ISSN: 2581-5989
PubMed - National Library of Medicine - ID: 101738774

International Journal of Dental Science and Innovative Research (IJDSIR)

IJDSIR : Dental Publication Service

Available Online at: www.ijdsir.com

Volume – 6, Issue – 1, January - 2023, Page No. : 175 - 179

Alveolar cleft bone grafting in a patient of Stickler Syndrome - A case report

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Citation of this Article: Dr. Soubhik Pakhira, Dr. Sreshthanu Saha, Dr. Anirban Raha, Dr. Arkajit Goswami, "Alveolar cleft bone grafting in a patient of Stickler Syndrome - A case report", IJDSIR- January - 2023, Volume –6, Issue - 1, P. No.175– 179.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

The rationale: Stickler syndrome is a rare autosomal dominant disorder comprising of loss of vision, degenerative changes of joints along with cleft palate, hypoplastic midface, and other craniofacial anomalies.

Patient concerns: Patient had kypho-scoliosis which was major concern for airway management during surgery.

Diagnosis: Already diagnosed as type 1 Stickler syndrome at childhood, Cone beam computed Tomography (CBCT) used for obtaining accurate dimension of the alveolar cleft defect by means of volume rendering. Prominent kypho-scoliosis of cervical, thoracic and lumbar vertebrae were evident from the routine radiographs.

Treatment: Secondary alveolar bone grafting was performed with closure of concomitant oro-nasal fistula.

Outcomes: Patient was post-operatively followed up at 6 months and 1 year interval up to 4 years with sufficient amount of alveolar bone formation.

Take – away lessons: Anticipation of difficult airway due limited neck mobility and kyphoscoliosis imparted well-planned intubation and extubation, which prevented intraoperative as well as post-op complications.

Keywords: Stickler syndrome, alveolar cleft, secondary alveolar bone graft, oro-nasal fistula, iliac-crest, autologo us marrow cancellous bone graft.

Introduction

Stickler syndrome was first described in 1967 as hereditary progressive arthro-ophthalmopathy [1]. Since then, several case reports have been published in various specialties. But there are not many case reports regarding surgical management of secondary cleft deformities in Stickler syndrome patients. Operating on such patients is challenging due to anticipated difficult intubation. A patient of type 1 Stickler syndrome having alveolar cleft defect with concomitant oro nasal fistula was treated successfully by alveolar bone grafting and followed up for 4 years.

Case report

A 12 years old male patient of type 1 Stickler Syndrome with alveolar cleft defect was referred to the Department of Oral and Maxillofacial Surgery for alveolar bone grafting from Department of Orthodontics and Dentofacial Orthopaedics. In spite of primary cleft lip and palate repair in early childhood, nasal regurgitation of oral fluids from oro-nasal fistula was chief complaint from the patient. Patient had alveolar cleft defect with small oro-nasal fistula on labial side in between maxillary right lateral and canine teeth. Other than the cleft defect he had hypoplastic midface with severe skeletal crossbite, congenital blindness due to vitreo-retinal degeneration of left eye, severe cervical scoliosis with prominent kypho-scoliosis of other vertebrae. He was undergoing palatal expansion treatment for past 2 years before presenting to the Dept. of oral and Maxillofacial Surgery. The alveolar cleft defect had volume of 2.56 mm^3 as rendered from pre-operative cone beam computed Tomography (CBCT). Mouth opening of the patient was adequate with U-shaped palatal vault. Postero-anterior thoracic radiograph showed marked double scoliotic curve of thoracic vertebrae, whereas Antero -posterior pelvis radiograph showed increased space in Sacro-iliac joints. Marked kyphosis was determined from the lateral thoracic view also. Other routine investigations were within their normal ranges.

From anaesthetic point of view, difficult airway devices were made available as a part of our anaesthetic preparation. The endotracheal intubation was managed with external manipulation via cricoid pressure and posterior displacement of larynx. Gingival mucoperioseteal flaps were raised as per Åbyholm's [2] technique. Conventional turnover flap derived from the lining of the fistula was utilised to make nasal layer. Bone trephine was used during the bone graft harvesting from anterior iliac crest. The bone grafts were mixed with PRF membrane (Platelet Rich Fibrin) and placed in the alveolar process with tension closures. Extubation and recovery was uneventful and the patient was discharged at 5th post-operative day.

Post-operative soft tissue healing was uneventful without any wound dehiscence and recurrence of fistula. Orthodontic treatment was resumed after 6 weeks. Good amount of alveolar bone formation was evident from occlusal as well as periapical radiographs on periodic follow-up at 6 months and 1 year interval up to 4 years.

