

## Encephalotrigeminal syndrome and the dentomaxillary apparatus - a case report

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### Summary

**Purpose.** This study is a case report regarding the correspondence between dentomaxillary lesions and a neuraxial dysfunction with genetic determinism - the encephalotrigeminal syndrome.

**Material and method.** K.V., a female of 29 years old reported to the Institute of Cerebro-Vascular Diseases "Vlad Voiculescu" - Bucharest for persistent headaches and asked for further investigations.

**Results.** The clinical and paraclinical examinations revealed the presence of the Sturge-Weber syndrome - and the correlated disorders in oral and maxillofacial territory.

**Conclusion.** Genetic or degenerative diseases may affect the dentomaxillary apparatus. Few data is available on these cases in specialty literature. In dental practice, the diagnosis of such oral dysfunctions requires special therapeutical approach.

**Key words:** dentomaxillary apparatus, Sturge-Weber syndrome, oral and maxillofacial dysfunctions, diagnosis and therapeutical approach.

### Introduction

Dentomaxillary lesions have multiple and variable causes, implying local and general factors. During embryogenesis and later on, during the growth of the dentomaxillary apparatus, there are certain genetic affections that induce functional and morphological disorders at this level.

Among the affections that could influence it, there are quoted growth disorders of the central nervous system such as encephalotrigeminal syndrome from which Sturge-Weber disease makes part of [1]. This disease consists of capillary and cavernous hemangiomas, localized in the cuta-

neous territory of the trigeminal nerve. Sometimes, most of the hemangiomas are venous and spread up to the subadjacent leptomeninges. On a cranial radiography, calcium deposits could be observed in the affected brain. Arterio-venous disorders often reach the facial region and induce functional and morphological alterations of the dentomaxillary apparatus. [2]

Brain and tegumentary lesions may appear separately. This disease is very rare. Most of the patients survive a long period of time, most often with mental disorders and hemiparesis.

The study is a case report that reveals the correlation between a genetic neuraxial

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dysfunction and growth disorders of the dentomaxillary apparatus.

### Case report

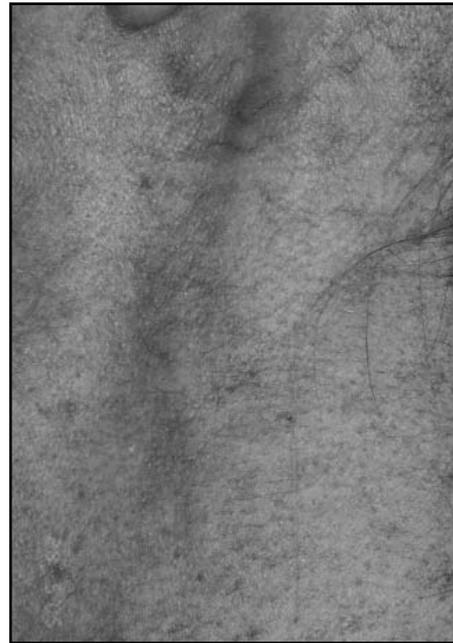
K.V., female of 29 years old, reported to the Institute of Cerebro-Vascular Diseases "Vlad Voiculescu", Bucharest, for persistent headaches and requested investigation. She was institutionalized with the registration number 5607/2005. A neurological clinical and paraclinical examination was performed and the patient was diagnosed with:

- migrainous syndrome;
- phakomatosis;
- arterio-venous malformation localized on the left side of the face and the left laterocervical region;
- elements of genodermatosis;
- left hemifacial congenital hypotrophy;
- right cortico-subcortical temporal congenital cyst;
- psychogenetic syndrome.

This case report intended to reveal the outstanding features of neuraxial pathology with genetic determinism and lesions of the dentomaxillary apparatus.

The orofacial inspection revealed the following lesions:

- cutaneous lesions associated with depigmentation in the left cervical and superior toracal regions;
- very important vascular lesion, that include the venous system in the left superior toracal and cervical regions. The superior vena cava and jugular vein have abnormal anastomosis (*Figure 1*). In a declive position of the head, cyanosis can be observed. In a normal position these venous eminences are no longer visible, revealing the variations of the venous pressure in the superficial veins of the head. Usually, patients with Sturge-Weber



**Figure 1.** Cutaneous angioma on the left lateral cervical region

syndrome have cutaneous angioma; in this case, the angiography shows that alterations affect the profound venous system [3].

- trophic disorders of the left hemiface. Muscular atrophy could be observed (the masseter and the platysma muscles (*Figure 2*)). Consequently, an obvious facial asymmetry could be noticed, affecting esthetics.

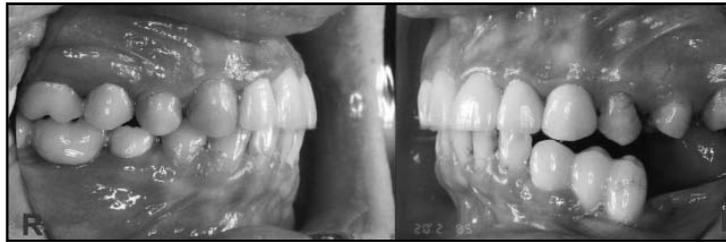


**Figure 2.** Muscular atrophy of the masseter and platysma muscles

- the oral examination reveals correct and incorrect treated carious lesions, bilateral singular superior edentations (*Figure 3*) with incorrect prosthetic



**Figure 3.** Occlusal view of the dental arches



**Figure 4.** Buccal view of the left and right dental arches. Incorrect prosthetic restorations in the right lateral and frontal region and an incorrect restoration of a mandibular lateral edentation on the left side (33 and 34 as abutment teeth)



**Figure 5.** Panoramic X-Ray of the patient

restorations in the right lateral and frontal region and an incorrect restoration of a mandibular lateral edentation on the left side. In the 3rd quadrant there is a fixed partial denture with the canine and the first premolar (33 and 34) as abutment teeth (semiincluded) that does not observe the correct occlusal plane. On the same side, a terminal edentation (not yet restored) is present (*Figure 4*).

The panoramic X-ray (*Figure 5*) certifies on the left mandibular hemiarch the presence of the first premolar (34) semiincluded and also of the second premolar and the molars (35,36,37,38) included in ectopic positions and incompletely developed. The retroalveolar radiographs show details of these teeth. The cephalometric X-Ray confirms the diagnosed anomalies (*Figure 6*).

The clinical examination confirms the affection of: integument, vessels, trophic and dentoperiodontal structures.

The electromyography [4] reveals the neurogenic denervation in the region with muscular atrophy.



**Figure 6.** Cephalometric X-ray

## Discussion

The 29 years old patient presents a series of malformations that correlate with the affection of embryonary and neuraxial structures. Usually in such cases there is a genetic determinism. For the present case, it was not possible to make a proper family anamnesis. The patient is still kept under observation with possible corrections of the dentomaxillary anomalies.

The electromyographic test suggests a neuraxial affection with secondary dentope-

riodontal lesions. The main reason is a deficiency in neuraxial trophic factors in the cephalic extremity.

## Conclusion

Genetic or degenerative diseases may affect the dentomaxillary apparatus. The dental practician can apply the proper treatment for the dentoperiodontal lesions, but the evolution of the disease requires continuous observation and further therapeutical approaches.

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