Dental Issues

The components of Sturge-Weber syndrome (SWS) have several consequences affecting oral health:

Increased Vascularity of the hard & soft tissues of the jaw on the involved side of the face. An otherwise simple extraction can become a major surgical management problem because of the very real danger of hemorrhage post-operatively. Flat bluish to red lesions that frequently blanch with pressure can be Intraoral extensions of facial port wine stains over the lips and onto contiguous cheek surfaces. Solitary vascular growths may also occur in these areas as well as on the palate and gums. Most oral angiomas are on the same side as the facial nevus and occur on the oral structures of the upper jaw. Uncommonly, mucosal lesions may be bilateral and/or involve the mandibular gingivae, floor of mouth or tongue. Tongue lesions have been described to resemble dilated blood vessels and to include enlargement of one side. Intraoral vascular lesions may range from small flat areas to very large proliferating masses of alarming proportions.

Swelling of the oral soft tissues is a fairly common manifestation of SWS. The problem becomes further complicated if anti-seizure medication is being taken. Dilantin in particular causes hyperplasia of the gingival. Hyperplasia is an increase in the size of the gums due to growth of new tissue. This tissue must be removed surgically, with the same concerns about post-surgical hemorrhage. Other oral findings associated with SWS include enlargement of one side of the jawbone containing the teeth (on the same side as the facial lesion) and a high arched palate, although one sided enlargement or diminished jaw size opposite to the facial lesion have been reported.

Dental findings may include multiple spaces between the teeth especially associated with alveolar hyperplasia. The increased vascularity of the bones of the jaw may also lead to premature eruption of permanent teeth, unusual eruption time and location of teeth, and periodontal problems such as pockets and loss of attachment of teeth to jaw, resulting in mobility and premature loss of teeth. Less commonly, both enlarged teeth and delayed tooth eruption on the same as the facial lesion may be seen. Microscopic examination of teeth related to oral vascular lesions has revealed angiomatous changes in the dental pulp.

Oral hygiene problems are frequently related to many of the gingival lesions in SWS. Patients with developmental delays of motor skills and/or cognitive skills may have difficulty with oral hygiene, thereby requiring support and care by others. Significant periodontal disease with loss of bony support for the teeth, especially related to areas of gingival masses or lesions and hyperplasia may occur. Gingival hyperplasia associated with anticonvulsant medication is well know to be related to oral hygiene, with meticulous oral and dental hygiene required to minimize gingival overgrowth.

It is very important, to accurately diagnose each patient with oral vascular lesions, especially with the similar Klippel-Trenaunay Syndrome (KT). Although SWS and KT gingival lesions may resemble each other, SWS lesions usually do not manifest with vascular involvement of underlying bone, while

KT lesions often include bony involvement with a significant risk of uncontrollable bony bleeding. There is one case report of "prominent vascularity" in the alveolar bone underlying a gingival lesion in SWS. Numerous reports of dental treatment of SWS patients including surgical treatment of vascular oral lesions indicate the need for caution and expertise but also report minimal bleeding without significant complications.

Oral Care and Treatment

Dental treatment requires care for the risk of bleeding. There are no reasons to avoid dental treatment in SWS, and individual modifications of techniques and equipment may be necessary such as in the use of rubber dam and clamps, to avoid localized trauma. In order to avoid accidental injury to the soft tissues of the mouth, the patient must be calm and cooperative through the entire visit. Resistant patient may have to be treated under sedation or general anesthesia so that the dentist can attain the necessary cooperation. Many medical centers and hospitals have Dental departments and all Provinces and States offer referral information for dentists with specialized training.

A specialist with appropriate training and experience should perform surgical treatment of oral lesions or dental extractions in the area of a lesion, because of the unique anatomy and challenging access. Many modalities of treatment can be successfully utilized for the surgical removal of intraoral vascular lesions, ranging from sclerosing (scarification) to conventional excision, freezing (cryosurgery) and laser excision. When properly performed, such procedures are safe and there is little risk of recurrence.

Anesthetic management of SWS requires careful pre-operative evaluation for associated anomalies. Vascular lesions of the nose, nasopharynx. lips, oral cavity larynx and trachea should be identified and tracheal intubation performed with soft, nonstylleted, well lubricated endotracheal tubes. Consideration of oral or nasal intubation is required for each individual and careful intubation and exctubation *is* mandatory. Ocular and neurological manifestations of SWS may require selection of anesthetic agents to avoid increases in intraocular and intracranial pressure. Safe anesthetic management requires a hospital facility.

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