



Autoimmune Polyglandular Syndrome Type 2: Case Report

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Introduction

Autoimmune polyglandular syndromes (APS) are polyendocrinopathies characterized by autoimmune dysfunction of multiple endocrine organs. According to the autoimmune manifestations it is classified into four main groups, designated APS-1, APS-2, APS-3, and APS-4.

Classification of APS	Diseases Associated
Type 1	<ul style="list-style-type: none"> Chronic candidiasis Chronic hypoparathyroidism Addison's disease (at least two present)
Type 2	<ul style="list-style-type: none"> Addison's disease (always present) Thyroid autoimmune disease and/ or Type 1 Diabetes
Type 3	<ul style="list-style-type: none"> Thyroid autoimmune disease associated with other autoimmune condition (excluding Addison's disease and/or hypoparathyroidism)
Type 4	<ul style="list-style-type: none"> Combination of organ specific autoimmune diseases non induced in previous groups

Table 1: Neufled and Blizzard's classification of APS (Rutigliano et. al, 2020)

Autoimmune polyendocrine syndrome type 2 (APS-2) is characterized by the triad of type 1 diabetes mellitus, Addison disease (adrenal insufficiency) and thyroid autoimmunity with hypothyroidism, hyperthyroidism, or Hashimoto thyroiditis. Report suggest that the prevalence of APS-2 is about 14 to 20 cases per million of population and that it affects females 3 to 4 times as often as it does males. The cause of APS2 is still unknown, although it may involve a combination of genetic and environmental factors. Individuals with APS-2 are prone to various oral issues due to autoantibodies attacking epithelial adhesion structures or tissue-targeting lymphocytes, causing inflammation and leading to specific pathological changes in the mucosal surfaces and skin.

(Sperling et. al, 2021)

This report describes the dental treatment of a child with APS-2 under general anesthesia, including the related oral symptoms. It emphasizes the importance of a collaborative team in managing the patient before surgery and the aftercare given during the early post-operative stage.

Case Report

A 9-year-old male with APS2 with clinical manifestations of Addison Disease, Hyperthyroidism, Hashimoto's Thyroiditis, Diabetes Type 1, and Sjogren Syndrome. Current medications include Prednisone, Lantus, Humalog, Plaquenil, Imuran, and Creon. The patient has no reported drug allergies. Clinical examination revealed multiple dental caries, gingivitis, enamel defects, oral candidiasis and xerostomia.

