

Management of Salivary Fistula in Goldenhar Syndrome - A Case Report

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Abstract: Objectives: To report a case of salivary fistula in Goldenhar Syndrome and discuss its management. Methods: A 14 year old girl presented with salivary fistula since birth. A diagnosis of Goldenhar syndrome with salivary fistula was made using clinical findings and X-Ray sialography. Results: Sialography revealed accessory parotid gland with fistulous tract draining on the left cheek. Intraoral transposition of the fistulous tract was done. Conclusion: Intraoral transposition of fistulous tract is an efficient way of management of congenital salivary fistula with accessory parotid gland in Goldenhar syndrome.

Keywords: Goldenhar syndrome, Congenital salivary fistula, oral intratransposition, accessory parotid gland

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I. Introduction

Salivary fistula can be a manifestation of various diseases ranging from congenital abnormalitis, infections, malignancy, post traumatic or post surgical. Goldenhar syndrome presents as congenital disorder with abnormalities of the ocular, auricular and vertebral system. Congenital salivary fistula along with pre auricular appendages has been reported as part of the Goldenhar Syndrome¹

II. Case Report

A 14 year old girl presented with complaints of discharge from a small opening on her left cheek which was present since birth as told by her parents. She developed excoriation around the opening for 2 years. Discharge was clear, watery in consistency and occurred mainly during meals. She also had two swellings of size 2.5x2.5 cms each in front of the left tragus. There was no history of trauma, surgery, fever or purulent discharge from the fistula. Patient did not suffer from tuberculosis neither had any contact with tubercular patient.

Examination revealed a fistulous opening about 3cms lateral to the left commissure. Oral examination revealed a normal Stenson duct opening opposite the second molar tooth. Parotid gland was normal. Two pre auricular swellings were present as described earlier. No other abnormality present. Fig 1.



Figure 1 - Preoperative picture showing salivary fistula with adjoining excoriation of skin

Investigation by a X-Ray sialogram revealed fistulous tract draining a accessory parotid gland. The position of the left parotid gland, stensons duct and its opening was normal. No connection was noted between the two systems. Fig 2

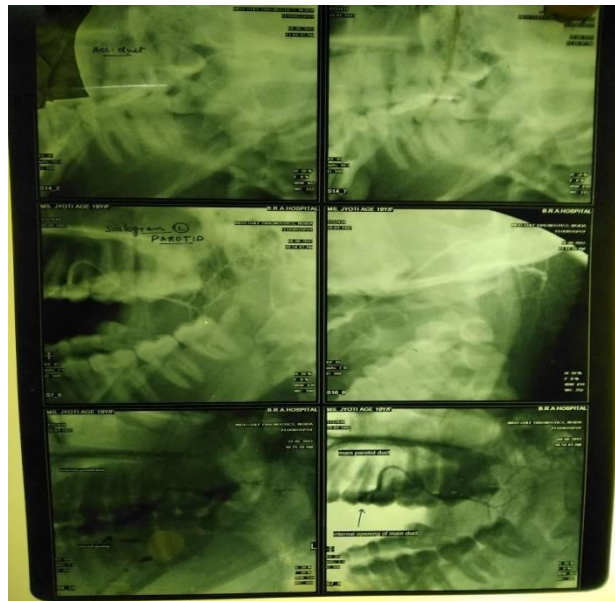


Figure 2 - sialogram showing normal and accessory parotid duct

Patient was taken up for surgery with the plan of reimplanting the duct in the cheek. The fistulous tract was cannulated by using prolene thread as a stillete. The tract was dissced free. Tract was pulled in by making an oblique tunnel in the cheek and duct to mucosa anastomosis was done Fig 3 . The patency and leak was checked by pushing in saline from the neo opening. Fig 4.



