

STING-Associated Vasculopathy with Onset in Infancy: A case report

Behbehani J., DDS; Loo C.Y., BDS, PhD, MPH, DMD, FAAPD; Laskou M. DDS, DMD, FAAPD (Tufts University School of Dental Medicine, Boston, MA)



Introduction

STING stands for Stimulator of Interferon Genes. STING-associated vasculopathy with onset in infancy (SAVI) is an autoinflammatory disease associated with enhanced type I interferon signaling. It is a rare disorder involving abnormal increased inflammation throughout the body especially in the skin, blood vessels, and lungs.

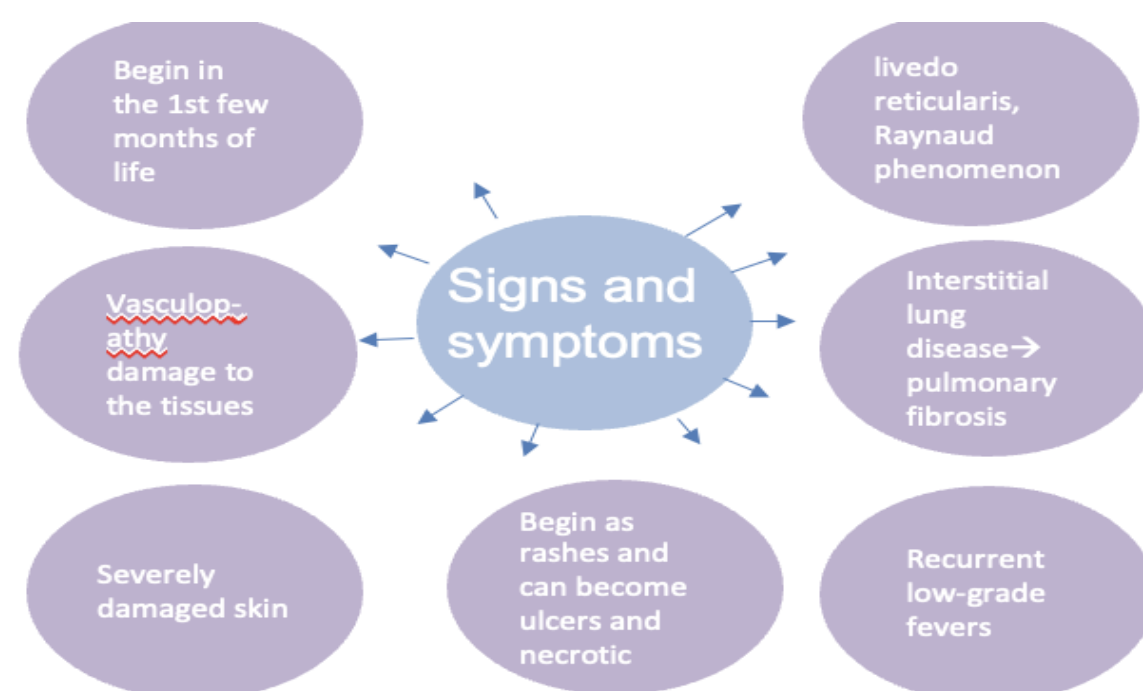
Prevalence: Unknown, only few affected individuals in the medical literature. 52 patients in 37 families reported. Verified by genetic sequencing

Inheritance: *de novo*, autosomal dominant

Etiology: Mutation in the STING1 gene, "gain of function" in TMEM173. STING produces beta-interferon, a cytokine that promotes inflammation, its overproduction leads to excessive inflammation and tissue damage.

Complications: Scarred ear, nasal septum perforation or finger/toes need amputation, childhood morbidity and increased mortality

Management: Minimally responsive to conventional immunosuppressive therapies, Janus Kinase inhibitors are promising treatment.



Case Report

A 7-year-old male with SAVI presented with an acquired saddle nose deformity, failure to thrive, midfacial hypoplasia, velopharyngeal insufficiency causing hypernasality, sleep related breathing disorder, and oligoarticular juvenile idiopathic arthritis.

Patient takes multiple immunosuppressive medications (adalimumab, hydroxychloroquine, azathioprine), antibiotics (Bactrim) calcium channel blocker (amlodipine), pain medication (acetaminophen) and is allergic to cephalosporins and all nuts.

Dental findings:

- Restricted maxillary arch, anterior crossbite
- Anterior alveolar bone loss and gingival recession
- Premolars are congenitally missing
- Dental crowding
- Caries on #K-MO, #L-DO

Management

1. Exam and Planning Phase:

- Medical clearance (Rheumatology)
- Preventative- periodic exams, prophylaxis, fluoride, sealants
- Extractions and stainless-steel crown
- Orthodontic consult- no treatment due to periodontal concerns and poor oral hygiene

2. Treatment Phase:

- OR full-mouth rehab with ENT
- Exam, prophylaxis, FMX
- Extractions: #E (excessively mobile), #L (non-restorable caries)
- Sealants on #19, #30, SSC on #K

3. Maintenance Phase:

- Three-month recalls: exams, caries risk assessment, anticipatory guidance, prophylaxis, fluoride



Fig.1 Facial front picture



Fig.2 Facial profile



Fig.2 Panoramic Radiograph

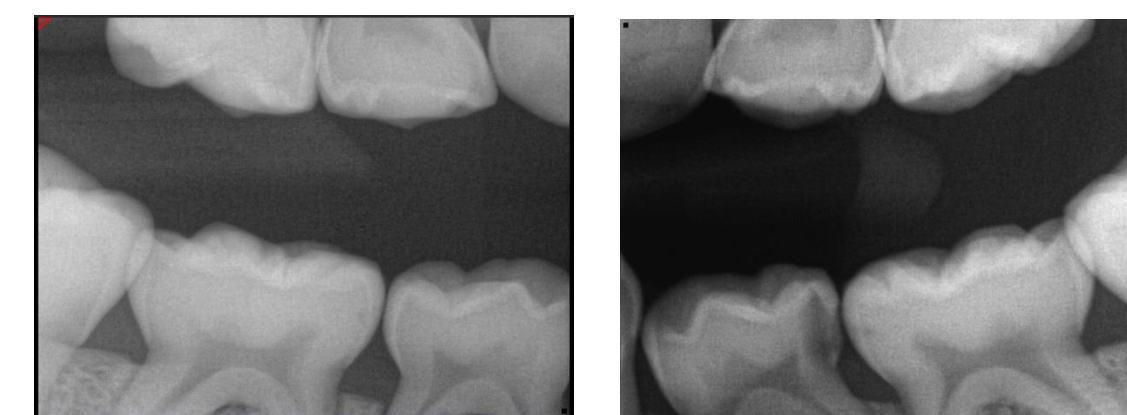


Fig.3 Bitewing radiographs



Fig.4 Anterior radiographs



Fig.5 Periapical radiograph of caries #L



Fig.6 Intraoral center photograph after treatment



Fig.7 Occlusal maxillary photograph after treatment



Fig.8 Intraoral right and left photograph after treatment

Conclusion

SAVI is a rare disorder that pediatric dentists may encounter. Each patient presents unique challenges, requiring adaptability in treatment approaches. Collaboration with other healthcare professionals is essential to ensure the best possible outcomes. It is crucial to consider the patient's medications and symptoms, as these factors may have oral and dental implications. Providing tailored dental care can significantly enhance the patient's quality of life.

References

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