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Management of oral findings in a child with an advanced case of dermatomyositis: clinical report

Christopher Hamlin, DDS
Jean E. Shelton, MD

Abstract

Pediatric patients with dermatomyositis present many challenges to the dental practitioner. A pattern of delayed eruption may be observed, and difficulty in exfoliation of the primary dentition due to lack of primary root resorption also may be seen. Primary teeth may require extraction to permit the permanent dentition to erupt at the appropriate time and in proper position. Medications used in controlling this disease can require special consideration in planning treatment procedures.

Dermatomyositis is an inflammatory disease process of obscure etiology which affects muscle and skin throughout the body. The disease often begins as proximal muscle weakness progressing to more general muscular involvement with potential subsequent difficulties in swallowing and respiration. Skin manifestations may precede or follow the muscle finding. Most characteristic among skin findings are heliotrope rash of the upper eyelids and an erythematous eruption in a butterfly pattern across the midface. These findings are in addition to general malaise, weight loss, and occasional periodic fevers. Children with dermatomyositis are likely to develop extensive soft tissue calcification. Prevalence figures for dermatomyositis are not known. The disease is more common in girls than boys. Peak age of onset is 5-10 years.[1]

Diagnosis usually is made on the basis of clinical findings. Laboratory findings are definitive. The following conditions confirm the diagnosis: electromyographic patterns of increased insertional activity; fibrillation potential; a typical myopathic pattern of short, small polyphasic motor units; positive sharp waves; bizarre high frequency repetitive discharges and elevated muscle enzyme levels (especially creatinine phosphokinase). Typical muscle biopsy findings of myositis (necrosis of muscle fibers) usually with evidence of regeneration, phagocytosis, and inflammatory exudate, often perivascular, also are present.[2]

Complications of the illness in children include such extensive calcification of soft tissue that mobility and function of all joints including the spine are restricted and eventually contracted. Arthritis and arthralgia may complicate the course. Ulcerations of the gastrointestinal tract with hemorrhage, another hazard, and involvement of the palatorespiratory muscles, are the two most frequent causes of death. Cardiac muscle involvement occurs in some patients. The disease may remit and progress alternately with some patients developing a chronic, smoldering course.[3]

Treatment of dermatomyositis generally is initiated with corticosteroids. Large doses may be required for extended periods.[4] Other immunosuppressive drugs such as azathioprine and methotrexate also have been used.[5] Physical therapy to maintain mobility is essential to all therapy.

Prognosis in children with dermatomyositis is favorable if the disease is treated early and vigorously. Some children are left with residual joint mobility and contracture problems. However, further disease exacerbations are unlikely.[6] Even with appropriate treatment, a small number of patients will develop the smoldering, chronic disease course exhibited by this patient. In these children the prognosis is more guarded and includes the likelihood of permanent disability in survivors.[7]

Clinical Report

TF is a six and one-half-year-old black female who presented at age five years to Children's Hospital of the King's Daughters in Norfolk, Virginia with complaints of joint pain (especially of the knees and left shoulder), weight loss, rash of the face and hands, and periodic fevers. On parental questioning, it was learned that weight loss, fever, and facial rash had been present approximately one and one-half years prior to her referral to Children's Hospital.

Initially, the diagnosis was made on the basis of proximal muscle weakness with marked muscle atrophy, severe limitations of range of motion, the presence of sub-

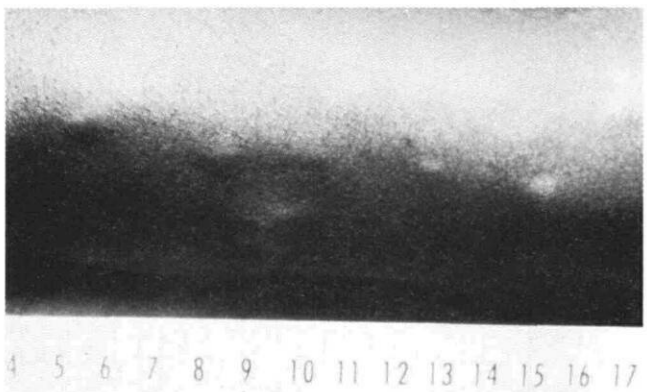


Figure 1. Subcutaneous calcifications were present at multiple sites.

cutaneous calcifications at multiple sites (Figure 1) giving a lumpy appearance to the left lateral thigh, and the characteristic rash over the face and upper eyelids. Creatinine phosphokinase was 39 UI/L (normal 21-214 UI/L). Diagnosis later was substantiated by electromyography showing marked loss of motor action potentials and rare fibrillations at rest. The muscle biopsy demonstrated muscle fiber necrosis, regeneration, atrophy, and periovascular inflammation. These are characteristic findings of dermatomyositis.

