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Oral-surgical management of an odontogenic keratocyst in a patient with Duchenne muscular dystrophy

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Abstract

Duchenne muscular dystrophy, a debilitating disease affecting male children, presents special considerations for the dentist faced with an oral-surgical problem. When general anesthesia is contraindicated, local anesthesia and a change in technique is indicated. One technique of outpatient surgical management of an odontogenic keratocyst, in a patient with Duchenne muscular dystrophy, is described.

Introduction

Duchenne muscular dystrophy, also known as pseudohypertrophic muscular dystrophy, is a sex-linked, recessive, degenerative myopathy of male children.[1,2,3] Usually diagnosed before six years of age,[1,4] it is the most common childhood myophathy, occurring once every 3,000-4,000 births.[5] Although often hereditary, Roses et al. reported that one-third of all cases may be spontaneous mutations,[6] and Zundel reported an even higher incidence of mutation.[3]

Clinical examination of children with Duchenne muscular dystrophy reveals a pseudohypertrophy of calves, deltoids, triceps, proximal muscle weakness, waddling gait, lordosis, and difficulty in standing.[3,7] Prior to diagnosis, the child may be considered clumsy or awkward,[8] and in many patients intelligence is decreased with language severely affected.[9]

The disease progesses from the pelvis to the shoulder girdle and upper limbs. Flexion contractures and scoliosis ultimately occur.[7]

The pathogenesis and biochemical defect of Duchenne muscular dystrophy are not yet understood.[2,6,10] Investigators have discovered significant increases in serum enzymes, especially creatine phosphokinase.[1,2,6,10,11]

Duchenne muscular dystrophy is a debilitating disease where most boys are nonambulatory by twelve years of age.[12] Although there is no cure, daily activity and respiratory exercises are stressed. Death occurs by the second or third decade from respiratory infection or heart failure.[1]

This paper will discuss one approach to the outpatient oral-surgical management of a patient with an odontogenic keratocyst whose medical history includes Duchenne muscular dystrophy and a seizure disorder.

Case Report

The patient, a twelve-year-old Caucasian male, was referred to Childrens Hospital of Philadelphia, (CHOP), Department of Dentistry for treatment of an oral swelling.

Chief Complaint

The patient complained of a swelling on the left side of his mandible. Traumatic origin was ruled out. He was examined by oral surgeons, who, after considering his medical history, referred him to CHOP on November 5, 1976.

Past Medical History

The patient was born on December 29, 1964. Labor was induced due to eclampsia and culminated in vaginal delivery. Developmental milestones were moderately delayed. When supporting the child upright at 18 months, the parents noted that he tended to stand on his toes.

An orthopedist diagnosed and treated him for tight heel cords, and recommended leg braces for two years. Six months after the braces were removed, toe walking recurred. On January 3, 1971, the patient was admitted to CHOP. Findings included a positive Gowers sign, lordotic gait, rubbery calves, relative hyperflexia, right equivocal Babinski response, and waddling. Serum creatine phosphokinase levels were elevated on two successive tests. There was no family history of neuromuscular disease.

A muscle biopsy was performed; the pathology report and clinical signs were consistent with Duchenne muscular dystrophy.

In September 1972, he was readmitted to CHOP for a convulsive disorder. Seizure control was achieved

and maintained with Dilantin 50 mg TID, and Phenobarbital 30 mg BID.

Oral Findings and Treatment

The patient presented with a class I molar relationship in the transitional dentition and interdental spacing was evident throughout the dentition. The oral hygiene was fair and maintained by the mother. A swelling was noted distal and buccal to the permanent left first molar.

A panoramic radiograph taken at the time of examination revealed a large, 4 cm x 5 cm, radiolucent lesion extending from the first molar distal to and entering the ramus. The second molar was displaced to the inferior border of the mandible, while the third molar was displaced into the ramus. The lesion was asymptomatic and consistent with a dentigerous cyst (Figure 1).