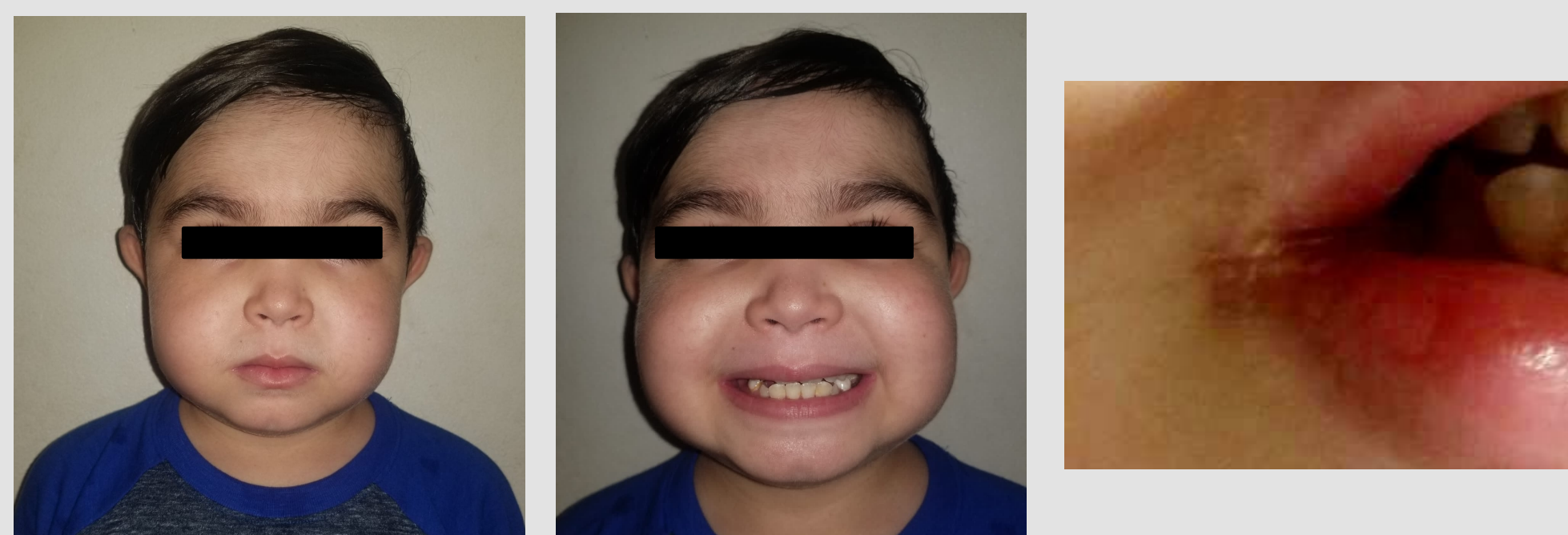


Figure 1: Pretreatment extraoral examination

Case Description

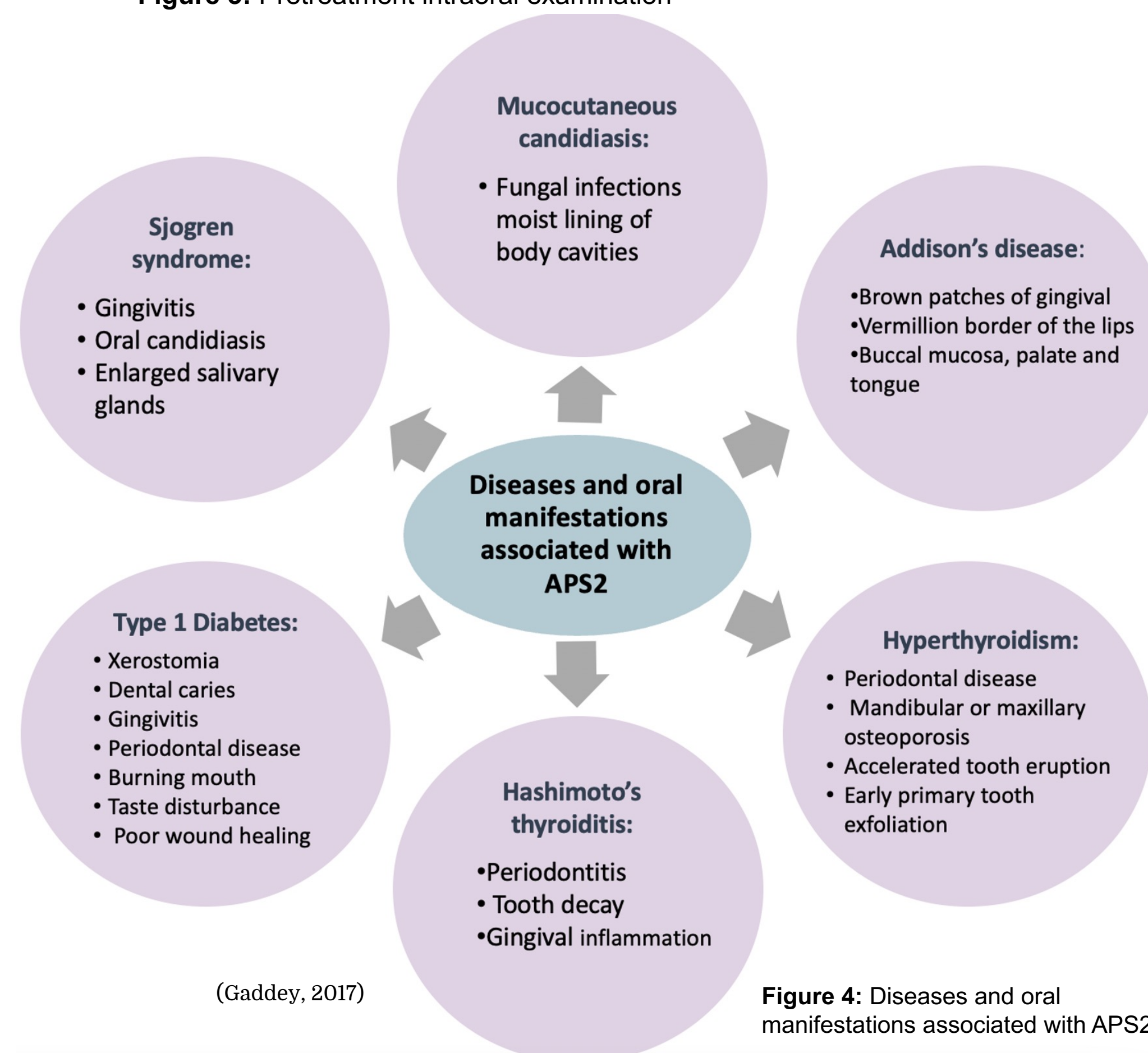
- Extraoral exam findings:**
 - Puffy face
 - Angular cheilitis
- Oral exam finding:**
 - Multiple dental caries
 - Gingivitis
 - Enamel defects
 - Oral candidiasis
 - Dry tongue



Figure 2: Pretreatment panoramic x-ray



Figure 3: Pretreatment intraoral examination



(Gaddey, 2017)

Figure 4: Diseases and oral manifestations associated with APS2



Figure 5: Dental Rehabilitation in operating room

Discussion

Preoperative:

A multidisciplinary team consisting of gastroenterologists, allergists, immunologists, endocrinologists, rheumatologists, and pediatricians was consulted for a patient at high risk for operative complications. The team recommended one day before to dental rehabilitation be admitted in hospital for a preoperative interventions such as administering hydrocorticoids through IV and reducing Lantus and Synthroid medication.

Operative :

On the day of the procedure, the patient's glucose levels were monitored, and specific considerations were taken for anesthetic management, including the risk of acute adrenal crisis. After general anesthesia induction and intubation, caries removal and resin restorations were placed on teeth num. 7 (MDLFI), 8(F), 9(F), 10(MDLFI), 19 (OB), and restorations on amalgam teeth num. 3(O), 30(OB). Sealants were applied on teeth 4, 5, 12 and 21. Local anesthesia (2% LIDO, 1/100K) was infiltrated on the area of extractions on teeth num. C, H, J and S with no complications, and hemostasis achieved.

Postoperative:

Instructions was discontinuing hydrocortisone, and changing to prednisone, and restarting Synthroid. A topical nystatin ointment was prescribed to treat oral candidiasis, resulting in a significant improvement of the lesions.

Conclusion

Overall, this case highlights the importance of a multidisciplinary approach and careful consideration of preoperative factors in patients with a rare autoimmune disorder.

Acknowledgements

Emilio Agrait Delfilló, DMD
Melba Esquilín Cruz, DMD

Cheryl Paulo Malavé, DMD
Rosana Hanke Herrero, DMD, MSD
Mariela Ramos, DMD

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