DOI: 10.7860/JCDR/2024/68606.19880

Case Report

Dentistry Section

Navigating into the Dental Management of a Child with Morphea: A Case Report

SHRAYANA BHATTACHARYA[1], SUDIPTA KAR[2], DIPANJAN DEBNATH[3], SHABNAM ZAHIR[4]

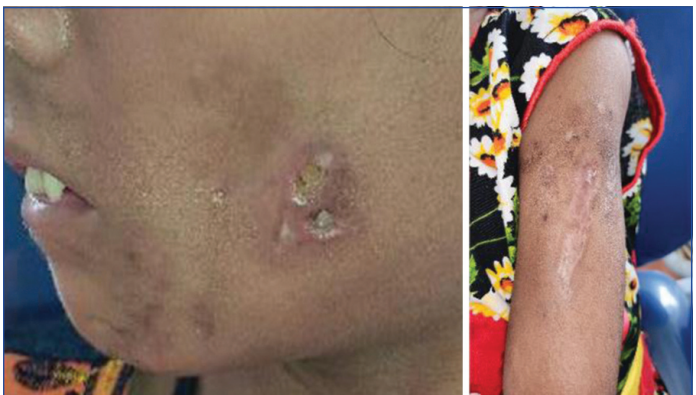
ABSTRACT

Morphea, a morphological variant of Localised Scleroderma (LoS) that typically occurs in children, is an inflammatory disease of connective tissue leading to skin and underlying tissue sclerosis due to increased deposition of collagen. The aetiology of morphea is quite elusive. Morphea, along with other regions also affects oral and perioral tissues; the most common affected implication being facial skin and tongue rigidity. The author reports a case of a 10-year-old female who reported to the Department of Paediatric and Preventive Dentistry for retained maxillary anterior deciduous teeth. The patient also presented with scarring on her face, chest, and arms. Her lips were thin, rigid and partially fixed producing microstomia with decreased mouth opening. Clinical examination, blood tests, Contrast-Enhanced Computed Tomography/High-Resolution Computed Tomography (CECT/HRCT) of thorax, Antinuclear Antibody (ANA) test, Antineutrophilic Cytoplasmic Antibody (ANCA) Test, Anti-dsDNA Test, systemic sclerosis/myositis profile, and histopathological evaluation confirmed the diagnosis of morphea. The management of morphea is multidisciplinary. Hence, the role of a paediatric dental surgeon in recognising the development of disease progression in a patient and avoiding any oral complications is very important.

Keywords: Localised, Paediatric dentistry, Scleroderma

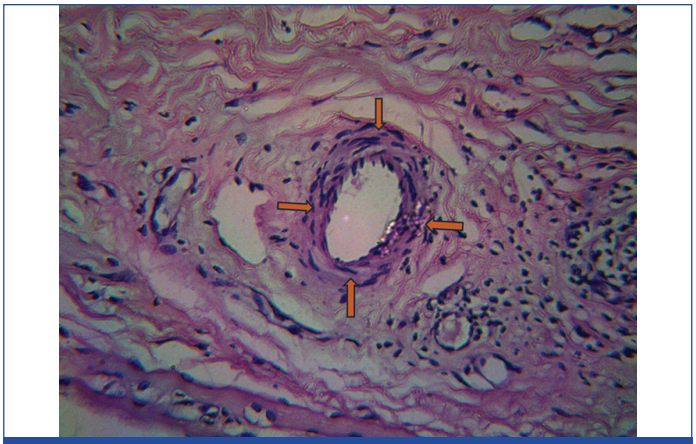
CASE REPORT

A 10-year-old girl reported to the Department of Paediatric and Preventive Dentistry in with the chief complaint of retained milk teeth in the upper front region with mobility. On taking the history, it was found that the patient started showing noticeable skin issues a year ago [Table/Fig-1]. The patient was a known case of Morphea. Histopathological examination revealed a thin epidermis, increased dermo-epidermal pigmentation, slightly increased hyalinisation and Multidirectional/multi-directional collagen formation in the dermis and hypodermis. There was mild lymphohistiocytic infiltration in the dermis, mostly in the perivascular and peri-appendageal regions [Table/Fig-2]. Other investigations- ANCA, ANA, dsDNA, systemic sclerosis profile, myositis profile along with other associated tests confirmed the diagnosis of Morphea. ANA scoring was 4+ implying coarse speckled pattern. ANCA score was negative and the dsDNA level was <10 IU/mL (Normal). CECT of the chest was found to be normal. The patient was undergoing treatment in a medical institution's paediatric and rheumatology departments. The patient was administered prednisolone, methotrexate, and folitrax and was under regular follow-up.



