

Case Report

Familial Trigeminal Neuralgia – Report of Two Cases

Yousef Al-Awadi, Kostadin L Karagiozov
Department of Neurosurgery, Ibn Sina Hospital, Kuwait

Kuwait Medical Journal 2003, 36 (2): 128-130

ABSTRACT

Familial trigeminal neuralgia is a rare condition with only 125 cases described until now. Two sisters with trigeminal neuralgia are presented, focusing on the clinical details with the surgical findings, and their relationship to the characteristics of the familial neuralgia group. The surgical

findings in both cases are discussed. The scope of the current theories for the pathology responsible for the condition, the differences between familial and the general trigeminal neuralgia group and the future possibilities for clarifying the cause of these differences are also highlighted.

KEY WORDS: familial condition, tic douloureux, trigeminal neuralgia

INTRODUCTION

There are very few reports in the literature on the familial occurrence of trigeminal neuralgia and its problems^[1-5]. The two major general series of surgically treated trigeminal neuralgia patients were those of Harris with 1433 cases^[6], and Jannetta with 2420 cases^[7]. The latter had detected statistical incidence of familial cases, but Harris reported it in about 1.2% of the cases only. Single families have also been reported on several occasions^[7,8]. With the exception of the association of familial incidence to Charcot-Marie-Tooth polyneuritis on two occasions only^[9,10], the majority of the cases did not reveal common causes or factors related to the clustered incidence. All these observations have led to the preliminary conclusion that the occurrence is either sporadic or related to an unknown factor. With the existing uncertainty on the nature of this occurrence, the issue on the best choice of treatment methods and the expectations for outcome remains open.

CLINICAL CASES

We have treated in the last nine years, two patients with trigeminal neuralgia. The clinical similarity of the two sisters was unusual and demanded a more detailed description. Data for only one generation before them were available and no other diseases except for hypertension and diabetes type II were discovered in their relatives. One of their sisters died of cancer (unknown type of malignancy) more than 10 years before the onset of trigeminal pain in both patients.

Case 1

Patient S.M., is a 70-year-old female who had right facial pain since 1997. The pain was episodic,

on short attacks of excruciating severity described as electric shocks. The sensory zones of V2 and V3 were affected from the very beginning, but at a later stage pain radiated to the V1 zone too. Minor stimuli on the affected areas as well as chewing, swallowing and dental care became intolerably painful. Being disabled by the pain, the patient underwent preoperative clinical evaluation. Her general and neurological examinations were within normal. Her routine laboratory studies of hematology and biochemistry were within the normal ranges too. EMG of the facial and trigeminal nerves was normal. CT and MRI scans of the brain did not show any abnormalities. Having taken carbamazepine for years, up to 1200 mg per day, the plasma levels were steadily in the upper therapeutic range and at times beyond the upper limits. The patient was offered a microvascular decompression and it was performed on the 28th of October 1998. After a small right upper lateral suboccipital craniectomy (Fig. 1), the upper cerebellar surface was retracted and the trigeminal cistern opened. The dissection of the trigeminal roots revealed a loop of the superior cerebellar artery in offending contact with the root "entry" zone, caudally (under) the roots. (Fig. 2) The offending loop was displaced and immobilized with Teflon felt strips and cotton balls. The post-operative period was unremarkable, and for the last three and a half years, she is pain-free.

Case 2

A 65-year-old female was having typical lancinating pain since early 1986 radiating to the V2 and V3 zones of the right trigeminal nerve. It was easily precipitated by wind, touch and meals. The most sensitive triggering area was the angle of the

Address correspondence to:

Kostadin L. Karagiozov, MD, PhD, PO Box 7801, Salmiya 22089, Kuwait. Phone/fax 571-1471. Email:kostadin@qualitynet.net

