

Trigeminal neuralgia associated with Chiari 1 malformation: symptom resolution following craniocervical decompression and duroplasty: Case report and review of the literature

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Abstract

Background: Trigeminal neuralgia (TN) may rarely be the presenting or only symptom of Chiari 1 malformation (CM). Isolated case reports have described resolution of TN following craniocervical decompression where TN is present in association with CM.

Case Report: This report discusses an unusual case of pure TN associated with CM that was successfully treated with craniocervical decompression and duroplasty and reviews the limited literature on the subject.

Conclusion: TN may be the sole presenting symptom of CM and can be successfully managed with craniocervical decompression. Clinicians should be aware of the association of TN with CM and consider surgical management.

Key Words: Chiari malformation, craniocervical decompression, duroplasty, trigeminal neuralgia

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INTRODUCTION

Trigeminal neuralgia (TN) may rarely be the presenting or only symptom of Chiari I malformation (CM). Isolated case reports have described resolution of TN following craniocervical decompression where TN is present in association with CM. This report discusses an unusual case of this nature that was successfully treated with craniocervical decompression and duroplasty and reviews the limited literature on the subject.

CASE REPORT

A 39-year-old female presented with severe right sided facial pain of sudden onset. Her pain was so severe that

she was admitted to a rural hospital for pain management. Her facial pain did not respond to oral medications including high dose carbamazepine, pregabalin, nonsteroidal antiinflammatory drugs (NSAIDs) and opiates. The patient was transferred to a tertiary hospital and investigation with magnetic resonance imaging (MRI) demonstrated a CM with tonsillar descent to the C1 level and very mildly increased T2 signal in the upper cervical cord [Figure 1]. There was no evidence of vascular conflict.

The patient proceeded to have a craniocervical decompression and duroplasty 8 weeks after first presentation of symptoms. At surgery tight arachnoid bands constricting the cerebellar tonsils were noted and the posterior aspect of the medulla

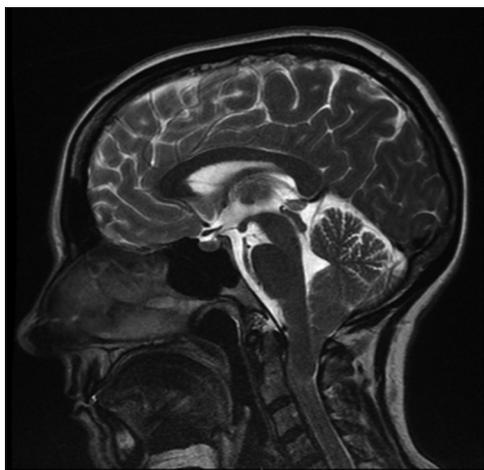


Figure 1: Preoperative T2WI MRI Brain demonstrating Chiari I malformation with subtle upper cervical cord signal change. The patient had severe and refractory right sided trigeminal neuralgia as the sole presenting symptom

had a gliotic and tented appearance consistent with chronic compression. Division of the arachnoid bands was performed and diathermy of the cerebellar tonsils without subpial aspiration. Native pericranium was used for duroplasty. There were no surgical complications. Following surgery the patient had immediate relief of facial pain and remains asymptomatic 1 year postsurgery with a satisfactory MRI appearance [Figure 2].

CONCLUSIONS

TN associated with CM is limited to a very small number of case reports. A 2008 case report with a literature review found only 19 cases in the English language literature,^[8] although in correspondence following this review several authors describe additional unpublished cases. A small number of cases in the non-English language literature can also be found.^[1,4,7,14] Pure TN is particularly uncommon as a presentation of CM and is limited to isolated case reports.^[2,7,9,10,13] Presentations of TN with CM may include bilateral symptoms or be secondary to hydrocephalus.^[5,11,12,14]

Postulated mechanisms of generation of TN due to Chiari malformation include (i) vascular compression at the nerve root entry zone, which could be affected by hydrocephalus or anatomic factors related to the Chiari malformation, such as a small posterior fossa; (ii) demyelination; (iii) microischaemic changes; and (iv) direct brainstem compression.^[8] The spinal tract of the trigeminal nucleus has been implicated due to its dorsal location and poor myelination which potentially renders it vulnerable.^[6,12]

Case reports describe a range of successful treatments in cases of TN associated with Chiari malformation including medical treatment with carbamazepine^[7] and craniocervical

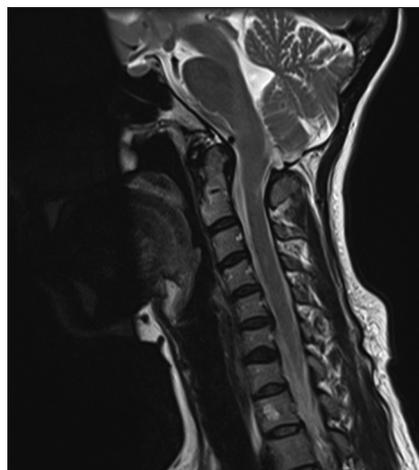


Figure 2:T2WI MRI scan one year postcraniocervical decompression and duroplasty. The patient remains asymptomatic

decompression^[3,10,12] as with this case. A bilateral TN case with associated CM was successfully treated with bilateral microvascular decompression (MVD),^[15] however, it is argued by some authors that retrosigmoid craniotomy for MVD may improve symptoms by indirectly decompressing the foramen magnum.^[6] Cases with associated hydrocephalus have been treated successfully with endoscopic third ventriculostomy^[11] and ventricular shunt procedures.^[5] Than *et al.*, in 2011,^[12] reviewed treatments for the 20 known cases described in the English literature at that time and found 15 of the 20 patients had been treated with craniocervical decompression with a reported 73% resolution of pain symptoms.

This case report contributes to the very limited number of case reports of Chiari malformation with pure TN, and those successfully treated with craniocervical decompression and duroplasty. Neurologists and neurosurgeons should consider this diagnosis in presentations of TN with concurrent CM and consider surgical treatment.

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