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CASE REPORT

Case Report: Dentigerous cyst marsupialization for a child with Hunter's syndrome [version 1; peer review: 1 approved, 1 approved with reservations, 1 not approved]

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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.

Open Peer Review

Approval Status

	1	2	3
version 1			
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Any reports and responses or comments on the article can be found at the end of the article.

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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 Hunter¹. 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 found². 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 II³.

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^{4,5}.

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).

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

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 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 functioning⁶. 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 patients⁷. 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 age⁸.

Downs *et al.*⁷ 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.*⁴ 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.

Grant information

The author(s) declared that no grants were involved in supporting this work.

Acknowledgement

We want to thank all the team worked in this operation; general anaesthesia team, oral surgeon and dental assistants for their great effort. Our gratitude also extends to the patient and her parents, who were very cooperative.

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Open Peer Review

Current Peer Review Status:

Version 1

Reviewer Report 02 January 2019

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Iyad Hussein

Department of Paediatric Dentistry, Hamdan Bin Mohammed College of Dental Medicine, Mohammed Bin Rashid University Of Medicine and Health Sciences, Dubai, United Arab Emirates

This article presents a case of a dentigerous cyst in a rare case of Hunter's syndrome. Overall it is good. Some corrections are suggested here.

Paragraph 1 Line 4

Add "Hunter's syndrome clinically resembles other mucopolysaccharide disorders such as Hurler's, San Filippo's, Morquio's, and Schere's syndromes, which are autosomal recessive. Hunter's syndrome is the exception being transmitted as an X-linked recessive disorder, thus mainly seen in males. Thus this does have a preference for males."

This is mentioned in the reference the authors quoted (number 7)¹.

Paragraph 1 Line 6: sex - as above comment.

Paragraph 3: English needs rewording.

I suggest the following:

"An eight year old girl, born as a second child to healthy consanguineous parents, was diagnosed with Hunter's syndrome. She was referred to the Paediatric Dentistry Department, Cairo University in June 2017 with a chief complaint of a hard swelling related to the lower left posterior area of her mandible, that was easily felt on palpation."

Paragraph 4 Line 2 should read: "first" not "fist"

Paragraph 4 Line 3 should read: "The adjacent..." not "Adjacent.."

Paragraph 5 Line 1 should read: "revealed a well defined..."

Paragraph 5 Line 8 should read: "...lower permanent incisors were also noted (Figure 1)."

Paragraph 6 Line 2 should read: "Examination of the aspirated.."

Paragraph 6 Line 6 should read: "A list.."

Paragraph 6 Line 8 should read: "keratinizing odontogenic tumour" not "Odontogenic keratocyst" according to Welbury et al².

Paragraph 6 Line 11 should read: "presentations"

References

1. Downs AT, Crisp T, Ferretti G: Hunter's syndrome and oral manifestations: a review. *Pediatr Dent.* **17** (2): 98-100 [PubMed Abstract](#)
2. Welbury et al.: Paediatric Dentistry 5th Edition. 2018. 604

Is the background of the case's history and progression described in sufficient detail?

Yes

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Yes

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Yes

Is the case presented with sufficient detail to be useful for other practitioners?

Partly

Competing Interests: No competing interests were disclosed.

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Reviewer Report 19 December 2018

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Onur Şahin

Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, İzmir Katip Çelebi University,

İzmir, Turkey

I appreciate your submission. This study is of interest to dentistry and medicine, to those who diagnose and treat paediatric patients with Hunter's syndrome. But, the manuscript should be revised and the author should provide the more detail data, to give useful clinical information to readers. So, I suggest the following revisions and advice before accepting it for indexing:

1. How was Hunter's syndrome diagnosed in this patients? You should give more detailed data about patient's medical history.
2. If patient's physical appearance and intra-oral views could be presented, your paper would be perfect.
3. More information is needed on how to apply marsupialization. Bone removed? Or buccal bone resorbed?
4. If the view of placed drain could be presented, your paper would be perfect.
5. In these patients, orthodontic treatment may be needed to erupt the tooth after marsupialization. This should be added to the discussion section.

Hunter's syndrome is a very serious disease affecting children. Aggressive surgery is usually not advocated for Hunter's syndrome. This study was to present that marsupialization could be the first choice treatment for dentigerous cyst in preadolescents. It should be useful article to oral & maxillofacial surgeons and general dental practitioners, if the authors perform more research and revision.

Thank you again for your submission.

Is the background of the case's history and progression described in sufficient detail?

Partly

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Yes

Is the case presented with sufficient detail to be useful for other practitioners?

Yes

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: implantology, oral and maxillofacial surgery, TMJ disorders, medication related osteonecrosis of the jaw, orthognathic surgery

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Reviewer Report 17 December 2018

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Nedal Abu-Mostafa

Oral and Maxillofacial Surgery and Diagnostic Science Department, Riyadh Elm University, Riyadh, Saudi Arabia

This report presented a case of an eight year-old girl diagnosed with Hunter's syndrome. She had hard bony swelling on the buccal vestibule related to the unerupted lower left first permanent molar. Radiographically: there was a well-defined radiolucency surrounding the unerupted lower left first molar with radio-opaque margin. Marsupialization was performed and a drain was placed. Follow up was done.

The case report is interesting however, I have some comments:

- Signs and symptoms of Hunter syndrome on the patient face weren't mentioned. As well, there are no intra-oral and extra-oral clinical pictures.
- The part of cystic lining that has been removed during marsupialization should be taken as incision biopsy. So the final diagnosis should depend on the histo-pathologic examination
- The panoramic radiograph shows only very minimal movement of lower left 1st molar. The eruption of the tooth and other permanent 1st molars could be achieved if orthodontic treatment has been carried out after 3 months of marsupialization.
- Discussion does not mention if there are similar previous case reports of hunter syndrome with dentigerous cyst that was treated by marsupialization.
- The conclusion should reveal the effectiveness of marsupialization as a conservative method for treatment of dentigerous cyst with Hunter syndrome with evidenced bone deposition. However the written conclusion put first the necessity of follow up and oral hygiene which was not evaluated the case report!

Is the background of the case's history and progression described in sufficient detail?

Yes

Are enough details provided of any physical examination and diagnostic tests, treatment given and outcomes?

Partly

Is sufficient discussion included of the importance of the findings and their relevance to future understanding of disease processes, diagnosis or treatment?

Partly

Is the case presented with sufficient detail to be useful for other practitioners?

Partly

Competing Interests: No competing interests were disclosed.

Reviewer Expertise: Oral surgery

I confirm that I have read this submission and believe that I have an appropriate level of expertise to state that I do not consider it to be of an acceptable scientific standard, for reasons outlined above.

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