ISSN : 0974 - 7532 Volume 8 Issue 10

Research & Reviews in

BioSciences

Regular Paper

RRBS, 8(10), 2014 [382-385]

Report of bilateral microtia in 2 successive female siblings in Fallujah general hospital

Samira Telfah Alaani*, Abdulqadir Abdulkareem Alrawi

Fallujah General Hospital, Fallujah, Anbar Province, (IRAQ)

E-mail : samiraalaani@hotmail.com; abd69k@yahoo.com

ABSTRACT

Microtia is a congenital anomaly, characterized by a small, abnormally shaped auricle (pinna). It is usually accompanied by a narrow, blocked or absent ear canal. Microtia can occur as the only clinical abnormality or as part of a syndrome. The estimated incidence of microtia is 1 in 5,000 to 1 in 20,000 births, and it is more common in males. Microtia can have a genetic or environmental predisposition.

Here we reported afamily with 2 succesive deliveries of female infants within 12 months period, with bilateral microtia with absence of any of the known risk factors & teratogens, in addition to the absence of previous family history of any kind of birth anomalies.

KEYWORDS

Microtia;
Fallujah general hospital.

INTRODUCTION

The external ear consists of the auricle and the external ear canal. There is a wide range of external ear abnormalities which are related to the size, shape, position of the ear or even the presence of preauricular pits or tags[1].

Microtia is a developmental malformation of the external ear, characterized by a small, abnormally shaped auricle. The prevalence of microtia has been reported to vary between 0.8-17.4/10,000 in different populations[2].

Microtia can occur unilaterally or bilaterally. The unilateral form is much more common, occurring in 79-93% of cases[3]. In unilateral microtia the right ear is more frequently affected (approximately 60% of the unilateral cases)[4]. Individuals with unilateral microtia usually have normal hearing in the other ear. Individuals with unilateral microtia usually have normal hearing in the other ear. Microtia is more common in males than females with a sex ratio of 1.5[5]. There are several grading systems for microtia. In the Marx classification[6], all of the features of anormal auricle are present in grade I, but the pinna is smaller than normal. In grade II, some anatomical structures are still recognizable. In the most common form, grade III (the peanut-shell type), only a rudiment of soft tissue is present[7].

The cause of microtia is still largely unknown, but most authorities would agree that the etiology is multifactorial. The exter-nal ear begins to develop in the sixth week of gestation and some reports implicate the drugs thalidomide and isotretinoin (Accutane) as causative agents[8] Most of the cases of microtia are isolated but can occur with other syndromes, most notably Goldenhar syndrome (oculoauriculoverte-bral dysplasia). Cervical vertebral defects are common, although

associated neurologic symptoms are uncommon. Hereditary etiolo-gies are infrequent[10].

More than 80% of microtia patients have aural atresia resulting in conductive hearing loss with air-conduction hearing typically reduced by 40-65 DB, whereas bone conduction is normal in more than 90% of the affected ears[11,12].

CASE REPORT

Aterm female baby weighing 3500 gram born at the 18th of July 2011 by spontaneous vaginal delivery to a 22 years old woman from Fallujah city, Garmah subdistrict. she is gravida 2 para 2, consanguineously married, her husband is 27 years old, he is her cousin, She gave history of infrequent antenatal care visits but she is not hypertensive, non diabetic & have no history of any other chronic or febrile illness, she have uneventful pregnancy with no history of using any drug or exposure to x.ray irradiation, they have fair income.There was no history of abortion & no family history of any kind of birth defects.

The baby born at home by midwife delivery & we receive her on 2nd day of life at the outpatient clinic, she was completely normal with the exception for the ears, *The baby had severely malformed ears with very minimal auricles and no visible external audi-tory canals*.(figure 1& 2). She is the 2nd baby, The 1st sibling (also female) was born 1 year ago with the same anomaly involving both ears, and died at the age of 15 days because of sepsis, that is the reason why this case was reported.

