MRI features in foetal microtia

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Research Article

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Abstract

Background: Atresia of the external auditory canal is positively correlated with the difficulty and success rate of operation after birth. At present, ultrasound screening often obtains images of body position, placenta and amniotic fluid, and the detection rate is low. Moreover, it is unable to evaluate whether the external auditory canal shows atresia.

Methods: We retrospectively conducted MRI features of 9 cases those were diagnosed foetal microtia from May. 2019 to Oct. 2020.

Results: Nine cases of microtia foetus were analysed: male, five cases; female, four cases; right ear, five cases; left ear, four cases; and degree I, one case (bilateral external auditory canal is shown); degree II, eight cases (affected external auditory canal is not shown, six cases of normal external auditory canal shown, two cases of normal external auditory canal not shown). All parturients underwent amniocentesis full exon gene detection, of which the results were negative. The magnetic resonance imaging (MRI) features of microtia, included abnormal external ear morphology, disappearance of normal structure, mass-like and small piece-like soft tissue shadow and equal signal on T2-weighted (T2W) imaging. The upper and lower diameters were significantly smaller than that of the normal side. The MRI features of external auditory canal atresia included disappearance of T2W linear high signal shadow in the temporal bone scale.

Conclusions: Foetal MRI can diagnose microtia and evaluate atresia of the external auditory canal.

Background

Congenital microtia is one of the most common surface birth defects. It is the second most common facial deformity after cleft lip and palate. The prevalence of microtia greatly varies from 0.83/10000 to 17.4/10000 worldwide[1], while it is 3.06/10000 in China[2]. It is usually manifested as abnormal shape, size or position of the external ear. Concurrently, > 90% of microtia cases show atresia[3], leading to conductive hearing impairment. Postnatal external auditory canal plasty is required, but the operation is complex and has many complications[4]. During screening microtia, ultrasonography is often affected by foetal position, placenta, amniotic fluid and other factors, and its detection rate is low. Recently, there are a few reports on microtia[5,6], but they are unable to evaluate whether the external auditory canal shows atresia. Foetal MRI has been widely been used in the diagnosis of foetal congenital diseases due to its large field of vision and high soft tissue resolution[7,8]. The author found that the foetal external auditory canal is filled with amniotic fluid, which can be used as a natural contrast agent to better evaluate the development of the external auditory canal. This study summarised the MRI data of foetal microtia, and further used MRI to evaluate whether the external auditory canal is atresia and reviewed the literature on microtia.

Methods

The nine cases were achieved from 2 hospital of Chongqing from May. 2019 to Oct. 2020. All of those were performed with amniocentesis full exon gene test and followed up to birth (or induct).

Philips 1.5T MRI was performed with 32 channel phased array body coil. The scan parameters were as follows: fetal head axial and coronal single shot turbo spin echo (SSH TSE) sequence (TE 15000ms, tr 120ms) and balance fast field echo (BTFE) sequence (TE 3.4ms, tr 1.7ms). Layer thickness is 4mm, layer spacing is - 1mm, FOV is 300 × 300, matrix is 300 × 218, nex1, turning angle is 90 °.

Results

There were five cases (55.56%) in the right ear and four (44.44%) in the left ear. All cases were single microtia. Single microtia refers to cases with or without external auditory canal abnormalities but without other organ malformations, excluding external ear-related syndrome. There were five male foetuses (55.56%), four female foetuses (44.44%), four normal deliveries and five cases of induced labour (all within 1 week after MRI).The minimum gestational age was 24 weeks, the maximum was 30^{+5} weeks, and the average was ($28\pm2^{+1}$) weeks. All pregnant women had no abnormal pregnancy history and no family history of external ear malformation. Amniocentesis full exon gene test was performed, and the results were negative. No gene defect related to microtia was detected.

The MRI features of microtia, included abnormal external ear morphology, disappearance of normal structure, mass-like and small piece-like soft tissue shadow and equal signal on T2-weighted (T2W) imaging, and the upper and lower diameters were significantly smaller than that in the normal side. The MRI features of external auditory canal atresia included disappearance of T2W linear high signal shadow in the temporal bone scale.(Fig 1)

In this study, microtia was divided into three grades: Grade I showed that the surface marker structure of the auricle was recognisable, the contour was smaller than the normal side, there was a small concha cavity and the external auditory canal was narrow or showed atresia. Grade II was the most common, the residual ear was peanut or sausage shaped, most of the auricle structure could not be identified and the external auditory canal showed mostly atresia. Grade III only had small skin polyps or bulges or even no ear[9].

Among the nine cases in this study, one case was grade I, eight cases were grade II and none was grade III. In one case of caesarean delivery of the female infant, left microtia degree I and bilateral external auditory canal existed. The other eight cases of microtia were degree II, including six cases of visible normal lateral external auditory canal and malformation ipsilateral external auditory canal atresia and two cases of bilateral external auditory canal that were not shown, consistent with the results of MRI.