Corticosteroid therapy (prednisone) at 2 mg/kg per day was begun. The patient showed initial improvement with remission of systemic symptoms. However, subcutaneous calcification continued to worsen. An exacerbation of clinical symptoms occurred five months later. Methotrexate then was added to therapy at 1 mg/kg weekly IV and corticosteroids were continued. An attempt was made to wean the patient off steroids while on methotrexate. During this period of decreased steroid dosage, the patient's mother related that the child's central incisors had loosened and tightened twice. An exacerbation of disease occurred a third time and steroids were restarted at 2 mg/kg per day in addition to weekly methotrexate. At this point, a dental evaluation was done.

Dental Findings

At age six and one-half the patient had the typical cushingoid-type facial features associated with steroid therapy. She had great difficulty entering the dental chair, as her range of motion was limited severely due to calcinosis of her muscles. She appeared shy and very apprehensive. The chief concern of the child's mother, and reason for referral to our office, was delayed eruption of the permanent teeth.

Dental exam revealed that no permanent teeth had erupted. However, the mandibular permanent central incisors were palpable ectopically to the lingual of the primary central incisors. Radiographs showed a lack of primary root resorption which may have forced the permanent teeth to move into an unfavorable position. There was no history of trauma to any of the primary teeth. However, lingual eruption of the mandibular incisors is estimated to occur about half of the time, and may be considered a normal pattern.[8]

The remainder of the child's oral examination for evidence of calcinosis of the tongue, mouth floor, and buccal mucosa was negative. The gingival tissue was normal and without inflammation. Oral hygiene was fair, and radiographs and visual exam for caries were negative. Temporomandibular joint function was limited somewhat, but no crepitus was noted. Periapical radiographs revealed extensive calcification of the pulp chambers and canals in all primary teeth and a lack of root resorption of the primary incisors (Figures 2-5).

Dental Consideration in Treatment Planning

A plan for extraction of the primary teeth was designed with the child's pediatrician. Usually a permanent tooth will erupt when it has achieved 75% of its root development, and age is not always a reliable criterion.[9] Extraction of the mandibular primary central incisors was necessary to allow normal positioning of the permanent successors.[8] Another concern was that prolonged delay in eruption, due to over-retention of the primary teeth, could cause the permanent teeth to lose some eruption potential. Root end completion of the mandibular permanent central incisor usually is accomplished by age nine.[8] Therefore, it was assumed that the extraction of the primary teeth would promote eruption of the permanent teeth.