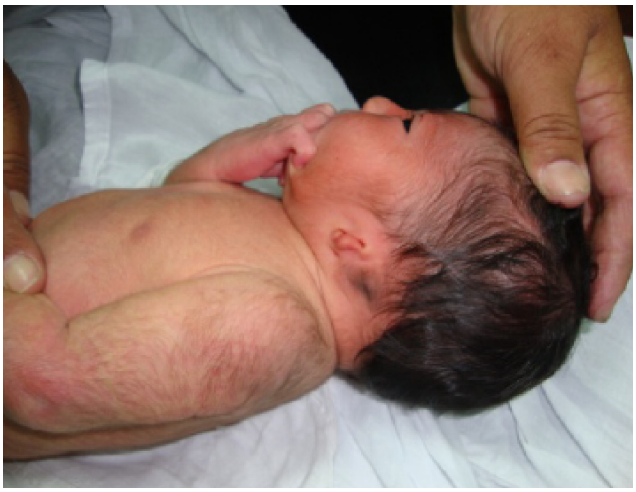

Figure 1: Left ear

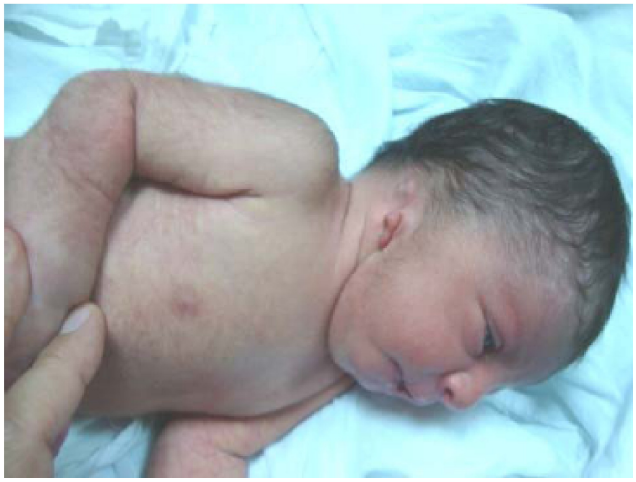

Figure 2 : Right ear

DISCUSSION

Microtia is not very common with an incidence range from 1 in 5,000 to 1 in 20,000 births, Although the etiology of microtia is poorly understood, both environmental and genetic factors have been implicated[13]. There are many risk factors for microtia, such as gestosis, anemia, race, high maternal or paternal age and multiple births[14].

Mothers with chronic type I diabetes are at significantly higher risk for having a child with microtia[15]. Certain medications such as isotretinoin, a widely used dermatologic drug, or mycophenolate mofetil (MMF) taken by the mother during pregnancy have also been implicated as microtia-predisposing factors. 83% of pregnancies with isotretinoin exposure result in spontaneous abortion or infants with serious birth defects including microtia[16]. MMF is an immunosuppressive agent, prescribed after solid organ transplantation. Patients who take MMF as immunosuppressive therapy during pregnancy are at risk for having a child with birth defects including microtia, cleft lip and palate[17].

Although there is strong evidence confirming the importance of environmental causes for microtia, it is believed that genetic components are also involved[18]. Estimates on the percentage of familial cases among microtia cases vary widely, ranging from 3-34%[18]. Both single gene defects and chromosomal aberrations have been reported in different microtia associated syndromes[18,19].

Recent reports have drawn attention to increases

Regular Paper

in congenital birth anomalies and cancer in Fallujah, Iraq blamed on teratogenic, genetic and genomic stress thought to result from depleted Uranium contamination following the battles in the town in 2004. An attempt to investigate the genetic familial causation of four individual cases of congenital anomaly has subsequently been made[20] The authors concluded that in these cases there was no familial genetic basis and that sporadic untargeted effects were responsible, causes which they described as epigenetic. The environmental aspect of the problem was studied in 2010 when hair sample analysis of 52 elements was carried out on the parents of children diagnosed with congenital anomalies, In addition soil and water samples were analyzed, Results indicated that man-made slightly enriched uranium was present in soil and in the hair of the parents[21].

Here we reported afamily having 2 succesive female siblings with bilateral Microtia within aperiod of 12 months. the 2 parents are young, healthy & they denied history of exposure to any of the risk factors & teratogens mentioned above, they have no prior family history of microtia or any birth defect. Chromosomal study for the parents & the baby failed to detect any chromosomal anomaly.

Knowing that the condition is multifactorial & accordingly both genetic & environmental factors can contribute to it's occurrence twice in this family.

CONCLUSIONS AND RECOMMENDATIONS

The absence of contributory family and drug history, in addition to the non-specific linkage to any environmental factor lead us to the belief that the successive occurrence of microtia in the two sisters is of sporadic occurrence. This defect could be due to environmental contaminants which are known components of modern weaponry. Investigations of metal contaminants, and elucidation of the types and body burden of metals, combined with simultaneous registry of the population's reproductive history, will allow the identification of families at high risk and will facilitate therapeutic measures to remediate the damages.

