

# Clinical Presentation of Oral Manifestations and Intraoral Somatosensory Changes in Fahr's Disease

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## Abstract

Fahr's disease is a rare congenital disorder characterized by abnormal calcium deposition with subsequent atrophy involving the basal ganglia, cerebral and cerebellar cortical regions. Very little information is available regarding non-neurological manifestations of this disease, and almost no information is available on oral findings. Moreover, information is unavailable regarding intraoral somatosensory changes in Fahr's disease. We report oral findings and intraoral somatosensory changes in patient with Fahr's disease. A 62-year-old Japanese woman was referred to the Department of Oral and Maxillofacial Surgery at Tokyo Women's Medical University Hospital with symptoms of bleeding gums. The patient had been diagnosed with Fahr's disease by the Department of Psychiatry 1 year earlier. Intraoral examination showed poor oral and dental hygiene with gingival hyperplasia on the buccal aspects of the upper and lower incisors. Generalized attrition of the teeth was seen. Panoramic radiography showed horizontal bone resorption, but no evidence of congenital absence of any teeth. Gingival bleeding associated with poor periodontal condition was diagnosed. As an additional symptom, the patient reported intraoral burning sensation of the tongue. Qualitative sensory testing was performed for the tongue, upper and lower gingiva and lip mucosa, showing heat and cold hyperalgesia of the tongue. Mechanical allodynia located in the upper and lower lip mucosa was also reported. In accordance with these clinical findings, we diagnosed primary and/or secondary burning mouth syndrome related to Fahr's disease. This case represents only the second instance for which intraoral findings of Fahr's disease have been reported. Somatosensory changes were also found to be associated with the present case of Fahr's disease. Intraoral somatosensory changes related to Fahr's disease may be due to progressive lesions associated with this disease. Continuous follow-up and qualitative sensory testing to assess disease progression may be needed for clarification of these issues.

**Keywords:** Fahr's disease; Intraoral; Sensory test; Burning mouth syndrome; Bruxism; Somatosensory change; Pain; Hyperalgesia; Allodynia.

## Introduction

Fahr's disease is a rare congenital disorder characterized by abnormal calcium deposition involving the basal ganglia, cerebral and cerebellar cortical areas and subsequent atrophy [1-3]. Very little information is available regarding the number of neurological manifestations of this disease and almost no information is available regarding oral findings. Considering the various metabolic abnormalities such as stunted physical and neuropsychological development in these patients, one could also expect abnormalities in the oral tissues. Moreover, no information is available about intraoral somatosensory changes in Fahr's disease. We therefore report oral findings and intraoral somatosensory changes in a patient with Fahr's disease.

## Case Report

A 62-year-old Japanese woman was referred to the Department of Oral and Maxillofacial Surgery at Tokyo Women's Medical University Hospital with symptoms of bleeding gums. The patient had been diagnosed with Fahr's disease by the Department of Psychiatry 1 year earlier. The patient was also on medication for diabetes, and had been previously prescribed prednisolone and bisphosphonate for lung cancer. Intraoral examination showed dental attrition, poor oral and dental hygiene with gingival hyperplasia on the buccal aspects of the upper and lower incisors (Figure 1). The patient reported

difficulty in tooth brushing due to tremors and muscle rigidity related to Parkinsonism. Panoramic radiography showed horizontal bone resorption, but no evidence of congenitally missing teeth. Gingival bleeding associated with poor periodontal condition was diagnosed in this case. Periodontal treatment involving oral prophylaxis and tooth brushing instructions were given and efforts were continuously made to maintain oral hygiene.

As additional symptoms, the patient reported an intraoral burning sensation of the tongue. After dental, bacteriological and radiographic examinations, possible causes of symptoms like infection by *Candida albicans* and other structural changes were ruled out, with the exception of calculus deposition and without symptoms associated with dental caries. Qualitative sensory testing using a spatula (put out from hot and cold water for thermal stimuli) and Q-tip (to investigate mechanical

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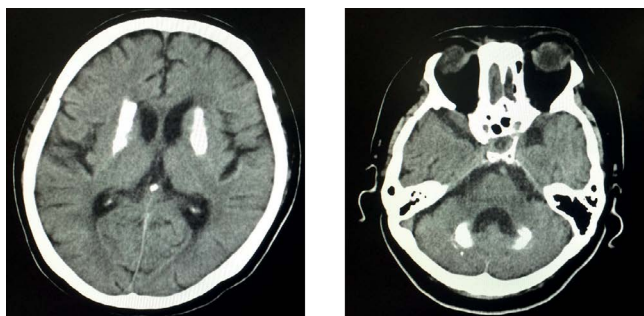
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**Figure 1:** Poor oral and dental hygiene, and poor periodontal condition with gingival hyperplasia on the buccal aspects of the upper and lower incisors. Generalized attrition of the teeth is evident.



