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Case Report | Open Access

Goldenhar syndrome: An oculo-auriculo-mandible syndrome was reported

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Introduction

In A 2-month-old girl with suffer from eye-facial problems was referred to our clinic. The baby was full-term [1,2] delivery without complications. She had limbal dermoid of left eye with 3mm diameter and both eyes with exotropic strabismus 30 degree. The left side of face is malformed included with external ear malformed, microtia or atresia. The mandibular hypoplasia on left side, on right side is nearly normal and vertebral [2-4] is also nomal. For management of facial features' baby was referred to pediatric doctor [2] and regular follow-up was advised (Figure 1). Clinical findings showed in Table 1. Causes: Up to now the cause of Goldenhar syndrome is unknown. However, it multifactors were considered. Among of factors, a genetic component, which would suspect for certain familial cases. The Goldenhar syndrome in the children of Gulf War veterans has been suspected, but the difference was not identified [5]. This syndrome may be related with the use of drugs [2] such as thalidomide, retinoic acid, tamoxifen, and cocaine by the pregnant mother. The pregnant mother with diabetes, rubella, and [2] influenza have also been suggested as risk factors [6]. This case is not familial relationship of genetic problem.as well as drugs and maternal diseases were confirmed.

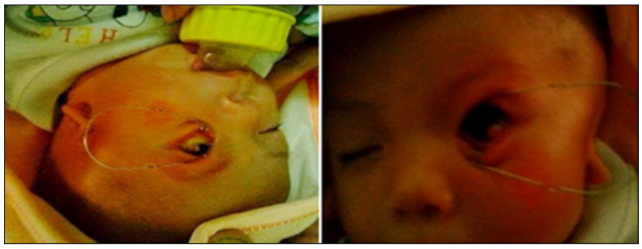

Figure 1: Clinical findings.

Table 1: Clinical findings.

S.no	Clinical findings	Lift Side	Right Side
1	Hemicranial deformities	Yes	No
2	Hemifacial deformities		No
3	Microtia or atresia will lead to deaf-ness later		No
4	Round limbal dermoid	3 mm diameter	No
5	Exptropic strabismus 30 degree (may result amplyopia,blindness later)	OU=both eyes	
6	Jaw deformity may result in a restrictive diet and malnutrition later,	+/-	+/-
7	The patient may suffer from light obstructive sleep apnea due to airway abnormalities	+/-	+/-

Epidemiology

This syndrome occurs from 1 in 3,500 and 1 in 26,000 live births. [3] The ratio of male-to-female is 3:2 [7].

Diagnosis

We diagnose this case with based on clinical signs. General features include unilateral facial asymmetry. Ocular signs include limbal dermoids [4] and strabismus may be develop amplyopia later. Microtia, partial to complete atresia of external acoustic meatus, preauricular skins, and deafness were seen. Microtia can affect one ear (unilaterally) or both ears (bilaterally). Microtia occurs in every 1 out of 6,000 to 12,000 births. The right ear is more commonly affected and microtia is often accompanied by atresia but our case is left ear. Abnormal sign of skeletal is mandibular deformities. Cardiac defects and renal abnormalities were not detected [1,8].

Treatment

This case will be regular checkup her ears for hearing as well as esthetics, eyes for seeing.as she grows suitable. Surgical treatment can help the child to develop normally e.g. jaw distraction/bone grafts, resection of ocular dermoid, repairing some deformities [9,10]. When grow up, some patients with Goldenhar syndrome will need of hearing aids or eyeglasses [1,8]. Stem cell grafting has been usefully on eye dermoids. These tissues that grow on the eye are follow-up for long time [1]. The treatment of the disease varies with age development and is [2] mainly cosmetic with safe cases. Surgical reconstruction of the external [2] ear may be done the age of 6 to 8 years [2]. Jaw reconstruction surgeries can be done in the early teens; epibulbar [2] dermoids should be surgically excised [7] Eyes and ears [2] can be corrected by plastic surgery [7]. Prognosis of the disease is good in [2] uncomplicated cases without any systemic associations. Successful [2] treatment requires a many concerned fields of otolaryngology, ophthalmology, pediatry, dermatology, orthopedy.

a) Palliative care: The role of palliative care is to relieve the suffering both of patient with disability and their families. Treatment included physical, psychosocial, and spiritual symptoms [9].

b) Quality of life: The period of follow-up of the patient is continuing for long time coming in the participation all activities with the same age child when the patient will grow up. All of these as a part of palliative care can be a reason make better quality of life for disable child as well as his/her family. Despite of a developing country the quality of life index of Vietnam is ranged 60 in 2018 [10].

Conclusion

This case is contributed in diversifying clinical signs of Goldenhar syndrome especially eye defected in order to follow- up and treatment as suitably as possible. Quality of life on congenital defected patient is necessary for both patient as well as his/her family.

References

1. https://en.wikipedia.org/wiki/Goldenhar_syndrome.
2. M Goldenhar (1952) Associations malformatives de loeiletdeloreille, en particulier le syndrome dermoïde epibulbaire-appendices auriculaires-fistula auris congenitaetses relations avecla dysostose mandibulo-faciale. Journal de génétique humaine, Genève1: pp. 243-282.
3. Touliatou V, Fryssira H, Mavrou A, Kanavakis E, Kitsiou-Tzeli S, et al. (2006) Clinical manifestations in 17 Greek patients with Goldenhar syndrome. Genet Couns17(3): 359-370.
4. ZakaurRab Z, Mittal S (2007) Optic Nerve Head Drusen in Goldenhar Syndrome. JK Science 9(1): 33-34.
5. Araneta MR, Moore CA, Olney RS, Edmonds LD, Karcher JA, et al. (1997) Goldenhar syndrome among infants born in military hospitals to Gulf War veterans. Teratology 56(4): 244-251.
6. Kapur R, Kapur R, Sheikh S, Jindal S, Kulkarni S, et al. (2008) Hemifacial microsomia: A case report. J Indian Soc Pedod Prev Dent 26: 34-40.
7. Kulkarni V, Shah MD, Parikh A (1985) Goldenhar syndrome: A case report. J Postgrad Med 31: 177-179.
8. Sudarshan P Gaurkar, Khushboo D Gupta, Kirti S Parmar, Bela J Shah. Goldenhar Syndrome: A Report of 3 Cases. Indian Journal of Dermatology.
9. Robin B Rome, Hillary H Luminais, RN, Deborah ABourgeois, Christopher M Blais, et al. (2011) The Role of Palliative Care at the End of Life. Ochsner Journal. Winter 11(4): 348-352.
10. https://www.numbeo.com/quality-of-life/rankings_by_country.jsp.

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