Case Report

1H-MR Spectroscopy in a Case of Isolated Intracranial Rosai-Dorfman Disease

Mariko DOAI, Hisao TONAMI, Naoko TSUCHIYA, Takuya AKAI, Nozomu KUROSE, Keiya HIRATA

1Kanazawa Medical University, Radiology, Ishikawa, Japonya 2Kanazawa Medical University, Neurosurgery, Ishikawa, Japonya 3Kanazawa Medical University, Pathology and Laboratory Medicine, Ishikawa, Japonya 4Kanazawa Medical University Hospital, Radiology department, Ishikawa, Japonya

Summary

A 61-year old female presented with a headache. CT and MR imaging showed a left frontal dural-based mass mimicking meningioma. 1H-MR spectroscopy showed elevated lipid peaks and decreased N-aspartate, choline and creatine peaks. No abnormal alanine peak was detected. The histological diagnosis was intracranial Rosai-Dorfman disease. Our case suggests that 1H-MR spectroscopy provides useful information in differentiating intracranial Rosai-Dorfman disease from meningioma.

Key words: Rosai-Dorfman disease, meningioma, MR imaging, 1H-MR spectroscopy

İzole İntrakranyal Rosai-Dorfman Hastalığında 1H-MR Spektroskopi

Özet


Anahtar Kelimeler: Rosai-Dorfman hastalığı; meningiomi; MRG; 1H-MR spektroskopi

INTRODUCTION

Rosai-Dorfman disease (RDD) is a rare benign histiocytic proliferation characterized by massive painless cervical lymphadenopathy. RDD commonly involves the cervical lymph nodes, and extranodal involvement is less common, while isolated intracranial involvement is even rarer with around 80 cases reported so far. They are mostly dural-based mass lesions presenting radiologically like meningioma, and histological analysis and immunophenotyping are essential to reach a correct diagnosis.

We experienced a case of isolated intracranial RDD in which 1H-MR spectroscopy provided useful information
in differentiating intracranial RDD from meningioma.

CASE PRESENTATION

A 61-year old female presented with a two-month history of headache. Her medical history showed hypertension and hypothyroidism, which were in good control with medication. Vital signs and neurological examinations were normal. Routine hematological and biological tests revealed no abnormalities.

Computed tomography (CT) showed a left frontal hyperdense dural-based mass lesion with surrounding white matter edema (Figure 1). Magnetic resonance (MR) imaging and $^1$H-MR spectroscopy were performed by a clinical 3-T system with a 32-channel head coil. MR imaging showed that the lesion was isointense relative to gray matter on T1-weighted images and hypointense on T2-weighted images, with an obvious dural attachment (Figures 2 and 2B). The lesion was homogeneously enhanced on Gd-enhanced MR images (Figure 2C). CT as well as MR imaging findings suggested meningioma. $^1$H-MR spectroscopy was performed with chemical shift imaging (TR: 2,000 msec, TE: 30 msec, voxel size: 10 10 15 mm). MR images in three orthogonal planes were used to locate the voxels for the $^1$H-MR spectroscopy and to minimize voxel contamination. The aim was to obtain an average spectroscopic representation of the largest possible part of the tumor while avoiding contamination of the sample by extra-tumoral tissues specifically bone, fatty scalp tissue and normal brain tissue. $^1$H-MR spectroscopy showed an elevated lipid peak (1.33 ppm), and decreased N-acetylaspartate (NAA) (2.02 ppm), choline (Cho) (3.20 ppm) and creatine (Cr) (3.04 ppm) peaks. No abnormal alanine peak was detected (1.48 ppm) (Figure 3).

The patient underwent left frontotemporal craniotomy with total removal of the tumor. The tumor was yellowish in color and relatively hard. Histopathological examination demonstrated numerous, proliferative histiocytes showing phagocytosed lymphocytes, plasma cells and red blood cells (emperipolesis) (Figure 4). Immunohistochemistry for S-100 protein was strongly positive for histiocytic cells (Figure 4B). Electron microscopic examination revealed deposit of lipofuscin in cytoplasm (not shown). On the basis of these histopathological features, the lesion was diagnosed as intracranial RDD.