Discussion

Microtia is the result of multiple factors and mechanisms. Although the pathogenesis remains unknown, genetic material mutation is an important pathogenesis[10,11]. Foetal microtia has significant genetic

evidence: monozygotic twins have higher consistency of disease than heterozygotic twins, and the incidence of microtia is higher in relatives with minor malformations, such as appendage and fistula[12,13]. However, all cases showed sporadic simple microtia without family history, and amniocentesis exon gene test was normal.

Microtia can be regarded as an independent malformation or part of multiple malformations or syndromes. The common congenital malformations of microtia, include facial, congenital cardiovascular and vertebral malformations, most of which belong to the spectrum of eye-ear-spine diseases[14,15]. The characteristics of microtia are more common in male and right than in female and left[10]. The reason could be that the male androgen can reduce the respiratory rate of mitochondria and increase the sensitivity to chemical hypoxia, and the maturity of the mitochondria in the right ear occurs later than that in the left ear[3]. In this study, male-to-female ratio for the right ear was 5:4, and that for the left ear was 4:5, which is consistent with previous literature reports.

Han Juan[16] et al. used CT scanning to study patients with an average age of 12.44 years and found that with the increase of auricle deformity, the proportion of external auditory canal atresia increased. Among the nine cases, one case of type I microtia had no atresia of external auditory canal, while the other eight cases of type II microtia had atresia of the external auditory canal, which was basically consistent with the previous study results. In this study, in addition to the nine cases, there was one foetus with 29 + 4 weeks of gestation, which was diagnosed with microtia by ultrasonography. MRI showed that the external ear was normal and the external auditory canal exists. After follow-up, there was no abnormality in the appearance of the newborn ear, and the external auditory canal existed. This is consistent with the previous results of Wang TT[17] et al. MRI has important diagnostic value for external ear malformation and can be used as a supplement to ultrasonography. Of the nine cases of microtia, eight were diagnosed by the first MRI examination, and one was diagnosed by repeated scanning on the second day. At the first scan, both sides of the foetal head were close to the placenta and uterine wall, the external ear structure was compressed, the display was unclear and the bilateral external ear structure and external auditory canal were not obvious. After repeated scanning on the second day, the external ear structure of this case showed abnormal shape on the diseased side, and the bilateral external auditory canal was not displayed. Therefore, when MRI is used to diagnose microtia, if the external ear structure cannot be clearly displayed in one scan, the patient can be asked to undergo repeated scan after activity (foetal position change) until the external ear structure can be clearly displayed, but the specific absorption rate in MRI should be controlled below 2W/kg[18].

The auricle and external auditory canal are developed from the first branchial arch, second branchial arch and first branchial cleft tissue from the 6th week of embryo development. By the 16th week of embryo development, the contour of the external ear is basically formed. At the 26th week, the external auditory canal can reach one-third of its full length. The whole process of development is completed at the age of 9-10 years[19]. In this study, two foetuses with gestational age < 25 weeks of gestation did not show normal lateral external auditory canal. The reason could be that the foetal age is small, the external auditory canal has not communicated with the amniotic sac, there is no amniotic fluid filling in the

external auditory canal and there is no contrast, so it cannot be shown on MRI. Moreira et al. used MRI to study 144 foetuses and found that the external auditory canal in most cases could not be displayed until 29 weeks of gestational age[20]. However, the minimum gestational age of the external auditory canal in this study was 25 + 4 weeks, and it was confirmed after induction of labour within 1 week. Therefore, the minimum gestational age of the external auditory canal should be further confirmed by collecting a large number of cases.

This study has some limitations: First, the sample size of this study was small, so we can further collect cases and increase the sample size in the future; second, we only used MRI to evaluate the existence of the external auditory canal but did not measure the length and width of the external auditory canal to evaluate the development of normal external auditory canal in foetuses of different gestational ages, which can be explored in the future; and third, foetal MRI can only show the hearing loss of the middle ear. Further, it is difficult to distinguish the anatomical details of the bone.

Conclusions

To conclude, microtia can be diagnosed by MRI, and MRI has unique and irreplaceable advantages in detecting external auditory canal atresia.

Abbreviations

magnetic resonance imaging(MRI)

T2-weighted (T2W)

Declarations

Ethics approval and consent to participate

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Ethical approval was waived by the local Ethics Committee of Chongqing health central for women and children in view of the retrospective nature of the study and all the procedures being performed were part of the imaging examination. All methods were carried out in accordance with relevant guidelines and regulations. Informed consent was obtained from all participant's parents.

Consent for publication

Written informed consent was obtained from the patient's parents for publication of those cases report.

Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

P.S. organized the study, M.C. collected the data and drafted the manuscript. Y.B. scanned the foetaus.All authors have read and approved the manuscript.

Acknowledgements

Not application

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Table

Table 1 Datum of nine cases

Num.	Female(F)/ Male(M)	Gestational Age(Weeks)	Right(R)/ Left(L)	Characteristic of MRI		Microtia Grade (I/II/III)
				Size (mm)	External Auditory Cannal Yes/No	
1	F	25^{+4}	R	7.7	N	II
2	F	26^{+6}	L	10.3	Y	I
3	F	24	L	7.8	N	II
4	M	28	L	8.4	N	II
5	M	30^{+5}	R	15.9	N	II
6	M	29^{+4}	L	14.8	N	II
7	F	27^{+5}	R	11.9	N	II
8	M	27^{+5}	R	16.2	N	II
9	F	29	R	13.5	N	II

Figures

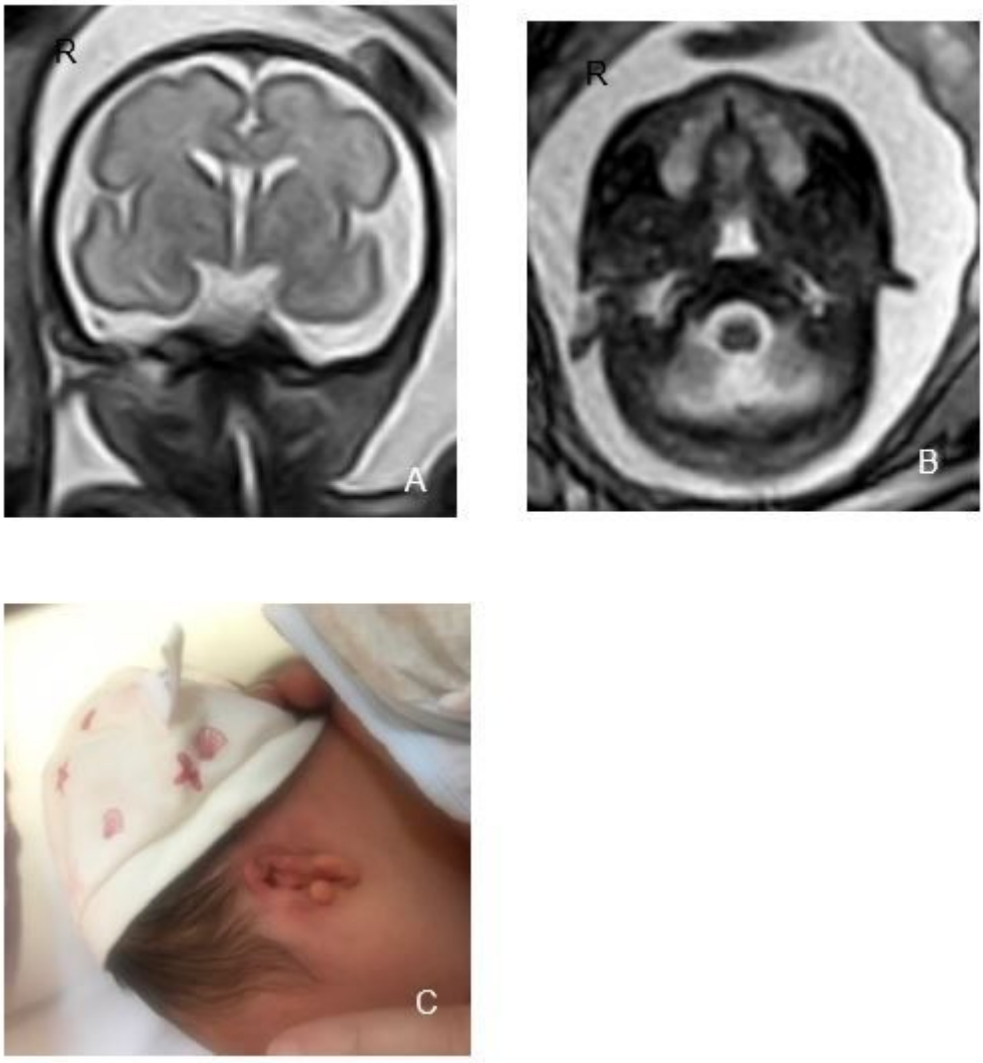

Figure 1

Case 6: female,34 years old, 29 + 4 weeks gestational age, left microtia. A: On coronal T2W, the right external auditory canal showed a linear T2W high signal shadow from the external superior oblique to the internal inferior; it was connected with the amniotic cavity; there was no T2W high signal shadow on the left temporal bone scale. B: On axial T2W. C: Postnatal left auricle deformity, external auditory canal atresia.