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Beyond the Liver: Primary Sclerosing Cholangitis in Dental Care

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Learning objectives

1. Define Primary Sclerosing Cholangitis (PSC) and its symptoms.
2. Define viable management of PSC.
3. Identify what oral manifestations may present.
4. Discuss key considerations related to providing effective oral hygiene care.

Abstract

Primary Sclerosing Cholangitis (PSC) is a rare, chronic cholestatic liver disease characterized by progressive inflammation and fibrosis of the bile ducts, resulting in strictures, cirrhosis, and eventual end-stage liver disease. The incidence of PSC is approximately 0.87 cases per 100,000 person-years, with a prevalence of 13.53 per 100,000, highlighting its rarity but clinical significance.

Clinical manifestations of PSC include jaundice, pruritus, fatigue, vitamin deficiencies, abdominal pain, and complications such as portal hypertension. The prognosis is variable, with an average life expectancy ranging from 10 to 20 years following diagnosis. Current treatment options are limited while liver transplantation remains the most effective treatment for advanced disease.

PSC has significant implications for oral health. Common findings include xerostomia, increased clinical attachment loss, progression of periodontal disease, oral lesions, and delayed wound healing. Xerostomia, often the most prevalent symptom, increases the risk for dental caries, mucosal irritation, and functional difficulties such as swallowing and speaking. Additionally, vitamin deficiencies and the use of immunosuppressive medications can further compromise oral health and healing.

Dental management of patients with PSC requires careful planning, including thorough medical history evaluation and collaboration with the patient's physician. Hepatic impairment may reduce drug metabolism, necessitating modifications in local anesthetic dosing and medication selection. Preventive strategies, including daily fluoride use and routine varnish applications, are essential to minimize oral complications.

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Introduction

Primary Sclerosing Cholangitis (PSC) is a cholestatic disease of the liver that affects the bile ducts of a patient due to fibrosis, which causes strictures of the bile ducts and leads to cirrhosis and with time, end-stage liver disease [1,2]. A 2023 study specified the overall pooled incidence rate for PSC was 0.87 events per 100,000 person-year, and the pooled prevalence rate was 13.53 events per 100,000 person-year [3]. This indicates the overall incidence and prevalence for PSC to be very low for the general population. With low incidence and prevalence, patients with PSC and liver disease could receive little consideration, but still require special attention regarding their healthcare needs. This brings about the need for further discussion and attention among healthcare providers to enhance general understanding within the profession, specifically in the dental setting.

Pathophysiology

Currently in the United States there are roughly 29,000 patients who have been diagnosed with PSC [1]. The disease is more common in males compared to females at about a 3:2 ratio [1]. Incidence over the years has seemed to increase, but some research has speculated that this may be due to the advancements in technology that enable healthcare providers to more accurately diagnose the disease [1]. However, when comparing this disease to other more common diseases, its reduced overall prevalence is illustrated by its statistics and the lack of awareness by many in the healthcare field.

The initial signs and symptoms of PSC generally reflect the development of progressing injury of the bile ducts from the liver. With this progression patients will begin to encounter strictures in bile ducts which lead to cholestasis and biliary cirrhosis that causes the patient to experience continual progression and dysfunction of the liver [3]. Continued progressive symptoms will appear as jaundice, pruritus, fatigue, vitamin deficiency, portal hypertension, varices, and pain in the right upper quadrant area. In later stage disease, a patient can start to experience ascites and encephalopathy and as the strictures advance and continually develop within the bile ducts, the jaundice will become more obvious followed by changes in the color of one's urine and stool [4].

The prognosis of PSC can be somewhat formidable as the disease is very insidious but can experience a gradual disease process without effective treatment. The life expectancy for PSC falls somewhere between 10 to 20 years on average [5]. However, depending on the level of symptoms a patient is experiencing can distinguish between overall survival. Research identified that patients who were asymptomatic and did not have decompensated cirrhosis at seven years would have an estimated survival of 75% in comparison to 96% for same age and sex matched healthy individuals [6].

The overall etiology of PSC is not clearly understood, as there seems to be no direct cause. Although it is understood by most who study the disease, there are some probable environmental and genetic correlations [4]. Most environmental factors seem to be related by association only [5]. Research has illustrated that patients who smoke and drink coffee experience less incidence of the disease [6,7]. Association has also been correlated for patients with PSC who experienced a higher-than-normal amount of time around undomesticated animals, such as farm animals [6]. Other possible factors believed to exist include reduced use of contraceptive hormones, increased urinary tract infections in females, and patients who were more likely to consume hamburger and steak cooked well-done, all of which experienced a higher incidence of the disease [6,8]. When studying genetic relationships with PSC, many studies have identified genetic susceptibility for patients developing the disease [9,12]. Researchers have been able to identify that risk of the disease for first-degree relatives, such as siblings, can have nine to 39 times greater risk for the development of the disease when compared to the general population [13]. Although the etiology is not well understood for PSC, and there is no direct cause correlated with the disease, both environment and genetics are believed to play a role in the causation of the disease.