Figure 1: Axial CT shows a left frontal hyperdense dural-based mass lesion (arrow) with surrounding white matter edema.
**Figure 2:** (A) Axial T1-weighted image shows an isointense mass lesion (arrow) with white matter edema. (B) The mass lesion shows hypointensity on T2-weighted image (arrow). (C) Post Gd-enhanced coronal MR image demonstrates homogenous enhancement of the dural-based mass lesion (arrow).

**Figure 3:** $^1$H-MR spectroscopy shows elevated lipid peak (arrow), and decreased N-acetylaspartate (NAA), choline (Cho) and creatine (Cr) peaks. No abnormal alanine peak was detected.
DISCUSSION

Rosai and Dorfman first described this rare disease entity in 1969, which was then known as sinus histiocytosis with massive lymphadenopathy, now named RDD\(^{(13)}\). Although the pathogenesis is not clearly understood, some have proposed the etiology to be either infection or immune deficiency\(^{(14)}\). RDD usually involves cervical lymph nodes. Involvement of an extra-nodal location has been observed in 43% of patients with RDD, whereas intracranial localization is rare and only 80 cases of RDD with intracranial involvement have been reported in the literature\(^{(3,11)}\).

The typical imaging findings of RDD include a dural-based, extra-axial, well-circumscribed enhancing mass with possible peripheral cerebral edema\(^{(15)}\). On CT, the lesion appears as a well-defined hyperdense mass\(^{(6,15)}\). On MR imaging, the lesion is isointense relative to gray matter on T1-weighted images and isointense or hypointense on T2-weighted images with homogeneous contrast enhancement\(^{(7,9,15)}\). These imaging appearances of intracranial RDD mimic meningioma\(^{(3,11,14)}\). Similar to previous reports, in the present case, CT as well as MR imaging did not aid in differentiating RDD from meningioma. On the other hand, \(^1\)H-MR spectroscopy showed an elevated lipid peak, and decreased NAA, Cho and Cr peaks. No abnormal alanine peak was detected. These \(^1\)H-MR spectroscopic findings suggest the lesion to be granulomatous lesions such as tuberculoma, not meningioma. The \(^1\)H-MR spectroscopic pattern of meningioma has been thoroughly described\(^{(2,5)}\). The most characteristic patterns of meningioma are the presence of alanine, high relative concentration of Cho and low concentration of Cr containing compounds\(^{(2,5)}\). NAA containing compound lipids are also absent or low\(^{(2,5)}\). Alanine signals are reported to be specific for meningiomas and it was absent in our case\(^{(2,5)}\). To our knowledge, only one case report has described using \(^1\)H-MR spectroscopy for isolated intracranial RDD. Symss et al. reported a case of intracranial RDD in which \(^1\)H-MR spectroscopy showed elevated lipid and NAA peaks. However, they have not done enough consideration of the \(^1\)H-MR spectroscopy\(^{(12)}\). In our case, \(^1\)H-MR spectroscopy showed an elevated lipid peak and a decreased NAA peak. The
discrepancy of an NAA peak may be derived from the voxel setting. In our case, the voxel was carefully adjusted to cover the depicted lesion areas shown on the MR images in three orthogonal planes in order to avoid contamination by extra-tumoral tissues. The elevation of the NAA peak in the previous reported case may be derived from the contamination of surrounding normal brain tissues. The high lipid peak shown in the 1H-MR spectroscopy in our case is thought to reflect abundant deposition of lipofuscin from the results of electron microscopic examination.

Tuberculoma has been reported to show a high lipid peak, more Cho and less NAA and Cr. Therefore, differentiation between intracranial RDD and tuberculoma by 1H-MR spectroscopy may be difficult.

Intra-or extra-axial locations, but is frequently located in the cerebral parenchyma. A solid extra-axial mass is reported to be extremely uncommon.

CONCLUSION
This case report is the first to demonstrate the detailed 1H-MR spectroscopic findings of RDD. Combined evaluation of CT and MR imaging findings and 1H-MR spectroscopy can help in narrowing down the differential diagnosis of RDD, especially from meningioma. Clinical usefulness of 1H-MR spectroscopy in the diagnosis of RDD needs to be confirmed in a larger series of patients.

REFERENCES

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