Discussion

Stickler syndrome has estimated incidence of 1 in 10,000, where around 20% of the patients have cleft palate[3]. It has been reported as most common syndrome associated with Pierre Robin Sequence (PRS). Individuals usually present physical abnormalities on airway examination and cardiac manifestations related to Stickler syndrome could be an issue of concern[4]. A retrospective study was conducted between 1997-2004

by Lee and Haywards[5] to evaluate the group of cleft palate children with Stickler syndrome in a children's hospital. In their findings, all patients had cleft palate and most common non-craniofacial abnormalities were hearing impairment, musculoskeletal abnormalities and refractive error. Later on in 2005, Rose et al[6] published clinical characteristics and diagnostic criteria of Stickler syndrome based on their study upon 90 patients of 38 families. Their set of diagnostic criteria made the process of identification of the syndrome simpler amidst the involvement of multiple complex genes. Patients with micrognathia in Stickler syndrome often present with considerable problems regarding airway management during anaesthesia. Most of the cases even have incidences of failed intubations.[7] The incidences of airway complications during anaesthesia as well as obstructive sleep apnoea (OSA)[8] attributed to skeletal abnormalities[9] as well as the malformations in head and neck region[10]. Regarding the secondary cleft surgeries in syndromic patient literary support is still scanty. Case reports of alveolar cleft bone grafting in patients of Stickler syndrome is no such exception.

Conclusion

In the case report we tried to enlighten the Gray areas of conducting major maxillofacial surgeries in syndromic patients. We were fortunate enough that major untoward situations of airway embarrassment during anaesthesia were prevented by well-executed planning.

Acknowledgement

Dr. Goutam Guha, Mch, Professor and Head, Dept. of Plastic and Reconstructive Surgery, IPGMER and SSKM Hospital, Kolkata.

References

1. Stickler GB, Pugh DG. Hereditary progressive arthro ophthalmopathy. Additional observation on vertebral anomalies,a hearing defect and a report of a similar case. Mayo Clin Proc. 1967.

2. Frank E. Åbyholm, Olav Berg land& Gunvor Semb: Secondary Bone Grafting of Alveolar Clefts: A Surgical/Orthodontic Treatment Enabling a Non-prosthodontic Rehabilitation in Cleft Lip and Palate Patients, Scand J Plast Reconstr Surg 15: 127-140, 1981.

3. Jackson, Oksana A. Alison E. Kaye, Alfred Lee, Nancy Minugh-Purvis, Marilyn A. Cohen, Cynthia B. Solot et al: Orofacial Manifestations of Stickler Syndrome, Ann Plast Surg 2020;84: 665–671

4. Joana Varandas et al: Airway Management in Stickler Syndrome Patient: A Case Report, Journal of Anaesthesia and Clinical Research,2017

5. Lee KH, Hayward P. Retrospective review of stickler syndrome patients with cleft palate 1997-2004. ANZ Journal of Surgery. 2008 Sep;78(9):764-6.

6. Rose PS, Levy HP, Liberfarb RM, Davis J, Szymko-Bennett Y, Rubin BI et al. Stickler syndrome: clinical characteristics and diagnostic criteria. American journal of medical genetics Part A. 2005 Oct 15;138(3):199-207.

7. Küçükyavuz Z, Özkaynak Ö, Tüzüner AM, Kişnişçi R. Difficulties in anesthetic management of patients with micrognathia: report of a patient with Stickler syndrome. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology, and Endodontology. 2006 Dec 1;102(6):e33-6.

8. Jeon SY, Kwon OE, Jang JW, Kang SY, Min JY, Kim SW. Surgical management in a severe OSA patient diagnosed with Stickler syndrome. Auris Nasus Larynx. 2021 Oct 1;48(5):1031-4.

9. McArthur N, Rehm A, Shenker N, Richards AJ, McNinch AM, Poulson AV, Tanner J, Snead MP, Bear

croft PW. Stickler syndrome in children: a radiological review. Clinical Radiology. 2018 Jul 1;73(7):678-e13.

10. Lucarini JW, Liberfarb RM, Eavey RD. Otolaryngological manifestations of the Stickler syndrome. International journal of PediatricOtorhino laryngology. 1987 Dec 1;14(2-3):215-22.