[Table/Fig-1]: Multiple plaque-like lesions on the left-side of the face, chin region and left upper arm respectively.

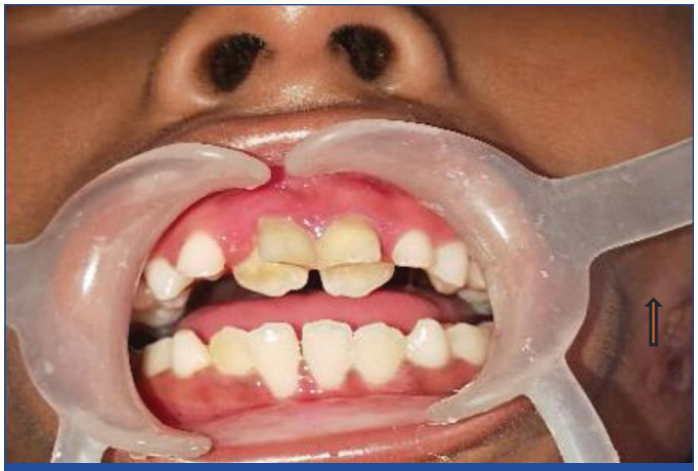
Upon extraoral examination, ill-defined, pigmented, indurated, and adhered lesions greater than 3 cm in size were found, numbering more than four and attached to deep planes. Plaques were visible



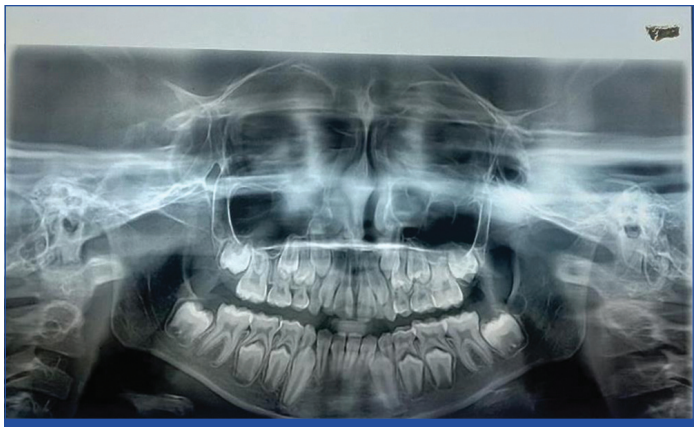
[Table/Fig-2]: Histopathology image showing the lymphohistiocytic infiltration in the perivascular region (marked with arrows). H&E stain, 40x magnification.

mainly on the facial area, torso and extremities [Table/Fig-1]. Intraoral examination revealed retained 51, 61, with palatal eruption of 11, 21 (as per FDI tooth numbering system). The patient had reduced mouth opening (2.5 cm approx.) and perioral rigidity to a certain extent [Table/Fig-3]. The patient's oral hygiene was assessed based on the Oral Hygiene Index- Simplified (OHI-S). The score was 2.6, hence the interpretation was moderate. (OHI-S score 1.3-3 is moderate) [1]. Deep pit and fissures were present in primary molars. However, no evident malocclusion, enamel defects, dental caries or periodontal compromise were observed. The OPG of the patient did not otherwise reveal any rank pathology. On examination, developing canine and premolars were found on all four quadrants, retained 51, 61 as well as erupting 11, 21. No bony abnormality or any developing lesion was observed. The root morphology was normal and no pathological resorption was observed [Table/Fig-4].

