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Case report

Reversible splenial lesion syndrome in sisters with sensorineural deafness as the first manifestation

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ABSTRACT

Reversible splenial lesion syndrome (RESLES) is a rare clinico-radiological disorder syndrome with unclear pathophysiology. Here, two sisters with sensorineural deafness as the chief complaint diagnosed with RESLES was reported. Although the sisters had the disease successively, they were divided into two types on imaging by isolated lesions of splenium of the corpus callosum (SCC) and extensive lesions of the corpus callosum. The clinical manifestations and lesions on MRI disappeared after 6 months. The sensorineural deafness of the sisters in this article may be caused by transcallosal auditory pathway (TCAP) injury. Auditory handicap has been found in previous RESLES cases, indicating that we know little about the connection between the SCC and the auditory pathway, and further research is needed.

1. Introduction

Reversible splenial lesion syndrome (RESLES) is considered to be a clinical and radiological syndrome with a unique benign process mainly caused by reversible lesions in the splenium of the corpus callosum (SCC) [1, 2, 3, 4, 5, 6]. With the deepening of clinical knowledge and the development of medical imaging, the reversible splenial lesion syndrome has been raised to attract more and more attention. The corpus callosum, as a structure connecting the left and right cerebral hemispheres, plays an important role in balancing and coordinating information in the cerebral hemispheres. The pressure of the corpus callosum is an important auditory conduction pathway [7]. The literature on corpus callosum injury and deafness has earlier case reports, and reports of deafness caused by reversible corpus callosum syndrome are rare [8]. This paper reported two cases of reversible splenial lesion syndrome with sensorineural deafness as the first manifestation, and reviewed with literature.

2. Case reports

2.1. Patient 1

The first patient was a 33-year-old female who started working in a rubber factory for 2 months before the onset of the disease. She was admitted to the hospital due to a bilateral sudden onset hearing impairment for 6 days, and 1 day later developed symptoms of delirium, aphasia, orientation, memory and computational impairment. Cranial MR showed: bilateral basal ganglia had a patchy high T2 abnormal signal; corpus callosum had patchy low T1, high T2, high FLAIR signals with unclear margins; and the DWI sequence presented a high signal (Figures 1 and 2). Audiometry showed bilateral sensorineural deafness. Cerebrospinal fluid (CSF) examinations were normal according to cell counts, as well as glucose and protein levels. Also, electroencephalography (EEG) had no epileptic discharges. Nutritional nerve and glucocorticoids were used for treatment. Methylprednisolone sodium succinate was administered intravenously at 80 mg per day for 5 days. After 3 days of treatment, the new cranial MRI became better: abnormal signals changed to spot-like lesions from patchy ones (Figures 1 and 2). After one week of treatment, the patient's delirium disappeared, language function recovered, orientation and calculation abilities were basically normal, and memory power was just slightly poor. Re-examination of cranial MRI showed that besides the area, the strength of abnormal signals turned to weak (Figure 1). After 2 weeks of treatment, the patient's hearing returned to normal and cognitive function returned to normal. Followed up for 6 months, the patient was back to normal with no complication.

2.2. Patient 2

Another patient was a 31-year-old female, the sibling of patient 1, who also had started working in the same rubber factory 2 months before the onset of the disease. She was hospitalized for 3 days with

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bilateral hearing impairment. Neurological examination: consciousness was clear but unresponsive, orientation was fair, while calculation was slightly poor. Cranial MRI revealed that the left frontotemporal parietal occipital lobe had patchy iso T1, slightly long T2 and high FLAIR abnormal signals; on DWI sequence, lesions showed diffuse restricted changes (Figure 3). Audiometry showed bilateral sensorineural deafness. For cerebrospinal fluid (CSF) examinations, the results were regular. Nutritional nerve and glucocorticoids were as pivotal therapies. Dexamethasone sodium phosphate was administered intravenously at 10 mg per day for 3 days. After 3 days of treatment, the bilateral frontotemporal parietal occipital lobe became patchy slightly longer signals on T1 weighted sequences, lesions on T2, DWI and FLAIR sequences had no change as before by cranial MR (Figure 3). One week after treatment, the clinical symptoms of the patient disappeared, except for mild hearing loss. For review, cranial MRI showed that bilateral frontal parietal lobe and corpus callosum had patchy iso/slight T1, slightly long T2 and high FLAIR abnormal signals (Figure 3). After 2 weeks of treatment, the patient's hearing returned to normal, other clinical symptoms disappeared. Cranial MRI showed that the lesions had resolved completely (Figure 3). Six months later, we received that the patient was generally good with normal life via telephone interview.