Initially, 1.8 cc of 2% lidocaine with 1:100,000 epinephrine was infiltrated on the crest of the alveolar ridge distal, lingual, and buccal to the left first molar. After aspirating 5 cc of straw colored fluid, a #15 B-P scapel blade was utilized to remove a 1 cm sample of tissue from the roof of the lesion. This tissue was submitted for histologic examination. The wound was packed with 45 cm of iodoform gauze coated with BIPPS dressing; the patient tolerating the procedure well.

Two weeks postoperatively the mother was instructed in irrigation techniques, and how to change the dressing.

The histological evaluation revealed the lesion to be an odontogenic keratocyst (Figure 2).

By May 1978, the tooth had erupted sufficiently so that removal and cyst enucleation could be performed. Local anesthesia was administered and an incision was made from the base of the ramus mesial to the second premolar. A buccal flap was reflected with a Cameron elevator and the cystic cavity revealed. The lining was gently curetted from the bony cavity and the second molar removed with elevators. Primary closure was achieved and the postoperative course was benign and uneventful. Figure 3 is a follow-up panoramic radiograph taken January 1979.

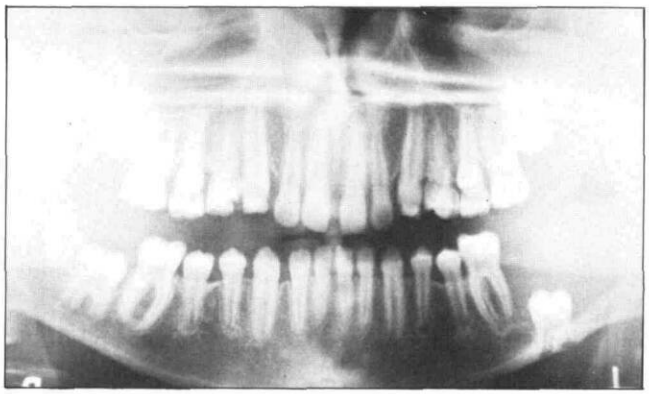

Figure 1. Panoramic radiograph taken November 1976. Note the cystic lesion displacing the second molar apically on the left side of the mandible.

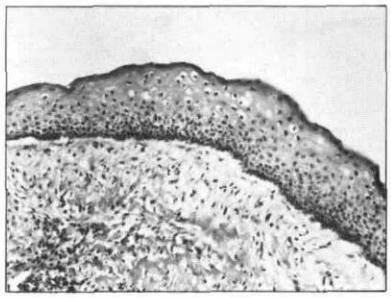

Figure 2. Photomicrograph of biopsy specimen reveals a surface epithelium covered by a thickened layer of parakeratin and a lining epithelium composed of a single layer of parakeratin, followed by a spinous cell layer and polarized basal cell layer (H & E, original magnification x 150).

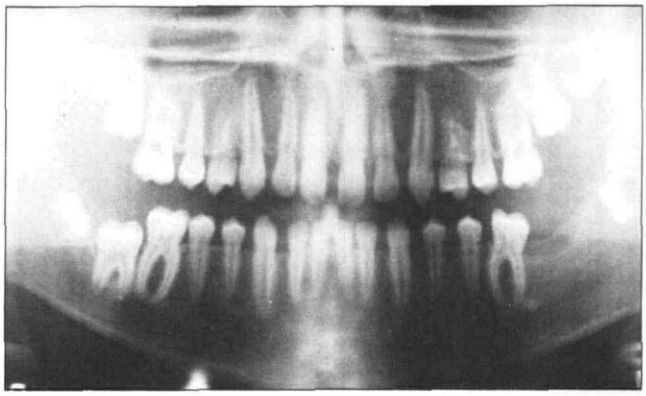

Figure 3. This panoramic radiograph is from January 1979, after second molar removal. Note the excellent healing of the cystic lesion.

Discussion

The dental management of patients with Duchenne muscular dystrophy can be complicated by behavior problems, malocclusions, and repeated respiratory infections.

General anesthesia is contraindicated in these children.[4] Atrophy of respiratory muscles eventually reduces the vital lung capacity and interferes with the ability to cough and clear the throat of salivary secretions.[4] Since the patient was manageable in the presence of the mother, an outpatient approach utilizing local anesthesia was used.

