Case 418

1. Leukodystrophy hypoxic type

【Progress】

He was introduced to central mother-children center for further investigation using follow up with MRI spectroscopy.

【Discussion】

Brain nerve cells own axons as transmitters and dendrites as receivers. Corpus callosum compose of axon whose number reach to two hundred million, and connect neurotic signals between right and left hemisphere, although the number of axons decrease as ages advance. Axons own myelin that enable electric signals to flow at high speed. Hypofunction of myelin leads to nerve hypofunction.

Leukodystrophy is divided into two types: hypo-myelinating and demyelination (1-3). Many diseases with leukodystrophy are known such as X-linked adrenoleukodystrophy, Metachromatic leukodystrophy, Krabbe's disease, Pelizaeus-Merzbacher disease (1). They arise from abnormality of genetic inheritance, inducing hypoplasia of myelin or demyelination. Hypoplasia of myelin results from protein abnormality needed to form healthy myelin (1-3). Their diagnosis is mainly made using MRI image with MRI spectroscopy and genetic testing. Specific symptoms and tissue biopsy are also useful for diagnostic information. MRI spectrum is useful to investigate the metabolite of the oligodendrocyte or glia cells (1, 4-6).

MRI image abnormality for leukodystrophy is usually made on T2WI. Hyper-opacity signal on white matter in infants or children is found in leukodystrophy. The degree of hyper opacity is less marked in hypoplasia of myelin than demyelinating. The dark stripes in pale white-opacity of white matter are often found in hypoplasia of myelin, indicative of normal axons that are known as leopard sign or tigroid sign, while light white opacity, indicative of hypoxia of axons (1, 4-6). When pale hyper opacity is found in infants indicative of hypoplasia of axons on T2WIMRI, it might be less development of axons (immature axons) or hypoplasia of myelin. Because of various degrees of hypoplasia of myelin existing, follow-up studies six months later using T2MRI with spectroscopy are necessary for their differentiation (1).

Meanwhile, MRI image abnormality for demyelinating leukodystrophy is found as thick high signal intensity from deep white matter to subcortical white matter with least black stripes (1).

Hyper signal intensity in deep white matter is often encountered in adults, usually more often as ages advance on FLAIRMRI or T2WI. It is not clarified these signal intensity changes bring about electric signal intensity abnormality or not. It is known that axons number decrease as ages advance that might cause hyper signal intensity in deep white matter or they might reflect the disorder of glymphatic systems.

In our three-year-infant, local pale high signal intensity with dark stripes mimicking tiger stripe sign in deep white matter is depicted on T2MRI that indicates immature axon or hypoplasia of myelin.

【Summary】

We presented a three-year-old infant presented with dizziness accompanying with his mother. Pale white signal intensity with black stripes is depicted on T2WI indicative of immature axons or hypoplasia of myelination. He was introduced to Stem Hospital Center of Mother & Children with MRI and MR spectroscopy. It is borne in mind that pale white signal intensity with black stripes that are intact axons, known as leopard or tiger stripe sign on T2WI indicate hypomyelination of axons, while thick high signal intensity without black stripes indicate demyelination axons.

【References】

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