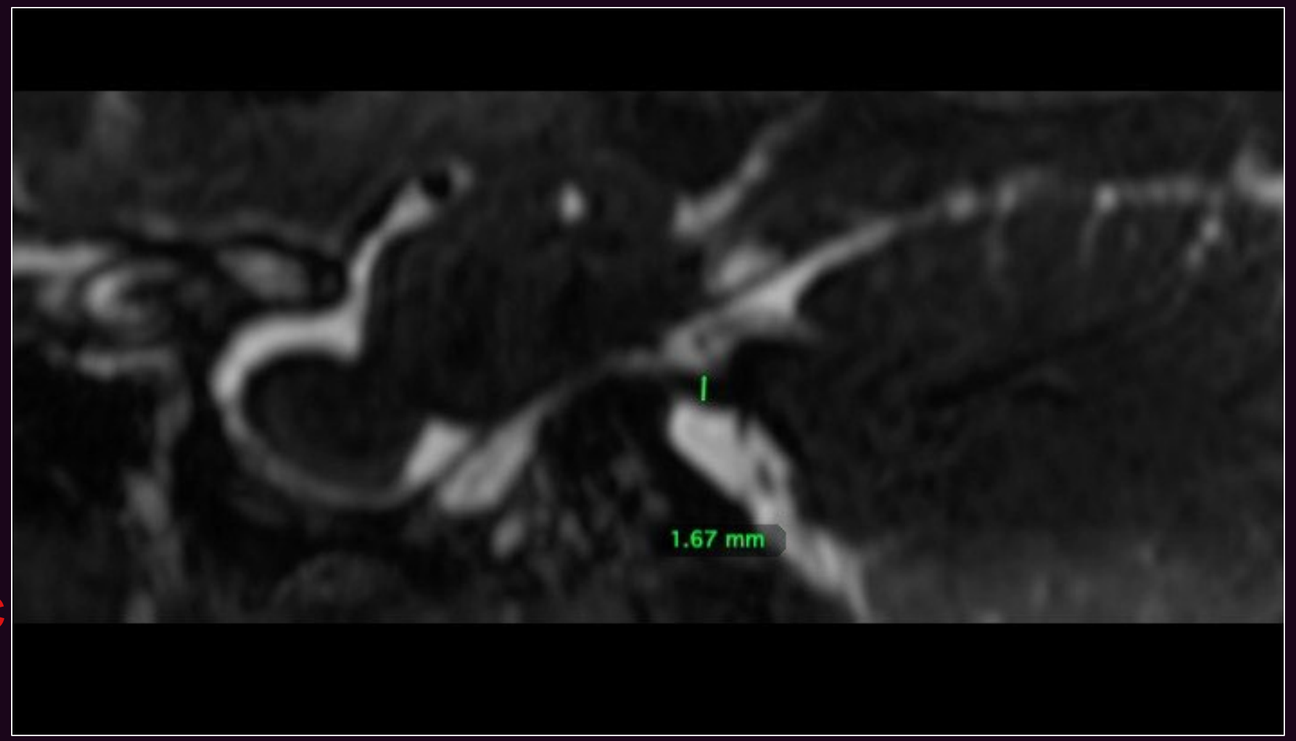


Fig 3. High resolution 3D FIESTA shows normal left Meckel's cave with normal left cisternal segment of the left trigeminal nerve, measuring 1.67 mm.

FIRST LINE MANAGEMENT

FIRST-LINE MANAGEMENTS PHARMACOLOGIC INVOLVING ≥2 TRIAL DRUGS BEFORE SURGERY.