The clinical and radiographic appearance of the lesion suggested it to be a dentigerous cyst. Only after histologic evaluation was the correct diagnosis of odontogenic keratocyst discovered. This diagnosis affects the prognosis, because compared to dentigerous cysts, they are difficult to remove and tend to frequently recur.[13-15] Thus, the importance of submitting biopsy material is demonstrated and should be considered when treating lesions of the type illustrated by this case.

The size of the cyst and possibility of nerve damage or fracture of the jaw precluded a one-appointment procedure. The two-step procedure, lasting eighteen months, was only possible with patient and parental cooperation.

Duchenne muscular dystrophy remains an enigmatic disease. Although prenatal screening and genetic counseling provide some hope for decreasing its occurrence, further research is needed to understand the cause and treatment of this disease.

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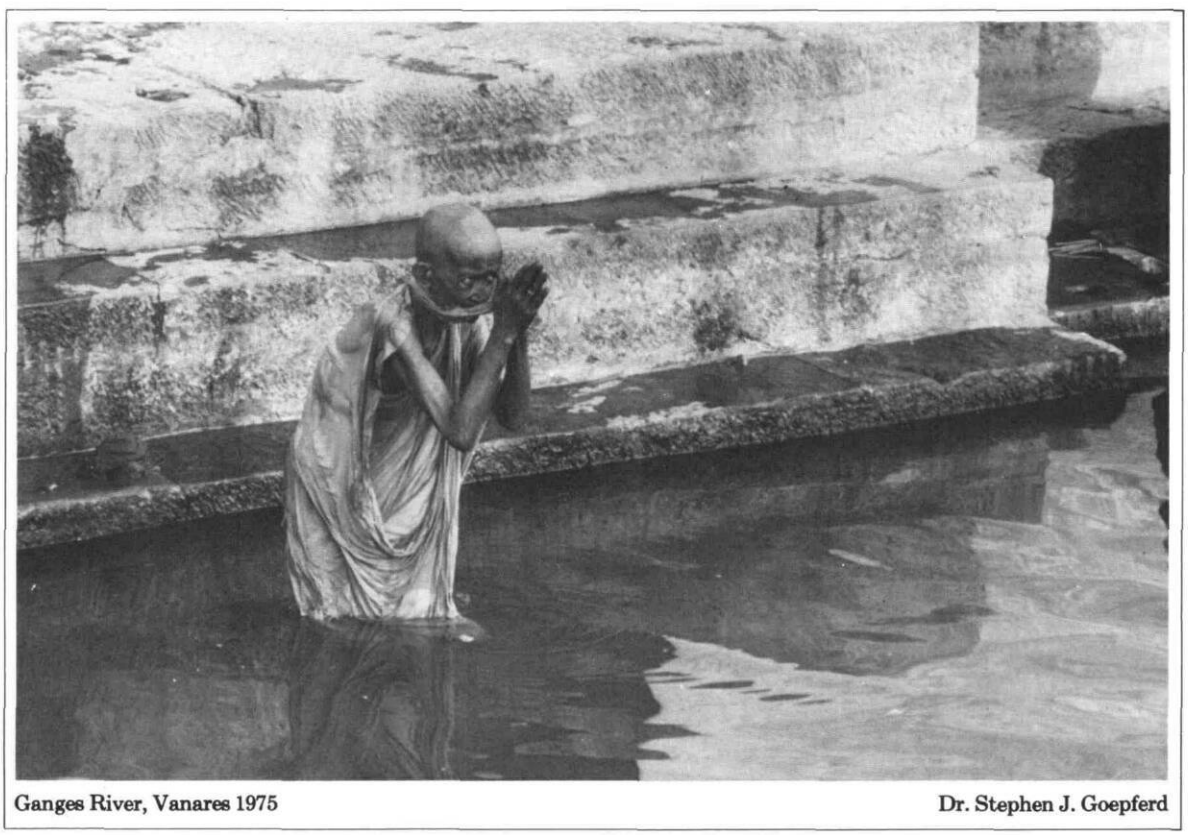

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