CASE REPORT

Case Report: Dentigerous cyst marsupialization for a child with Hunter’s syndrome [version 1; referees: awaiting peer review]

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Abstract

Hunter’s syndrome or mucopolysaccharidosis (MPS) type II is an inherited disorder caused by enzyme iduronate-2-sulfatase deficiency. It is characterized by involvement of the nervous, cardiovascular, respiratory, and musculoskeletal systems, along with numerous oral manifestations. This is a case report of an eight year-old girl diagnosed with Hunter’s syndrome, who was referred to the Pediatric Dentistry Department, Faculty of Dentistry, Cairo University with a chief complaint of hard swelling related to the lower left posterior area. Radiographic examination revealed well defined corticated radiolucency surrounding an unerupted lower left first molar. Aspiration was done and cytopathologic examination revealed cystic fluid mixed with blood. The case was diagnosed as a dentigerous cyst. Cyst marsupialization was done under general anaesthesia. From this case report we concluded that in Hunter’s syndrome patients more conservative approaches are more valuable. Regular dental follow up is advised to maintain good oral hygiene, and to detect any complications as early as possible.

Keywords

Dentigerous cyst, Marsupialization, Mucopolysaccharidosis, Hunter’s syndrome, Case report.

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Introduction
Hunter syndrome, or mucopolysaccharidosis type II (MPS II) is a rare metabolic disorder. It was first described in 1917, named after physician Charles A Hunter. It is inherited as an autosomal recessive trait, and results in lysosomal enzyme iduronate-2-sulfatase deficiency. Prevalence of MPS II-H has been reported to be 1 in 170,000 cases, and no predilection for sex and ethnicity has been found. Recently enzyme replacement therapy has emerged as a new treatment. Supportive management and physical therapy are also very important in the management of MPS type II.

Systemic manifestations of Hunter’s syndrome are macrocephaly, moderate to severe developmental delay, dysmorphic facies, skeletal abnormalities, joint contractures, hepatosplenomegaly, cardiac valvular disease, as well as corneal clouding, and puffy eyelids. Intraoral manifestations include an enlarged tongue, hyperplastic gingivae, broad arches with interdental spacing, anterior open bite, hypoplasticity, peg-shaped teeth with delayed development and eruption.

Case report
An eight year-old girl diagnosed with Hunter’s syndrome. She was born to healthy, consanguineous parents as their second child. She was referred to the Pediatric Dentistry Department, Cairo University in June, 2017 with a chief complaint of hard swelling related to the lower left posterior area that can be easily felt on palpation. The patient has no previous dental history, it was her first dental visit.

Clinical examination showed a hard bony swelling obliterating buccal vestibule related to the unerupted lower left first permanent molar. Adjacent primary teeth; lower left first and second primary molars were sound with normal mobility and no pain on percussion.

Radiographic examination revealed well defined corticated radiolucency surrounding the unerupted lower left first molar. There was significant root resorption related to the roots of the first and second primary molars. Delayed eruption of the permanent first molars was also present, with the lower right molar displaced towards the inferior border of the mandible. Enlarged dental follicles of the lower second permanent molars, as well as shortened lower permanent incisors. (Figure 1).

Aspiration was performed using a sterile plastic syringe; cystic fluid mixed with blood was found (Figure 2). Examination of aspirated fluid was done performed using a light microscope, by a pathologist in the Department of Oral and Maxillofacial Pathology. It revealed red blood cells with cholesterol crystals. Final diagnosis was reached by exclusion. List of differential diagnosis was as follow: dentigerous cyst, unicystic ameloblastoma, odontogenic keratocyst. Odontogenic keratocyst; was excluded because it gives white cheesy material on aspiration. Dentigerous cyst and unicystic ameloblastoma, have similar clinical and radiographic presentation, but it was diagnosed as a dentigerous cyst as they are common in patients with Hunter’s syndrome.

Cyst marsupialization was the choice of treatment, and the patient was referred to her physician to be prepared for the surgical intervention under general anaesthesia. Two weeks later, surgery was performed by the dental team; oral surgeon, pedodontist and dental assistants. Marsupialization was performed and a drain was placed. The patient was scheduled for follow up visits every two weeks to check the drain for two months post-surgery.

After a one year follow up period it was found that clinically on palpation, there was no hard bony swelling related to the lower left first permanent molar, but the tooth was yet to erupt. Radiographically there was an increase in bone density around the lower left first permanent molar indicating normal bone healing and shrinkage of cyst size. The lower left first permanent molar had moved towards occlusal plane (Figure 3).

Figure 1. Panoramic radiograph showing well defined corticated radiolucency surrounding an unerupted lower left first molar.

Figure 2. Photograph showing plastic syringe filled with blood stained fluid.

Figure 3. Panoramic radiograph showing lower left first permanent molar moved occlusally with increased bone density around.
Discussion
Hunter’s syndrome patients suffer from permanent, progressive cellular damage which affects all organs and systems functioning. The patient was very apprehensive, exhibited aggressive behavior, had a large tongue, and limited mouth opening. Consultation with the physician and general anesthesia team was done to ensure the surgical procedure would have least possible risk.

Delayed development and eruption of teeth, dentigerous cysts, and enlarged dental follicles are common features in Hunter’s syndrome patients. In the presented case enlarged dental follicles are evident in the lower second permanent molars. Enlarged dental follicles are due to pools of chondroitin sulfate B. These lesions contain dense, fibrous connective tissues and large amounts of acid mucopolysaccharides. These areas of destruction tend to worsen with age.

Downs et al. stated that cystically involved teeth in Hunter’s syndrome patients should be removed, however, in our case cyst marsupialization was considered for a number of reasons. First was to be more conservative, to avoid total loss of the first permanent molar. Second was to avoid risk of pathologic fracture of mandible, which may have occurred if cyst inoculation was performed. The enlarged tongue and limited mouth opening also made invasive surgery more difficult.

Marsupialization is not an aggressive technique but it requires meticulous postoperative care. Wound infection and delayed healing were expected. A drain was placed at the site of surgery and both the patient and her parents were educated and motivated to apply good oral hygiene measures and to maintain the drain in place. Surprisingly, normal healing with no infection occurred. This agrees with Savitha et al. who stated that aggressive surgery is usually not recommended for Hunter’s syndrome patients to avoid any complications.

Although, the presented treatment modality doesn’t eliminate the pathologic condition at the time of surgery, and requires multiple postoperative follow up visits, it provides a conservative, non-aggressive line of treatment with good healing for a systemically compromised, syndromic case.

Conclusion
Hunter’s syndrome is a complex medical condition that necessitates regular dental follow up to maintain good oral hygiene, and to detect any complications as early as possible. With regards to dental lesions, less aggressive with more conservative approaches are recommended.

Patient perspective
Although the treatment required long and exhausting follow up visits, the patient and her parents were pleased with the more conservative treatment performed. The hard swelling gradually disappeared with time, and the patient kept her tooth.

Consent
Written informed consent for publication of the clinical details and images was obtained from the patient’s father.

Data availability
All data underlying the results are available as part of the article and no additional source data are required.

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References
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