At the first scheduled appointment, preventive therapy was advocated. Fluoride gel was applied and the patient was referred for physician's consent for the extraction of 51, 61 [Table/Fig-5]. At the second appointment, extraction of 51, 61 was done, following the physician's consent [Table/Fig-6]. The treatment aimed to improve the oral condition of the patient compared to what it was before. Deep pits and fissures need preventive measures to reduce the chance of

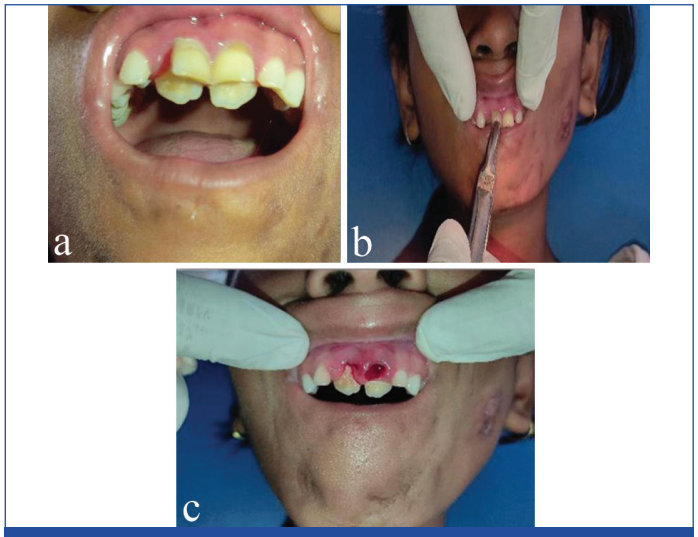


[Table/Fig-3]: Intraorally, the palatal eruption of 11, 21 and retained 51, 61 is seen as well as reduced mouth opening. (indicated by an arrow).



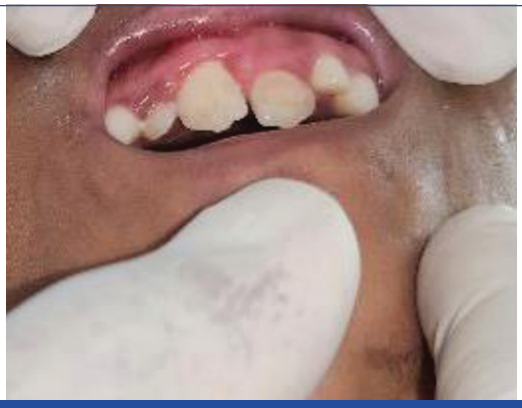
[Table/Fig-4]: OPG of the patient showing normal morphology.

[Table/Fig-5]: Shows preventive therapeutic procedure being performed, during first appointment.



[Table/Fig-6]: a) Pre-op; b) Intra-op; c) Post-op of the extraction procedure.

future decay. Extraction of retained deciduous incisors was done to prevent any chance of developing malocclusion. After a week, the patient was called for follow-up. Healing was checked and found to be satisfactory. Oral prophylaxis was performed [Table/Fig-7].



[Table/Fig-7]: Satisfactory healing after one week was observed and oral prophylaxis was done.

For dry mouth symptoms, viz., dry feeling in the mouth, bad breath, slight difficulty in swallowing, biotene oral rinse was prescribed. The patient and the concerned guardians were motivated to maintain oral hygiene, for the betterment of overall health status. Oral functions like mastication, speech and deglutition appeared to be normal.

DISCUSSION

The LoS, also known as “morphea en plaque”, is a serious condition that inflames and hardens the skin and soft tissue, leading to impaired function, disfiguration, and a diminished quality of life [2]. The Mayo Clinic Criteria, which Peterson LS et al., reported in 1995, first classified morphea into five sub-categories: linear, plaque, bullous, deep, and generalised. [3]. Laxer RM and Zulian F presented the Padua criteria, which identified five subtypes of morphea: circumscribed, generalised, linear, pansclerotic, and mixed. However, bullous morphea and deep morphea were not included in these subtypes. The Padua criteria noticed that the involvement of deep-type lesions could co-exist with circumscribed lesions [4]. In 2017, the European Dermatology Forum proposed a classification system with five subtypes: limited, generalised, linear, deep, and mixed [5]. Morphea classification systems have limitations. Authors specialise either in adult or paediatric medicine, limiting the ability to categorise morphea subtypes. The criteria, highly based on differing perspectives and experiences of morphea, are created primarily by dermatologists or rheumatologists. Determining consistent subsets of morphea patients was challenging due to the lack of unbiased study of demographic or clinical variables in the existing categorisation standards.