2.3. Patient consent

The patient/next of kin has consented to the submission of the case report for submission to the journal.

3. Discussion

As early as 2004, medical imaging changes of reversible damage to the corpus callosum were found from some patients presenting with mild encephalitis/encephalopathy, antiepileptic drugs, metabolic disorders, tumors, and viral infections, then called 'Mild Encephalopathy/Encephalitis with Reversible Splenial Lesion (MERS)' [9, 10, 11]. In 2011, Garcia-Monco JC, et al. described and summarized the clinical and iconography features of RESLES, defined as a clinical and radiological syndrome of a unique benign process dominated by reversible lesions in the SCC [12]. Takanashi J, et al. classified RESLES according to medical imaging changes into two types: isolated SCC lesion (type 1); large SCC lesion extending into white matter and/or entire corpus callosum (type 2) [9,13,14]. In this paper, although the two sisters had the same sensorineural deafness as the first and chief clinical symptom, they respectively belonged to type 1 and type 2 of RESLES because of district iconography. Imamura T, et al. also reported sisters with RESLES, but they were 6 and 2 years old separately [15].

The causes of RESLES were complex, most of which were caused by discontinuation of antiepileptic drugs, infection, high-altitude cerebral edema, poisoning or abnormal metabolism (hypoglycemia and hypernatremia) [4, 16, 17, 18]. Studies have reported that RESLES related to neuroleptic malignant syndrome in schizophrenic patients. In this article, 2 cases were physically fit and had no history of drug dependence and addiction. From clinical manifestations and laboratory examinations, drugs, infections and metabolic abnormalities could be excluded. Combined with the working environment of the patient (rubber processing factory), organic solvent poisoning was not excluded. Unfortunately, there was no relevant evidence. Although chronic alcoholism can also cause reversible corpus callosum damage [19], there was no relevant medical history in this case, so Marchiafava-Bignami disease can be excluded. Marchiafava-Bignami disease (MBD) is a neurological disease with symmetrical demyelination in the CC, which is related to chronic alcoholism and malnutrition [20, 21, 22, 23]. Susac's syndrome is distinguished by inflammation and occlusion of pre-capillary arterioles, clinically accompanied by branch retinal artery occlusion, encephalopathy and hearing loss [24, 25, 26]. The two patients in this article do not have symptoms related to branch retinal artery occlusion, so susac's syndrome is not considered.

It has been ever thought that RESLES was more common in children. Common symptoms are considered to include cognitive impairment, convulsions, mental abnormalities, coma, ataxia, drowsiness, visual disturbances, and delirium [5, 6, 27]. However, reports of sensory deafness caused by RESLES are rare. A case has been spoken that a patient with a pineal tumor suffered from transient sensorineural deafness after surgical resection of the splenium part of the corpus callosum [28]. It is first found that the sisters in this article had RESLES with sensorineural deafness as the initially manifestation.

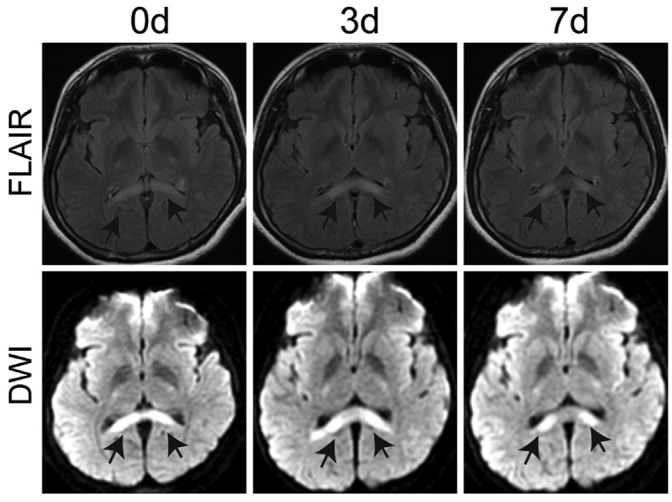

Figure 1. MR imaging of patient 1. On admission, the corpus callosum could be seen high FLAIR abnormal signal with unclear margins, lesion was high signal on DWI. After 3 days of treatment, the corpus callosum could be seen high FLAIR abnormal signal with unclear margins, the lesion was high signal on DWI. After 7 days of treatment, the lesions had basically recovered, as seen on DWI.