CONSET

Conset for publication have been obtained from the two parents.

ETHICAL APPROVAL

Ethical approval was granted by the scientific committee in Fallujah general hospital

REFERENCES

[1] P.E.Kelley, M.A.Scholes; Microtia and congeni-tal aural atresia. Otolaryngol Clin North Am., **40(1)**, 61-80, Feb **(2007)**.

[2] E.E.Castilla, I.M.Orioli; Prevalence rates of microtia in South America. Int.J.Epidemiol., **15(3)**, 364-8 **(1986)**.

[3] M.B.Forrester, R.D.Merz; Descriptive epidemiology of anotia and microtia, Hawaii, 1986-2002. Congenit.Anom (Kyoto), **45(4)**, 119-24 **(2005)**.

[4] J.Harris, B.Kallen, E.Robert; The epidemiology of anotia and microtia. J Med Genet, **33(10)**, 809-13 **(1996)**.

[5] O.Sanchez, J.R.Mendez, E.Gomez et al; Clinico-epidemiologic study of microtia. Invest.Clin., **38(4)**, 203-17 **(1997)**.

[6] H.Marx; Die Missbildungen des ohres, In: A.Denker, O.Kahler, (Ed); Handbuch der Spez Path Anatomie Histologie Berlin, Germany, Springer, 131 **(1926)**.

[7] H.Okajima, Y.Takeichi, K.Umeda et al.; Clinical analysis of 592 patients with microtia. Acta Otolaryngol Suppl, **525**, 18-24 **(1996)**.

[8] A.F.Jahn, K.Ganti; Major auricular malformations due to Accutane (isotretinoin). Laryngoscope, **97(7 Pt 1)**, 832-835, Jul **(1987)**.

[9] B.Brent; The pediatrician's role in caring for patients with congenital microtia and atresia. Pediatr Ann., **28(6)**, 374-383, Jun **(1999)**.

[10] F.Alasti, A.Sadeghi, M.H.Sanati et al.; A mutation in HOXA2 is responsible for autosomal recessive microtia in an Iranian family. Am.J.Hum.Genet, **82(4)**, 982-91 **(2008)**.

[11] H.Weerda; Classification of congenital deformities of the auricle. Facial.Plast.Surg., **5(5)**, 385-8 **(1988)**.

[12] J.Harris, B.Kallen, E.Robert; The epidemiology of anotia and microtia. J.Med.Genet, **33(10)**, 809-13 **(1996)**.

[13] E.E.Castilla, I.M.Orioli; Prevalence rates of microtia in South America. Int.J.Epidemiol, **15(3)**, 364-8 **(1986)**.

[14] P.Mastroiacovo, C.Corchia, L.D.Botto et al.; Epidemiology and genetics of microtia-anotia: A registry based study on over one million births. J.Med.Genet, **32(6)**, 453-7 **(1995)**.

[15] R.S.Stern, F.Rosa, C.Baum; Isotretinoin and pregnancy. J.Am.Acad.Dermatol., **10(5 Pt 1)**, 851-4 **(1984)**.

[16] A.Perez-Aytes, A.Ledo, V.Boso et al.; In utero exposure to mycophenolate mofetil: a characteristic phenotype? Am.J.Med.Genet.A, **146(1)**, 1-7 **(2008)**.

[17] C.W.Cremers; Meatal atresia and hearing loss. Autosomal dominant and autosomal recessive inheritance. Int.Pediatr.Otorhinolaryngol, peer-00552670, version 1 - 6 Jan 2011, **8(3)**, 211-3 **(1985)**.

[18] T.Klockars, S.Suutarla, E.Kentala et al.; Inheritance of microtia in the Finnish population. Int.J.Pediatr Otorhinolaryngol, **71(11)**, 1783-8 **(2007)**.

[19] Samira Alaani, Mozhgan Savabieasfahani, Mohammad Tafash, Paola Manduca; Four Polygamous Families with Congenital Birth Defects from Fallujah, Iraq, Int.J.Environ.Res. Public Health, **8**, 89-96 **(2011)**.

[20] Samira Alana, Muhammed Tafash, Christopher Busby, Malak Hamdan; Eleonore Blaurock-Busch, Uranium and other contaminants in hair from the parents of children with congenital anomalies in Fallujah, Iraq, Conflict and Health, **5**, 15 **(2011)**.