Medical Considerations in Treatment

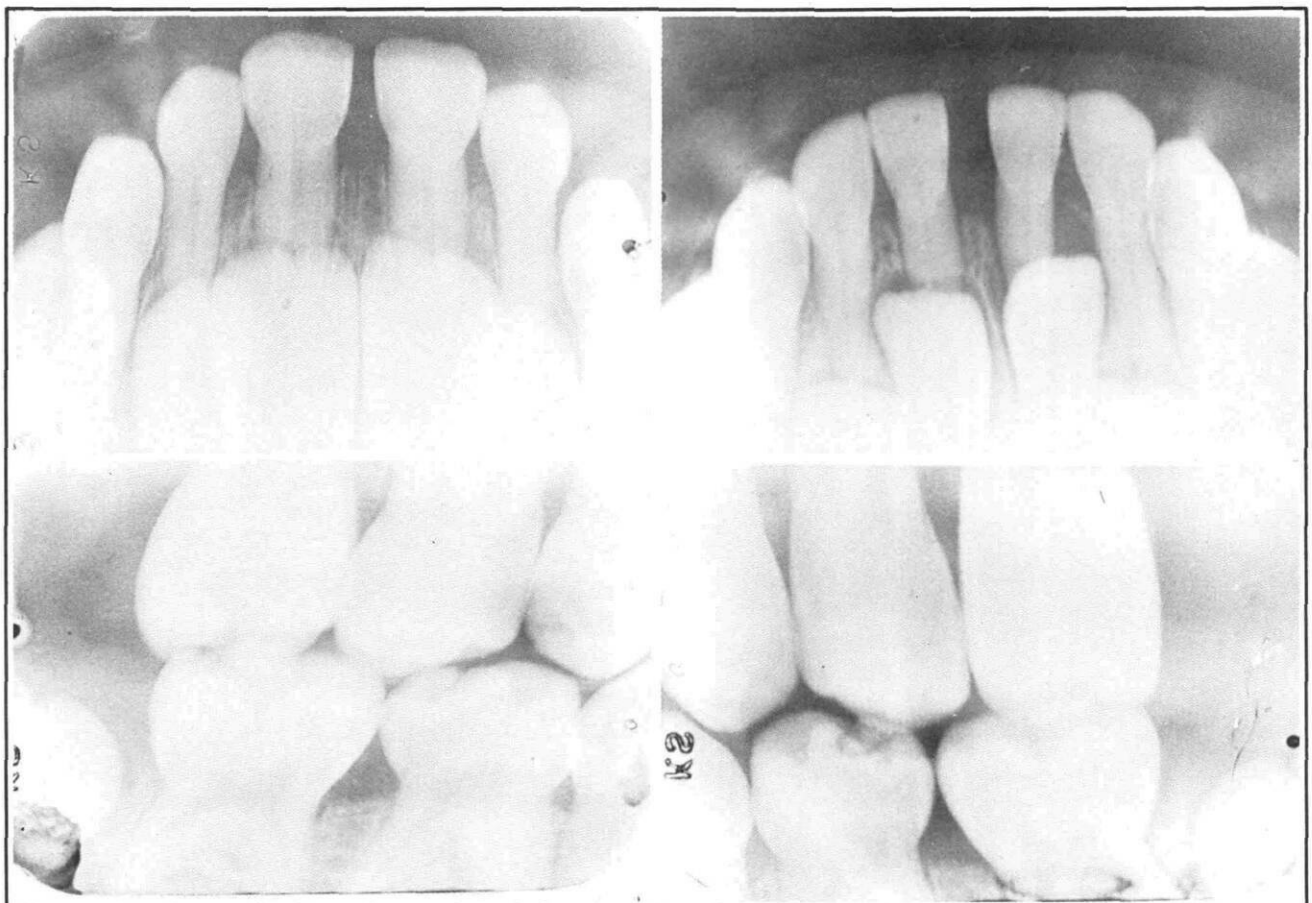
Since the patient was on large doses of steroids (prednisone 2 mg/kg per day and methotrexate 1 mg/kg per week), special precautions were taken prior to oral surgery. With methotrexate administration, a complete CBC and platelet count had to be drawn the day of the proposed extractions to avert any unnecessary postoperative bleeding complications.[5,8] Prophylactic antibiotic therapy was required due to possible introduction of infection and compromised immune function of the patient, possibly induced by chemotherapy.

Treatment

The patient was scheduled for extractions in the pedodontic office. CBC and platelet counts taken at the hospital revealed: HCT 38, HB 12.9, WBC 8,200, normal differential, platelets 729,000, and ESR 28. (Elevated erythrocyte sedimentation rate is seen in conditions such as chronic granulomatous infections, carcinoma, and rheumatoid arthritis. Normal ESR is 1-20).[10] These values were adequate for the planned procedure.

An hour before the appointment the patient was started on 500 mg of oral penicillin and given a preoperative oral sedative of hydroxyzine pamoate 50 mg/10 cc.[a] The ex-

[a] Vistaril suspension; Pfizer, Inc., 235 E. 42nd St., New York, N.Y. 10017



Figures 2-5. Periapical radiographs revealed calcification of the pulp chambers and canals in the primary teeth. There was also evidence of delayed root resorption.

tractions were accomplished with local anesthetic (1.8 cc Xylocaine with epinephrine 1:100,000) without difficulty. The patient tolerated the procedure well. After adequate hemostasis was achieved the patient was dismissed to her mother's care and was placed on oral penicillin 250 mg/5cc every 6 hours for 7 days.