**Figure 2:** Intracranial CT showing bilateral calcification of the basal ganglia and dentate nucleus. No evidence of tumor or nerve dislocation is apparent.

allodynia) was performed on the tongue, upper and lower gingiva and adjacent lip mucosa [4]. The results of this qualitative sensory testing showed heat and cold hyperalgesia of the tongue and mechanical allodynia of the upper and lower lip mucosa. Moreover, paradoxical heat sensation was observed on the upper gingiva. Bilateral calcification of the basal ganglia and cerebellar dentate nucleus were observed in intracranial computed tomography (CT) (Figure 2), but no evidence of tumor or nerve dislocation was seen. In accordance with these clinical findings, we diagnosed primary and/or secondary burning mouth syndrome (BMS) related to Fahr's disease.

## Discussion

Fahr's disease is characterized by abnormal deposition of calcium in areas of the brain that control movements including the basal ganglia, thalamus, dentate nucleus, cerebral cortex, cerebellum, subcortical white matter, and hippocampus [5]. Most cases initially present with extrapyramidal symptoms. They may also present with cerebellar dysfunction, speech difficulty, dementia and neuropsychiatric symptoms [6]. The diagnostic criteria for Fahr's disease have been modified and derived from those described by Moskowitz et al. [3,5,6] and present case was diagnosed following the suggested diagnostic criteria.

- Bilateral calcification of the basal ganglia visualized on

neuroimaging. Calcification in other brain regions can also be observed.

- Progressive neurological dysfunction, which generally includes movement disorder and/or neuropsychiatric manifestations. Age of onset is typically in the fourth or fifth decade, although this dysfunction may also present in childhood.
- Absence of biochemical abnormalities and somatic features suggestive of a mitochondrial or metabolic disease or other systemic disorders.
- Family history consistent with autosomal-dominant inheritance.

Intraoral characteristic features of Fahr's disease have been reported in which several missing teeth were evident, along with tooth mobility, generated gingival inflammation and recession with associated periodontitis [7]. In the present case, gingival inflammation and bleeding were observed; however, no absence of teeth was evident. A previous case report has reported radiographic findings showing partial anodontia, as well as partial development of the maxillary and mandibular regions [7]. Generalized rarefaction of bone, resembling that seen in hyperparathyroidism, was also found. In this case, horizontal bone resorption was observed without any other tooth or bone abnormalities. The poor dental hygiene in this case may be explicable by Parkinsonism (motor impairment, apathy, depression, and dementia) [8]. There is no evidence of bruxism and clenching in patients with Fahr's disease. However, an earlier study showed an association of bruxism and clenching with motor symptoms of Parkinson's disease [9]. The dental attrition in the present case could thus have been due to bruxism associated with Parkinsonism on this patient with Fahr's disease. Although, tooth attrition during oral examination was evident in the present patient there was no evidence of involuntary movement or mandibular dystonia during clinical examination.

Bruxism is defined as a repetitive jaw-muscle activity characterized by clenching or grinding of the teeth and/or by bracing or thrusting of the mandible [10]. Since some researchers have proposed alternatives to the so-called gold standard tool for diagnosis about sleep bruxism (e.g. polygraphic and audio-video recording) [11,12], it is necessary to measure masticatory muscle activities with polysomnography to diagnose about sleep bruxism. Further studies may be needed to investigate the correlation between sleep bruxism and Fahr's disease using polysomnography.

Moreover, somatosensory abnormalities were reported as non-motor symptoms of Parkinson's disease. In general, abnormalities in nociceptive and mechanical thresholds have been described in Parkinson's disease [13]. These have been attributed to anomalies of central nociceptive processing and sensorimotor integration through the affected basal ganglia and dopaminergic pathways [14].

In the present case, we identified heat and cold hyperalgesia and mechanical allodynia of the intraoral region. BMS was also diagnosed by taking into account the findings from intraoral qualitative sensory testing and other clinical findings. No previous reports have described abnormalities of sensory loss or gain, or burning sensation in the mouth in patients with Fahr's disease. In this case, because of the lack of evidence for peripheral organic changes in the oral mucosa, the sensory changes and burning mouth sensations may be best explained by anomalies of central nociceptive processing in the basal ganglia and/or dopaminergic pathways. We therefore think that an association may exist between these reported somatosensory changes and Fahr's disease.

To the best of our knowledge, this case represents only the second

instance for which intraoral findings of Fahr's disease have been reported. Somatosensory changes were also found to be associated with the present case of Fahr's disease. Intraoral somatosensory changes related to Fahr's disease may be due to progressive lesions associated with this disease. Continuous follow-up and qualitative sensory testing to assess disease progression may be needed for clarification of these issues.

#### Conflict of Interest

None of the authors have any conflicts of interests to declare.

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