Winsome Cheung 2019.11.19
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Neurofibromatosis (Von Recklinghausen's disease)

Neurofibromatosis type 1 (NF1), also known as Von Recklinghausen's disease, is a common autosomal dominant condition that affects the nervous system, the skin**,** the oral cavity, and bone. NF1 occurs in every 1 out of 2,500 births. It is a genetic disorder that has a high risk of both benign and malignant tumor formation. Although this disorder can be inherited, about 50% of NF1 patients have no family history of the disease. Individuals may have this disorder through gene mutations. Clinically, there are seven components of the disorder including six or more Café au lait patches, two or more neurofibromas, freckling in the axillary or inguinal area, optic gliomas, two or more Lish nodules, and bone lesions. Skin pigmentation is the most prominent clinical presentation out of all the component, and it tends to grow in size and quantity over time. These spots are either present at birth or developed during early childhood.

Despite the prominent extra oral clinical presentations, NF1 patients have some unique intraoral traits as well. Development of neurofibromas on the tongue, on or around peripheral nerves, palate, vestibule, and buccal mucosa. NF1 patients may have gingival enlargement and pigmentation, facial skeletal abnormalities, plexiform neurofibromas in oral soft tissue, perioral soft tissues with subsequent periodontitis, overgrowth of the alveolar bone, impacted and supernumerary teeth. There is no prevalence of tendency in sex or race for the disorder but it is typically diagnosed in early childhood. Biopsy can identify neurofibroma lesions. Radiographically, there is increased size of coronoid notch, condylar deformity, increased bone density and enlarged mandibular foramen.

Patients who have Neurofibromatosis may also have neurological complications that can lead to cognitive impairment, learning difficulties, and behavioral issues. They also have an increased risk of having high blood pressure, gastrointestinal symptoms, cerebrovascular disease, and epilepsy. Treatment for neurofibroma is surgical excision. Laser is used for small neurofibromas but is not effective if there are numerous tumors. Occupational therapy is helpful for coordination difficulties and attention problems. Differential diagnosis of Neurofibromatosis are neurofibromas. Cutaneous neurofibromas can resemble other benign cutaneous tumors such as melanocytic nevi, lipomas, and schwannomas. Cafe´-au-lait macules are associated with several other syndromes. Subcutaneous neurofibromas clinically resemble lipomas, which are much commonly seen than neurofibromas.

The prognosis of NF1 is unpredictable due to the possibility of reoccurrence and malignancy transformation. Early diagnosis of NF1 is important in order to ensure that patients and their families are counseled appropriately. Most patients with NF1 can be diagnosed by careful history taking and clinical examination. Genetic counselling is encouraged for parents who carries the gene of NF1. Diagnosis is based on a series of clinical presentations criteria. As a dental hygienist, it is essential to be able to recognize these criteria, so we may help to refer the patient for a timely diagnosis and treatment. Early diagnosis and intervention may improve an individual's quality of life.

Oral health status is often jeopardized in patients with neurofibromatosis type1. Therefore, NF1 patients should be closely monitored for disease complications. It is important to stress the importance of a follow up in patients with oral NF1 presentations because of the high rate of recurrence. Patient education is vital towards oral hygiene maintenance and regular dental check-ups must be emphasized to improve the oral health status in patients with NF-1.

Reference:

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