Both teeth were submitted for histologic examination. The biopsy report was essentially normal, except for the finding of extensive calcification evident in the coronal and root portion of the pulp chamber and canal. Also, minimal resorption was observed, confined to the apical third of both roots. The cessation of resorptive activity could correlate with the onset of symptoms at age three and one-half; at that age, approximately one-third of the primary root would have been resorbed.[9,11] This suggests root resorption may have ceased due to the systemic calcification related to the disease process.

This patient has been observed regularly every three months, and radiographically in the lower anterior area approximately every six months (Figure 6). Both mandibular permanent incisors erupted approximately six months after extraction of the primary teeth at age seven. It is anticipated that the primary teeth will continue to be over-retained due to calcific changes. Future radiographic monitoring will determine when the next extractions will be necessary. The criterion used for extraction will depend largely on root maturity of the permanent successors.

Observations

Oral manifestations in patients with dermatomyositis

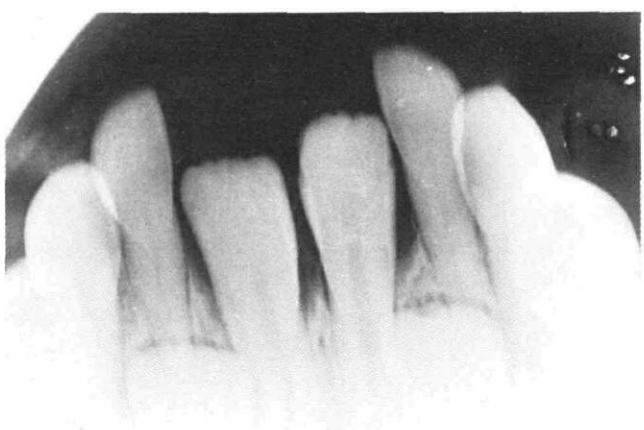


Figure 6. Eruption of the permanent central incisors progressed following removal of the primary central incisors.

rarely have been reported in the literature.[3,12,13] Oral soft tissues including the tongue, mouth floor, salivary glands, buccal mucosa, and muscles of mastication may show evidence of calcium deposits.[3] Anomalies of root formation have been reported once in the past, along with calcific obliteration of the pulp canals in both primary and permanent teeth.[12] Minimal root resorption has been noted in the primary dentition, however management of the situation and long-term care was not reported.[13]

Summary

This report may be unique because the severity of this child's disease may have arrested the normal process of root resorption, requiring the selective extraction of primary teeth at appropriate times.

When treating a child who has undergone this particular regimen of chemotherapy, one has to consider that the extraction of primary teeth may involve careful dental and medical planning. In this particular child, both pediatrician and pedodontist worked closely to manage all stages of the patient's dental treatment.

Dr. Hamlin has a private pediatric dentistry practice, 1806 Hampton Blvd., Norfolk, Va. 23517, and is a staff member at The Children's Hospital of the King's Daughters and DePaul Hospital, Norfolk, Va. Dr. Shelton is an assistant professor of pediatrics, Eastern Virginia Medical School, Department of Pediatrics; Norfolk, Va. Reprint requests should be sent to Dr. Hamlin.

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Quotable Quote

The Anglo-American common law traditionally has been uncomfortable with scientific evidence. Great reliance has been placed on the lay jury system and partisan advocacy to provide justice. The result, however, is often a rather unstudied lack of reality in the legal system. What clearly is accepted scientifically in the outside world often has been rejected inside the courtroom; and the reverse, brazen charlatanism, has been allowed to sway lay juries with procedures and opinions having little or no credibility in scientific circles.

An effort to provide reform in both these areas has been in progress over a long period of years and is based on the so-called Frye rule, which originated in the federal courts. Under this rule, trial court judges are instructed to allow into evidence the results of scientific tests or procedures only when they have been recognized as reliable and useful by the scientific community concerned with the procedure or technique. The trial judges are expected to make affirmative efforts on their own to ascertain the state of acceptance of the procedure in the appropriate scientific community. This can be done by examining the scientific literature and by calling nonpartisan, unbiased experts in the field in question to advise the court concerning the state of the art of the technologic procedures whose results are offered in the court. The judges also may have received information (not yet evidence) from the contending parties regarding the science involved to support or deny admission into formal evidence. In addition, the trial judges must evaluate the competence of the particular witness to have performed the test in question and to offer an expert opinion on the meaning and relevance of the results for the case in hand.

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