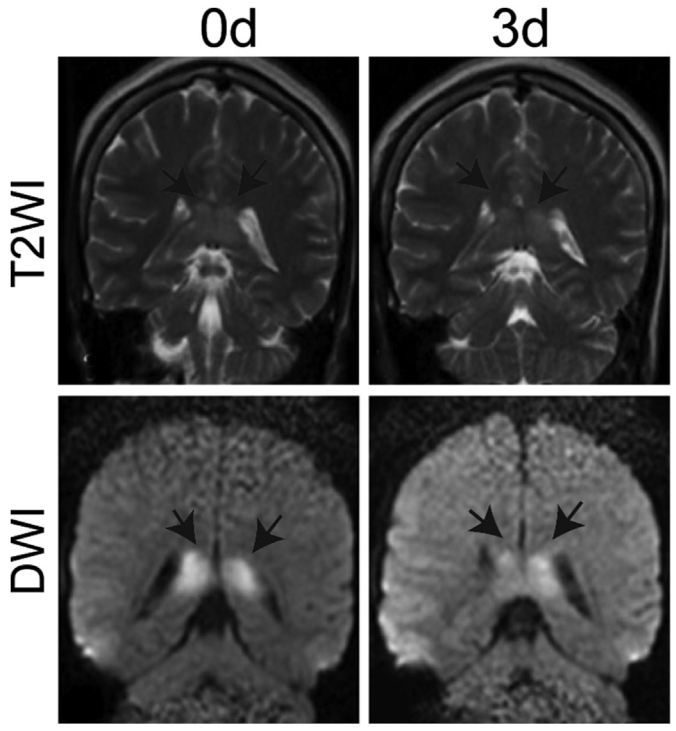

Figure 2. MR imaging of patient 1. On admission, T2-weighted image revealed lesions in the splenium of the corpus callosum, lesion was high signal on DWI.