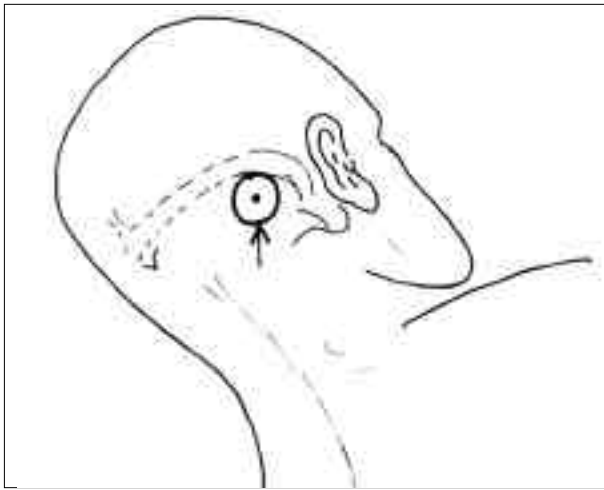


Fig. 1: Schematic representation of the site of craniotomy (pointed with an arrow), just under the turn of the transverse into the sigmoid sinus, usually round shaped.

mouth. Initially she responded to 400 mg of carbamazepine, however after two years, the doses had to be increased to 1600 mg with gradual appearance of toxicity signs. Unfortunately, she had extracted all the teeth on the right side at the advice of a dentist before the treatment. She was admitted in April 1992 and investigated for surgery. Her general and neurological condition was normal. Hematological and biochemical tests were also normal. Her CT and MRI did not show any lesion related to the condition. On 22 April 1992, she was operated. After a right superior lateral suboccipital craniectomy (Fig. 1) and retraction of the superior surface of the cerebellum, the trigeminal nerve within the arachnoid cistern was approached. It was found involved in arachnoid adhesions and compressed by a loop of the superior cerebellar artery (Fig. 3). The loop was dissected and displaced away from the nerve roots. Its position was maintained with loose Teflon felt looping strips and balls. Post-operatively there were no complications and control of pain was good.

ANALYSIS AND DISCUSSION

The average incidence of trigeminal neuralgia is 1/100,000. The coincidence of trigeminal pain within one family will therefore be of a very low probability. Because of the severity of complaints, such pain syndrome is not expected to be missed. Reviewing all published cases, currently the total number stands at 125, with the majority of families being two affected members^[11]. Cases of familial trigeminal neuralgia were published for the first time by Patrick in 1914 and since then, its clinical description has been gradually completed^[12]. There are several characteristic features of the familial cases:

1. The average age of onset is earlier compared to the general group and there is predominance of

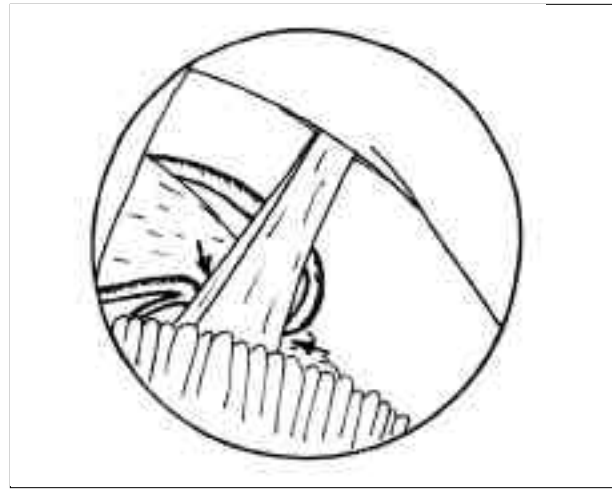


Fig. 2: Schematic representation of the surgical microscopic findings. The trigeminal root, exiting from the pons and the entering dura of Meckels cave is exposed after retraction of cerebellum. A tortuous artery loops under the trigeminal root and the arrows indicate the offending areas of the trigeminal "root entry" zone.

