

## INTRODUCTION

Wiskott-Aldrich Syndrome (WAS) is a rare genetic condition of immunodeficiency characterized by micro-thrombocytopenia, recurrent infections, and eczema.<sup>1</sup> This syndrome has a propensity for bleeding due to a low number of platelets and small platelet size. One of the possible complications that can occur is a liver clot. A liver clot is a rare, postoperative bleeding complication involving an incomplete clot formation that continues to bleed slowly and grows over a surgical site. It generally presents 24-48 hours after an invasive dental procedure as a dark red, gel-like pedunculated mass that resembles tissues of the liver.<sup>2</sup> The objective of this report is to detail an occurrence of liver clots around stainless steel crowns (SSCs) and to discuss its management in a child with WAS.

## ETIOLOGY AND EPIDEMIOLOGY

### Wiskott-Aldrich Syndrome (WAS):

- A rare X-linked immunodeficiency caused by a mutated WASp gene resulting in cytoskeletal and signaling abnormalities in lymphocytes<sup>3</sup>
- Mutated WAS proteins impair platelet development and function and ability to mount a robust immune response against pathogens
- Presents in males and affects 1-10 per 1 million with the average age at the time of diagnosis being 24 months<sup>2,4</sup>

### Liver Clot:

- Many factors contribute to a liver clot formation:<sup>4</sup>
  - Venous hemorrhage from vascular damage during dental surgery
  - Presence of foreign bodies (i.e. residual dental cement)
  - Infection and delayed wound healing
  - Deviation from post-operative instructions and care
- Only one case series described liver clots in healthy pediatric patients after receiving SSCs.<sup>5</sup>

## DIAGNOSIS AND MANAGEMENT

- Diagnosis of a liver clot is based on medical history, dental history, and clinical presentation
- Differential diagnoses include pyogenic granuloma, peripheral giant cell granuloma, and angiosarcoma
- Previously reported treatment regimen involves liver clot removal with a high-speed suction or curette, followed by saline irrigation and direct pressure for hemostasis.<sup>2,4</sup> Local or systemic hemostatic agents may be indicated for continuous bleeding.<sup>6</sup>

## CASE REPORT

A 4-year-old male patient with Wiskott-Aldrich Syndrome was admitted to CHOC due to persistent gingival bleeding around primary molar SSCs and increased swelling of the gums bilaterally. The child received a platelet transfusion and underwent full mouth oral rehabilitation under general anesthesia in a hospital setting 48 hours prior to admission. The patient's mother reported that the patient had been chewing on the insides of the mouth and avoided eating due to oral discomfort for the past 2 days. Clinical examination revealed purple to red gelatinous masses around all posterior molar SSCs with mild, slow bleeding around the periphery (**Figure 1**). The diagnosis of liver clots was made based on medical history, dental history, and clinical presentation.



**Figure 1.** Liver clots presenting as purplish-red gelatinous masses presenting around SSCs of the mandibular posterior quadrants



**Figure 2:** After removal of liver clots, mild oozing of residual blood observed around SSC margins that was controlled with gauze/pressure for 10 minutes

### Patient Management:

After admission, medical/dental management consisted of the following:

- Pre-operative labs drawn prior to removal of liver clots (PLT count of 41,000/mm<sup>3</sup>)
- Intranasal Versed sedation administered by the ED physician
- Benzocaine 20% topical anesthetic applied around perimeter of gingival mass
- Liver clots removed with curettage and gauze (**Figure 2**)
- Sites irrigated with saline water
- Direct, firm pressure applied with gauze for 10 minutes until hemostasis achieved
- Oral Amicar prescribed for 3 days (1.25g/5mL q6h)

### 1-week Follow-up:

- All SSCs intact with surrounding soft tissues healing normally (**Figure 3**)
- No evidence of liver clots or residual bleeding present



**Figure 3:** 1-week follow up images showing SSC and extraction sites healing within normal limits

## DENTAL IMPLICATIONS

- Although postoperative bleeding after dental surgery is typically self-limiting in healthy children, patients with bleeding disorders are at an increased risk of excessive bleeding from minor wounds.
- Possible complications, including liver clots, should be considered for these at-risk patients with an understanding and knowledge of prevention and appropriate management and care.
- In this case report, a pediatric patient with WAS developed multiple liver clots after SSCs were placed, even after supportive measures were taken during surgery including pre-operative platelet transfusion and the use of local hemostatic agents.
- WAS is characterized by thrombocytopenia with oral manifestations including petechiae, spontaneous gingival bleeding, and reduced ability to form blood clots.<sup>3</sup> Additionally, because the platelets are smaller, WAS patients often exhibit more bleeding at any given platelet count.<sup>1</sup>
- To prevent excess bleeding, additional precautions that should be taken include systemic hemostatic agents, such as aminocaproic acid or tranexamic acid, and postoperative hospital admission for observation.<sup>7</sup>

## CONCLUSION

This case report presents a rare case of liver clot formation after placement of multiple SSCs in a pediatric patient with WAS, including its presentation and management. A liver clot is a rare postoperative bleeding complication characterized by a poor clot formation and delayed hemostasis after an invasive dental procedure. Although previously reported in adults after extractions or periodontal surgery, liver clots can also present in the pediatric population in association with SSCs.<sup>5</sup> The best prevention and management for a medically compromised patient is completing a thorough preoperative evaluation of the patient and treatment planning in collaboration with the medical team.<sup>6</sup> Prompt diagnosis and management of liver clots are necessary to avoid excessive or persistent bleeding.

### REFERENCES:

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