Figure 3 - Intraoperative picture of tract being pulled through an oblique channel in cheek

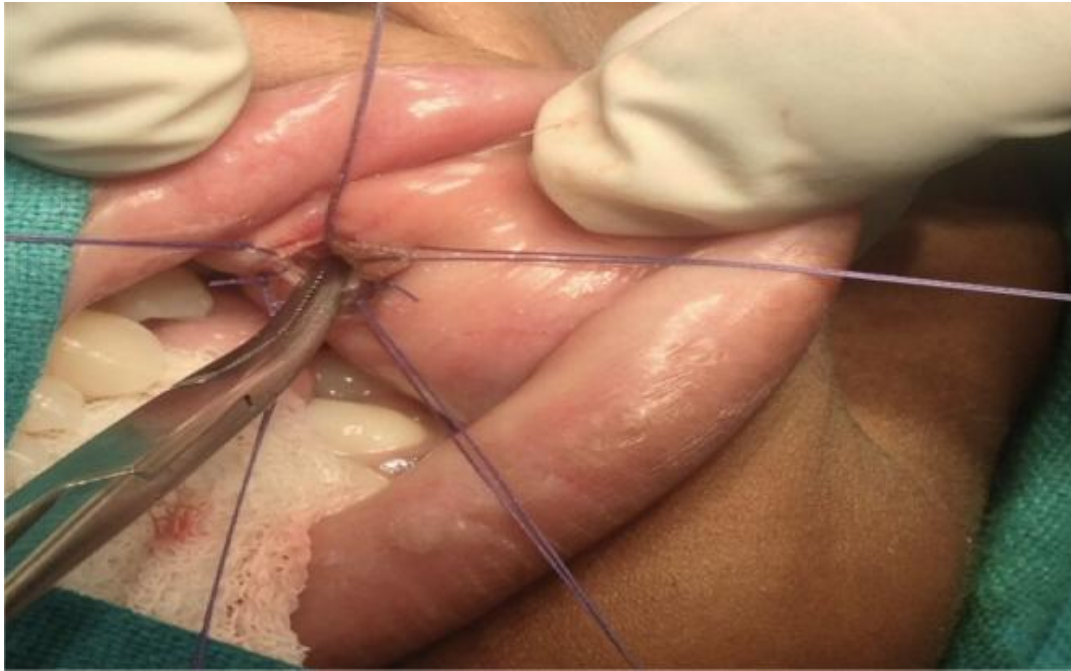


Figure 4 - Completed duct to mucosa anastomosis

Postoperatively dressings were opened after 48 hours without soakage. No discharge from the cheek was seen after surgery. Fig 5.



Figure 5 - Postoperative picture

III. Discussion

Goldenhar syndrome (oculoauriculovertebral dysplasia) is a rare congenital anomaly involving the first and second branchial arches.² First described by French ophthalmologist Maurice Goldenhar in 1952, its incidence has been described between 1:3500 to 1:5600, with a male to female ratio of 3:2. It can have a myriad of phenotypic presentation involving auricles (pre auricular swellings, microtia, hearing loss) 83%, facial (mandibular hypoplasia, hemifacial microsomia, salivary fistula, cleft lip and palate) 65-75%, ocular (microphthalmia, epibulbar dermoids etc.) 66%.^{3,4} Other features include tracheoesophageal fistula, hypoplastic vagina, inguinal and umbilical hernias. The OAVS decidedly has the clinical spectrum of microtia, which also includes preauricular tags and pits, as one of its defining features.^{5,6}

Salivary fistula is a cause of great distress to the patient both functionally as well as cosmetically. It occurs due to the presence of an ectopic accessory parotid system (EAPS). The accessory parotid is present in about 21% of

the normal population.⁶ The pathology behind the ectopic system is the separation of a part of the parotid tissue proper during its lateral migration. Multiple methods of managing such as superficial parotidectomy along with excision of fistulous tract (intraoral/extraoral approach), intraoral transposition of the tract and chemical cauterization using bouthinm toxin/trichloroacetic acid.⁸ Another method is ligation of the fistulous tract with spontaneous regression of the accessory parotid gland.

However there is no large scale study comparing the methods due to paucity of cases. Reported literature comprises of short case series and a few case reports only. Transposition of the fistulous tract is a simple procedure as described and it avoids the morbidity and radicalness of parotidectomy. Chemical cauterization warrants the need of repeated procedures and has a risk of formation of sialoceles and recurrence.⁹ Oral transposition scores over them in terms of ease of surgery, complete in single setting and use of ethicon straight needle as improvised microneedle cautery system obviates the need for high end expensive instruments.

IV. Conclusion

Goldenhar syndrome with salivary fistula is a rare case with only about 16 cases reported previously. Identifying salivary fistula as part of syndrome helps in assessing the other facets of the disease which could be missed otherwise. There is no evidence to support any of the described methods for management of salivary fistula in this syndrome, however intraoral transposition seems to be the easiest complete procedure associated with least morbidity.

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