Manifestation of the clinical signs and symptoms of the disease is well documented. Many patients can experience several complications: jaundice, vitamin deficiency, abdominal pain, bone disease, chronic itching, varices, polyps, hepatomegaly, splenomegaly, oral disease, and possible malignancies [14-17]. As these symptoms progress, the disease can become more debilitating and impose continued issues for the patient. To complicate the situation, PSC can often be associated with inflammatory bowel disease. It is estimated that about 70 to 80 percent of PSC patients present with both diseases [18]. However, only a small portion of the patients with inflammatory bowel disease like Ulcerative Colitis or Crohn's disease develop PSC, which equates to about five percent of the patient population [14]. An unhealthy gut microbiome can be associated with systemic inflammation that may manifest in the oral cavity [19].

Oral manifestations associated

A connection has been established between a patient having infections in the oral cavity with the presence of liver disease, however, the causative relationship between the two has not been directly correlated to any specific cause [20,21]. When considering the effect of periodontal disease on a patient with liver disease, we have found that patients with periodontal disease can have an increasingly negative effect on the progression of a patient's liver disease as well. This shows a greater level of importance for these patients to ensure oral health professionals are giving special consideration to this population [22].

Xerostomia is a studied effect in liver disease patients, which is experienced frequently [23,24]. With xerostomia present, this can lead to an increase in other oral health issues such as decay, periodontitis, mucosal lesions, and stomatitis [25,26]. Other side effects due to xerostomia are changes in taste, difficulty eating, swallowing, talking, and wearing a dental prosthesis [27].

Chronic liver disease patients take multiple medications including immunosuppressants which in turn changes the microbiota in the oral cavity and leads to continued oral health issue [28]. These medications have demonstrated to cause gingival hyperplasia, increasing the incidence of periodontitis as it is difficult to maintain meticulous oral hygiene [29].

As identified, this population of patients are more susceptible to vitamin deficiencies including A, D, E and K. This can lead to increased susceptibility to decay, inflammation and periodontal disease progression, prolonged bleeding, and delayed wound healing. Additionally, severe deficiency can contribute to intraoral ulcers.

If cirrhosis advances, transplantation may be recommended. Routine pretransplatation dental screenings are empirically sug-

gested as infections are a major cause for mortality among immunosuppressed post liver transplant patients [27]. One study suggests those in the field of organ transplantation agree that teeth with obvious infection and non-restorable teeth must be extracted prior to transplantation, although there are no generally accepted guidelines to suggest mandatory dental treatment prior to organ transplantation [29]. Patients may require a medical consult with a referral for treatment to be completed by a periodontist or oral surgeon as they are able to carefully provide and monitor extensive oral procedures in order to prevent the danger of profuse bleeding [25]. Oral infections such as periodontal disease in combination with systemic conditions such as liver disease may affect bone structure and have negative effects on the periodontium [27].

Management

Treatment of PSC can be somewhat limited in its effectiveness in combating the disease since the disease is rare and idiopathic in nature [19]. Researchers have indicated the use of Ursodeoxycholic Acid as a plausible treatment initially to see how it affects the overall health of the patient, and then it is recommended to decide on its continued use based on improved or stable lab results [5].

The final treatment option for PSC patients that will be discussed is a liver transplant. A liver transplant is considered to be the most effective way to treat PSC when patients are in advanced stages [24]. Due to the progressiveness of the disease, about 40% of patients will require a liver transplant, and the time between diagnosis and liver transplantation is about 20 to 25 years [24,25. Once a liver transplant has been completed, there can still be a 25 percent risk of recurrence of PSC, which could lead to the need of an subsequent transplant [24].

Importance of oral health

Patients with liver impairment, such as those with Primary Sclerosing Cholangitis (PSC), may seek preventive oral care or emergency treatment. A comprehensive review of the patient's medical history is essential to guide treatment decisions and determine whether a referral or medical consultation is necessary.

Common oral findings in these patients include xerostomia (the most frequently observed), increased clinical attachment loss, progression of periodontal disease, oral lesions, and delayed healing. Because xerostomia is prevalent, patients should be encouraged to use daily fluoride products and receive regular in-office fluoride varnish applications to help prevent caries.

Careful consideration should be used prior to the administration of anesthesia. It is pertinent to consider how hepatic impairment can lead to reduced metabolism of local anesthetics, which may require a lower dosage of these drugs or even be totally avoided. When considering changes or additions of medications in these patients, the treating physician should be included in all discussions [30,31].

Conclusion

When considering liver disease such as PSC, it is important for dental professionals to be aware of the additional contraindications that can present in patients and ensure all possible negative outcomes be reduced. While liver disease patients can have increased oral cavity issues, the reverse can also be true where these oral cavity issues can inherently cause progressively worsening liver issues if the oral cavity problems are not resolved in a timely manner. It is imperative that the oral health community be educated on the adverse effects of PSC and similar liver diseases.

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