Disparate categorisation schemes for morphea have created uncertainty for researchers and physicians, posing a significant obstacle to multisite investigations, which are essential in rare conditions such as morphea, and confusing doctors [6]. LoS classifications are not mutually exclusive, so several subtypes may arise linked with the same patient [7]. Morphea is more prevalent in Caucasian women, with a frequency of 0.4 to 2.7 individuals per 100,000 per year and a female-to-male ratio of 2.4-4.2:1 [8]. Patients with linear scleroderma younger than 18 comprise about 67% of the patient population [9].

Morphea rarely affects the mouth, but a study conducted by Prasada S et al., found that oral manifestations are more common in young patients, particularly in females (78%), which is consistent with the presented case [2]. Another case report by der Vecken V et al., described a woman with morphea who had a pale linear lesion on the right side of her face, extending from her nose to her upper lip, with gum recession between her central and lateral incisors on the same side [10]. Instances of oral manifestations are

infrequent and have seldom been documented in literature. These occurrences may exhibit dental irregularities, deficiencies in root development, root decay, misalignments of the teeth, and restricted mouth openings due to stiffened masticatory muscles. There have also been reports of gingival recessions, loss of alveolar bone density, and complications pertaining to the temporo-mandibular joint (such as arthritis) [8,2,11]. Tang MM et al., also reported a case of progressive gum recession and bone resorption in a 20-year-old female, with histopathological findings showing collagen hyalinisation and perivascular lymphocytic infiltrates, which are comparable to the author's case [12]. A few of the differential diagnoses of morphea can be Scleroderma (systemic sclerosis), Lipodermatosclerosis, Eosinophilic fasciitis, Trauma-induced fat necrosis (intramuscular injections), Chronic graft-versus-host disease, Atrophoderma of Pasini and Pierini [13,14].

Clinical studies suggest various therapeutic strategies for treating LoS, including vitamin D, D-penicillamine, phenytoin, cyclosporine, corticosteroids, methotrexate, interferon, and psoralen-UVA photochemotherapy [15,16]. Because of its anti-inflammatory and anti-proliferative properties, methotrexate is frequently used as a disease-modifying medication. Folic acid supplements are necessary for methotrexate patients to avoid side-effects. Topical calcineurin inhibitors have proven effective in reducing skin thickening, hyperpigmentation, hardness, erythema, and contraction [17]. Relatively positive results have been obtained by combining topical treatment with vitamin D analogue and phototherapy with low-dose UVA1 and 0.005% calcipotriol ointment. Mycophenolate mofetil has been found effective in treating Juvenile Localised Scleroderma (JLS) that is resistant to treatment [16].