Figure Legends

Fig 1: A. External view showing severe degree of scoliosis, degenerated left eye. B. Intra oral view showing oro nasal fistula and alveolar cleft. C. 3-D simulation image of the cleft defect. D. 3-D reconstructed image of face in CBCT showing the cleft defect. E. Occlusal radiograph showing palatal expander in place. F. Postero-anterior view of chest showing double curvature of thoracic vertebrae. G. Lateral cephalogram showing midface hypoplasia. H. Lateral view of thorax showing severe form of kyphosis. G. Antero-posterior and lateral view of pelvis showing increased space between Sacro- iliac joint.

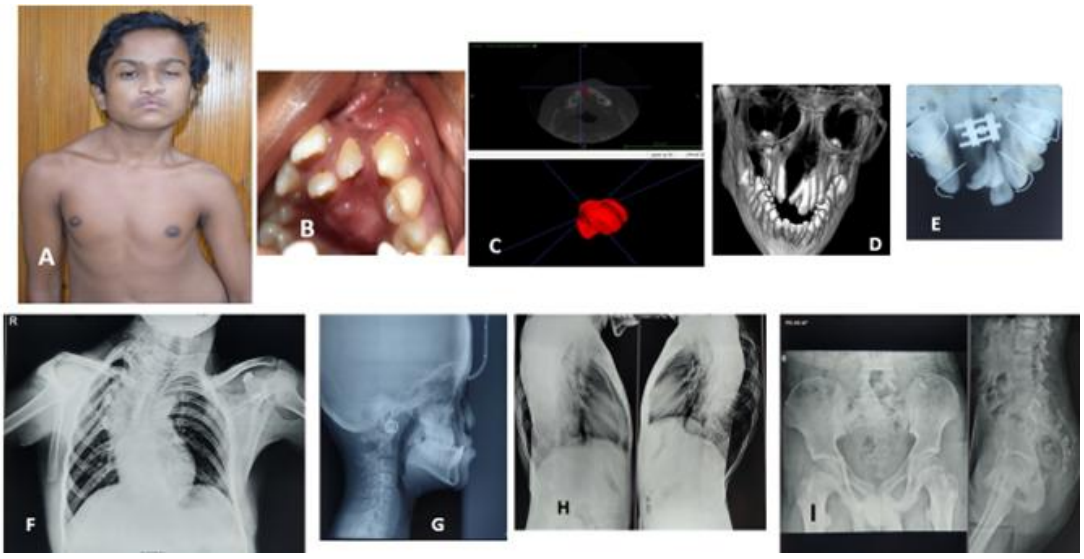

Fig 2: A. Marking of the incision of gingival mucoperiosteal flap. B. Exposure of the alveolar cleft defect with closure of the nasal layer. C. Exposure of the anterior iliac crest. D. Cancellous bone graft. E. Filling of the defect with cancellous bone graft mixed with PRF. G. Closure of the oral layer.

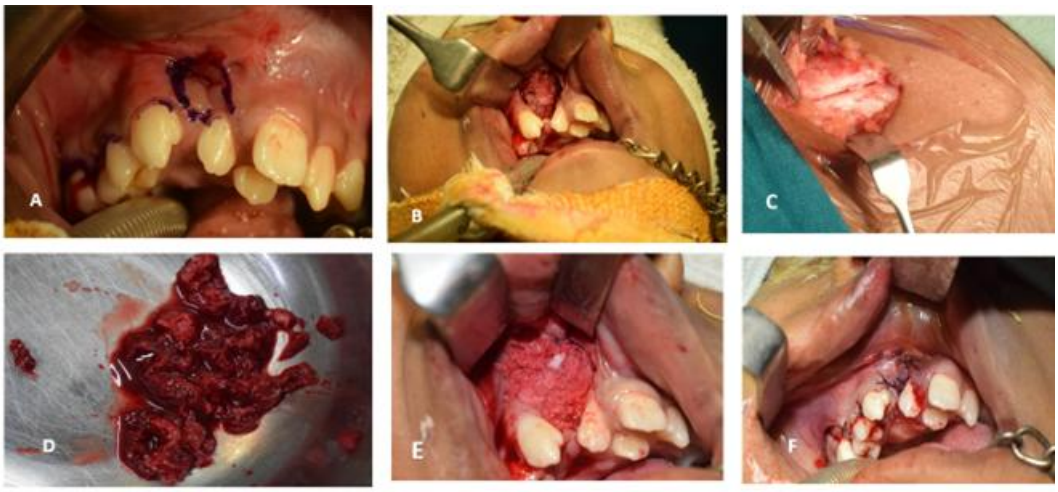

Fig 3: A. Post- operative intraoral view after 1 year. B. Post -operative CBCT image showing bone formation in the defect.