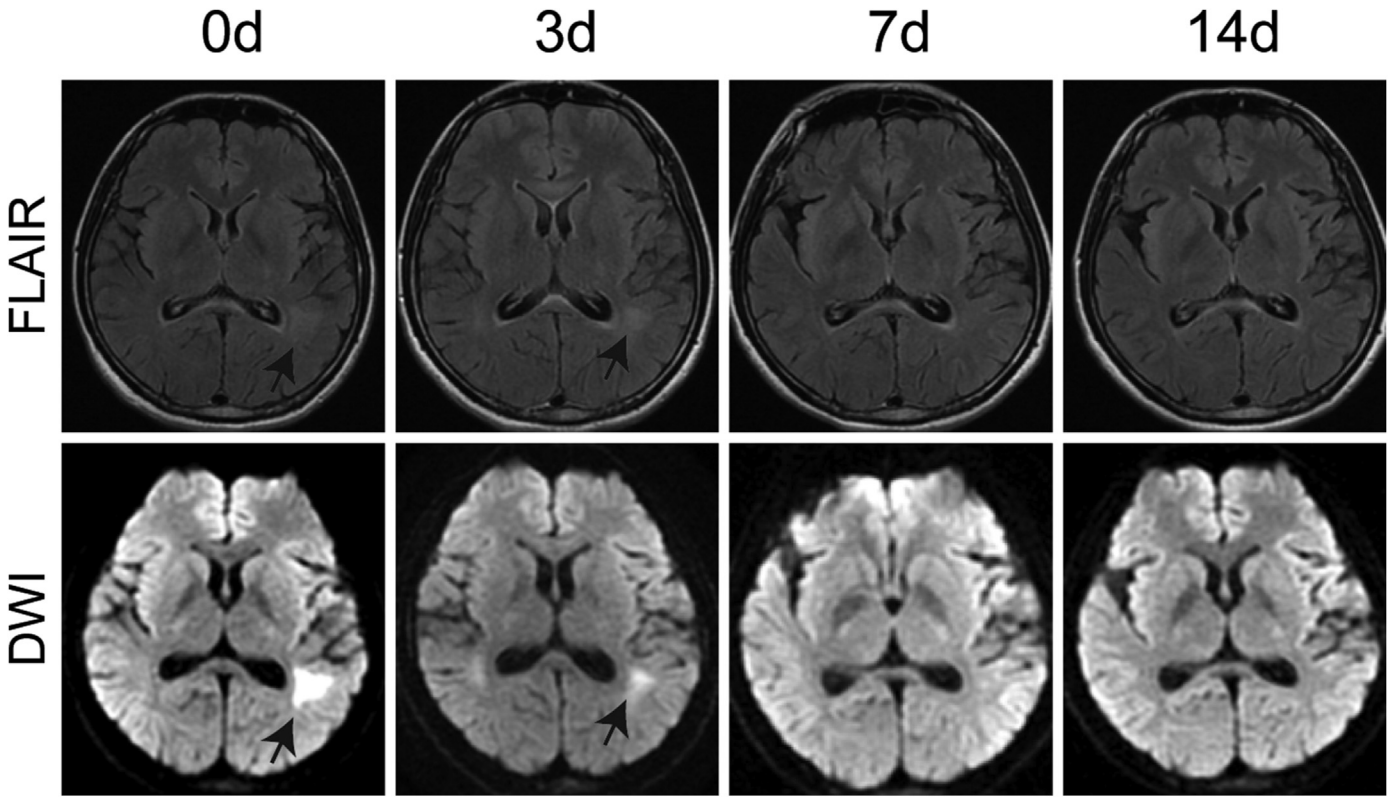

Figure 3. MR imaging of patient 2. On admission, the left frontotemporal parietal occipital lobe had high FLAIR abnormal signals, lesions showed diffuse restricted changes on DWI. After 3 days of treatment, lesions in the diffuse white matter and entire corpus callosum could be seen high FLAIR abnormal signal with unclear margins, and high signal on DWI. After 7 days of treatment, the lesions had basically recovered, as seen on DWI. On day 14, the lesions had resolved completely.

The functional localization in the cerebral cortex is well understood, but the relationship between the corpus callosum and auditory conduction pathways is poorly known [29]. The main function of the corpus callosum is to coordinate the integration of the two hemispheres of the brain. Anatomically, SCC is the posterior and the largest component of the corpus callosum, while it is the most vulnerable part [3]. It is believed that the fibrous tracts in the SCC connect the parietal, temporal, and occipital lobes on both sides and integrate visual functions, so that lesions in this area often present visually abnormal [3, 6, 30, 31]. Besides, anatomical studies have revealed that the SCC contains fibers from the primary and secondary auditory cortex and other auditory response areas, which plays a vital role in auditory conduction and integration [7]. Its auditive pathway across the corpus callosum is known as the transcallosal auditory pathway (TCAP), suggesting that any damage to the conduction fibers of this pathway can lead to auditory handicap [32, 33]. Recent studies have shown that the auditory cortex is the basis of hearing, and the integrated regulatory function of the corpus callosum is equally important [29, 31]. Rock C et al. applied optogenetic methods to determine the mechanism of corpus callosum projection fibers on the auditory cortex [29]. Pujol J et al. examined 22 patients with vascular periventricular leukoencephalopathy by head MRI and found that 4 patients had a loss of hearing function in the left ear, which may be related to damage to the auditory pathway of the cerebral hemisphere [34]. The sensorineural deafness of 2 patients in this article may be caused by TCAP injury, due to typical white matter lesions in the centrum semiovale on MRI and DWI in Case 2. Although there are sisters suffering from reversible corpus callosum syndrome [35], there have been no reports of deafness, and further researches are needed.

In summary, RESLES has an anatomical basis for sensorineural deafness, and its exact pathophysiological mechanism needs to be further studied. Genetic testing was not performed due to limitations. The intrinsic relationship of the co-morbidity of sisters or families also needs further studies.

Declarations

Author contribution statement

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The authors declare no conflict of interest.

Additional information

No additional information is available for this paper.

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