Research on mice models of systemic sclerosis has demonstrated that Janus Kinase (JAK) inhibitors are very successful in blocking the fibrotic pathway [18]. The TGF-beta-mediated route of STAT protein activation is successfully blocked to do this. Moreover, tofacitinib, a JAK 1/3 inhibitor, has been reported to improve skin sclerosis in case report of the treatment of morphea and SSc [19]. In patients with SSc and morphea, tofacitinib has been the most extensively studied JAK inhibitor to date, demonstrating its efficacy and dependability as a therapeutic choice [19]. The fusion protein called abatacept binds to CD80 and CD86 and prevents CD28 from interacting with the cell, hence suppressing T-cell activation [20]. Reconstructive treatment is suggested as a viable option for lesions that have stopped showing signs of disease activity. The main aim of this treatment is to enhance the cosmetic appearance of the affected area. For individuals experiencing craniofacial abnormalities or localised tissue atrophy, structural fat grafting is the best treatment option. Because adipose tissue has a high concentration of stem cells, structural fat grafting is a useful technique for generating new vascularisation and substantive structural modification [16]. Although, it needs some improvement, the Localised Scleroderma Cutaneous Assessment instrument (LoSCAT) is a clinically promising instrument for evaluating morphea. It is the only clinical measure for morphea created per the strict guidelines of Outcome Measures in Rheumatologic Clinical Trials (OMERACT) and the only clinical scoring system for morphea that takes into account both activity and damage [6]. High-frequency ultrasound is a non-invasive tool for diagnosing morphea and evaluating treatment response, with 100% sensitivity and 98.8% specificity [21]. Infrared thermography, a technique measuring infrared heat, is useful for detecting active lesions in children with morphea but has a sensitivity of 92% and a specificity of 68% [22]. Dermoscopy is an affordable, non-invasive, and easily accessible method that can quickly diagnose skin lesions even in outpatient settings. It allows for the observation of pigmented skin structures and also aids in identifying, differentiating, monitoring, and predicting the prognosis of several inflammatory dermatoses [23].

Most patients diagnosed with JLS have a positive prognosis and are able to achieve remission. However, the disease may take a longer course for some patients. A study has shown that approximately 12.5% of patients, all of whom had the linear variant, experienced continued disease activity even after a decade of follow-up. Longer disease activity was associated with a delay in the start of therapy. Pansclerotic, linear, and mixed types of morphea are more likely to cause functional damage. Should treatment be discontinued, disease reactivation is most commonly seen within two years, making it imperative to ensure close monitoring during this period [24].

Odontological management is a personalised approach that is determined by the dentist based on the patient's needs. Encouraging patients to practice good dental hygiene is essential to preventing gingival recessions. In cases of xerostomia, sialogogues such as pilocarpine must be prescribed. Additionally, patients can consume foods that stimulate salivation like menthol candy to keep their oral cavity moist [25]. To preserve range of motion and prevent or postpone contractures, patients with restricted mouth opening and range of motion need to get routine physical and occupational treatment. Bilateral commissurotomy may be suggested in exceptional circumstances to widen the patient's mouth. To maintain proper dental health care, it is very advised that people with LoS see their dentist frequently. To prevent any oral issues for our patient, which is crucial for their well-being, we concentrated on encouraging good dental hygiene and follow-up.

CONCLUSION(S)

Dental surgeons and medical doctors must identify cases of morphea that are limited to the oral mucosa. They must possess knowledge of the clinical presentation of morphea and conduct a thorough examination of its morphology, especially if the oral mucosa has solitary white streaks or plaques. It is essential to differentiate correctly and monitor closely to avoid serious local issues. Quite unfortunately, dermatologists or rheumatologists have diagnosed most patients with intraoral lesions at an advanced stage instead of dentists. Consequently, it is critical to identify and treat skin and oral mucosa involvement in LoS with particular attention.

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PARTICULARS OF CONTRIBUTORS:
1. Postgraduate Trainee, Department of Paediatric and Preventive Dentistry, Guru Nanak Institute of Dental Sciences and Research, Hooghly, West Bengal, India.
2. Professor, Department of Paediatric and Preventive Dentistry, Guru Nanak Institute of Dental Sciences and Research, Hooghly, West Bengal, India.
3. Pedodontist, Private Practioner, Sodepur, West Bengal, India.
4. Professor and Head, Department of Paediatric and Preventive Dentistry, Guru Nanak Institute of Dental Sciences and Research, Hooghly, West Bengal, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Shrayana Bhattacharya,
2A/7, Uttarpara Housing Estate, 88B, G.T. Road, Bhadrakali,
Hooghly-712232, West Bengal, India.
E-mail: bshrayana09@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Dec 07, 2023
- Manual Googling: Feb 02, 2024
- iThenticate Software: Jul 18, 2024 (10%)

ETYMOLOGY: Author Origin

EMENDATIONS: 9

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Dec 05, 2023**
Date of Peer Review: **Jan 30, 2024**
Date of Acceptance: **Jul 20, 2024**
Date of Publishing: **Sep 01, 2024**