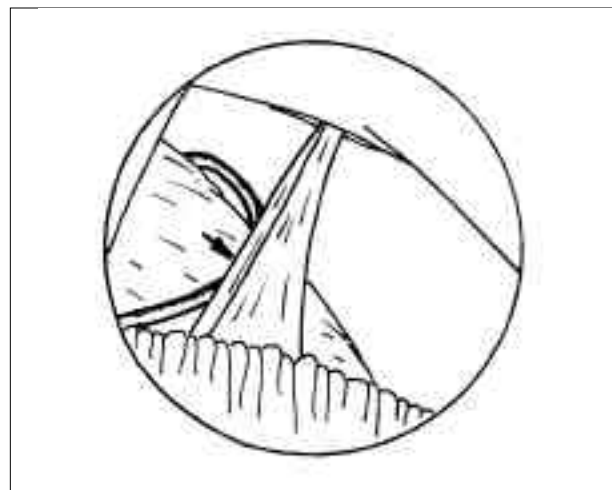


Fig. 3: Schematic representation of the surgical microscopic findings. The structures shown are the same as in Fig 2, but the 'offending' area of the 'root entry' zone is on the medial side of the nerve.

females 1.6/1.

2. The right side is more frequently affected and bilateral cases are more common^[13].
3. The maxillary branch is the one most frequently affected, a finding different from the general group of operated trigeminal neuralgia patients.

All these clinical differences make the familial group clearly distinctive. Except for the age of onset and bilateral affection, our patients match these clinical characteristics. The responsible underlying pathology of the trigeminal nerve is not certain. The reasons are:

1. Only small proportion of the cases have verified and analyzed pathological findings, and they were operated by microvascular decompression in the last decades.
2. Part of the cases remain with unproved pathological findings because of the application of ablative functional methods such as the

percutaneous technique (percutaneous glycerol, balloon compression or radiofrequency thermocoagulation), away from the site of presumed trigeminal pathology.

These two factors impede the accumulation and analysis of data. In these circumstances the two existing theories of early atherosclerosis and associated tortuosity of vessels and secondly the existence of biochemical changes in myelin, are unproved at present. In the future we expect, that thorough reporting and cumulative analysis of cases will provide significant pathological findings for the familial trigeminal neuralgia. A wider application of the microvascular decompression as a method of treatment for familial trigeminal neuralgia is anticipated.

REFERENCES

1. DiCorato MP, Pierce BA. Familial Trigeminal Neuralgia South Med J 1985; 78:353-354.
2. Duff JM, Spinner RJ, Lindor NM, *et al.* Familial trigeminal neuralgia and contralateral hemifacial spasm Neurology, 1999; 53:216-218.
3. Herzberg L. Familial trigeminal neuralgia. Arch Neurol 1980; 37:285-286.
4. Kirkpatrick DB. Familial trigeminal neuralgia: case report. Neurosurgery 1989; 24:758-761.
5. Knuckey NW, Gubbay SS. Familial trigeminal and glossopharyngeal neuralgia. Clin Exper Neurol 1979; 16:315-319.
6. Harris, W. An analysis of 1433 cases of paroxysmal trigeminal neuralgia (tic-douloureux) and the end-results of gasserian alcohol injection. Brain 1940; 63:209-214.
7. Bracale C, Graziussi G, Avella F. Familial essential trigeminal neuralgia. Riv Neurobiol 1982; 28:299-302.
8. Braga FM, Bonatelli AD, Suriano I, Canteras M. Familial trigeminal neuralgia. Surg Neurol 1986; 26:405-408.
9. Testa D, Milanese C, La Mantia L, Mastrangelo M, Crenna P, Negri S. Familial trigeminal neuralgia in Charcot-Marie-Tooth disease. J Neurol 1981; 225:283-287.
10. Coffey RJ, Fromm GH. Familial trigeminal neuralgia and Charcot-Marie-Tooth neuropathy. Report of two families and review. Surg Neurol 1991; 35:49-53.
11. Fleetwood, IG, Innes AM, Hansen SR, Steinberg GK. Familial trigeminal neuralgia. Case report and review of the literature. J Neurosurg 2001; 95:513-517.
12. Patrick HT. The symptomatology of trifacial neuralgia. JAMA 1914; 62:1519-1525.
13. Pollack IF, Jannetta PJ, Bissonette DJ. Bilateral trigeminal neuralgia: a 14-year experience with microvascular decompression. J Neurosurg 1988; 68:559-565.