FIRST LINE MEDICATIONS

- CARBAMAZEPINE 200–1200 MG/DAY
- OXCARBAZEPINE 300–1800 MG/DAY

SECOND LINE MEDICATIONS

- LAMOTRIGINE, GABAPENTIN, PREGABALIN, BACLOFEN, BOTULINUM TOXIN TYPE A, PHENYTOIN

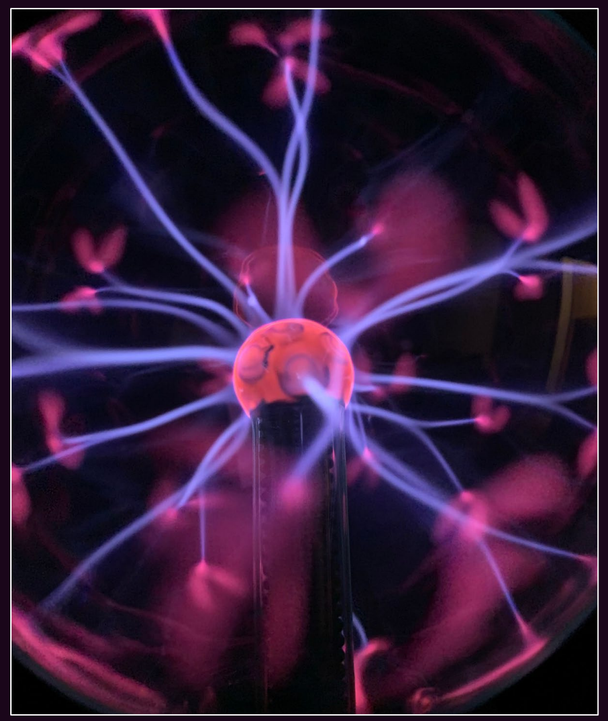
ALTERNATIVE MANAGEMENT

MICROVASCULAR DECOMPRESSION DOES NOT APPLY. AN ABSENT MECKEL CAVE MEANS NO NEUROVASCULAR CONFLICT TO DECOMPRESS.

SURGICAL ROUTE IS ABLATIVE OR RADIOSURGICAL.

BALLOON COMPRESSION	LOWEST CORNEAL RISK (<1%)
RADIOFREQUENCY THERMOCOAGULATION	CAN SPARE V1
GAMMA KNIFE RADIOSURGERY	NON-INVASIVE, DELAYED ONSET
GLYCEROL RHIZOLYSIS	-

CASE SPECIFIC MANAGEMENT



01 AVOID PROCEDURES THAT WORSEN CORNEAL SENSATION. FAVOR DIVISION-SPARING RADIOFREQUENCY OR BALLOON COMPRESSION.

02 IMAGING DRIVES THERAPY: THIN-SECTION 3D T2 (CISS/FIESTA) + TOF-MRA + PRE/POST-GADOLINIUM 3D T1 AT 3.0 T TO CONFIRM ABSENT CAVE AND EXCLUDE VASCULAR / MASS / DEMYELINATING LESIONS.

03 DOCUMENT THE CORNEAL REFLEX AFTER ANY PROCEDURE

OUTCOMES

MINIMAL ENTITY-SPECIFIC DATA

ONLY TWO CASE SERIES (7 + 3 PATIENTS). HOWEVER, ONLY WITH DIRECT EVIDENCE (3 PATIENTTS), WHERE ALL CHOSE GAMMA KNIFE OVER DECOMPRESSION, ALL REPORTED PAIN IMPROVEMENT

EXTRAPOLATED PAIN-FREE RATES (CLASSICAL/IDIOPATHIC TN)

- MEDICATION: ~90% INITIAL CONTROL; ~23% WITHDRAW FOR SIDE EFFECTS
- BALLOON COMPRESSION: 68% AT 4–11 YR
- RADIOFREQUENCY: 58% AT 3–9.3 YR
- GAMMA KNIFE: 32–63% LONG-TERM
- GLYCEROL RHIZOLYSIS: 28% AT 4.5–8 YR

COMPLICATIONS AND PROGNOSIS

- FACIAL HYPESTHESIA, CORNEAL HYPESTHESIA (6–10%), MASTICATORY WEAKNESS, KERATITIS.
- RELIEF ACHIEVABLE, BUT RECURRENCE COMMON; LONG-TERM RISK UNKNOWN



TAKE HOME POINTS

1

ABSENT MECKEL CAVE IS A RARE BUT RECOGNIZABLE STRUCTURAL CAUSE OF TN. THE ONLY SUBSTANTIAL SERIES, SHOWCASES ALL PATIENTS HAD IPSILATERAL TN AND IPSILATERAL NERVE ATROPHY.

2

SUSPECT IT WITH RED FLAGS SUCH AS FIXED HYPESTHESIA, DEPRESSED CORNEAL REFLEX, OR V1–V3 INVOLVEMENT WITHOUT NEUROVASCULAR CONFLICT.

3

DEDICATED HIGH-RESOLUTION MRI IS DIAGNOSIS-DEFINING — 3D T2 CISTERNOGRAPHY + TOF-MRA + PRE/POST-GADOLINIUM 3D T1

4

CORRECT CLASSIFICATION CHANGES SURGICAL APPROACH NO NEUROVASCULAR TARGET → MVD DOES NOT APPLY AND ABLATIVE/ RADIOSURGICAL APPROACH IS PREFFERED AFTER FAILED PHARMACEUTICAL MANAGEMNT

TAKE HOME POINTS

5

MEDICATION REMAINS FIRST-LINE — CARBAMAZEPINE OR OXCARBAZEPINE; TRIAL ≥2 AGENTS FIRST

6

INDIVIDUALIZE FOR V1 / CORNEAL INVOLVEMENT — lowest-corneal-risk techniques; document corneal reflex after intervention

7

OUTCOME DATA ARE MINIMAL — REPORT AND FOLLOW THESE CASES TO BUILD THE EVIDENCE BASE