Case of Langerhans Cell Histiocytosis That Mimics Meningioma in CT and MRI

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INTRODUCTION

Langerhans cell histiocytosis (LCH) is a rare disorder histologically characterized by the proliferation of Langerhans cells. There are various clinical presentations including single organ involvement (unifocal), disseminated disease (multifocal), and systemic disease (systemic). LCH typically presents as a mass or bone lesion, with or without calcification. Imaging findings can be variable, ranging from a well-defined mass to a destructive lesion with bone erosion.

Case Report

A 13-year-old female patient sought treatment complaining of chronic headaches and dizziness that she had been experiencing for the preceding year. Head CT results showed a high density mass on the right calvaria with calcification spots and an adjacent skull defect with uneven borders. Non-contrast MRI revealed a heterogeneous area of signal intensity in T1WI and a high-intensity mass with low-intensity shadows within it in T2WI. Contrast MRI showed an intensified mass of 3.5×3.8×2.0 cm with a heterogeneous signal.

The lesion was connected with the dura mater through the wide base, showing a round or elliptical transparent bone defect. The edge of the defect may be clear or unclear, and the defect can involve inner and outer areas of cranial bones. Imaging often shows a round or elliptical transparent bone defect. The edge of the defect may be clear or unclear, and the defect can involve inner and outer areas of cranial bones. Cranial and intracranial changes measured by MRI include:

1) Lesions of the craniofacial bone and skull base with or without soft-tissue extension;
2) Intracranial extra-axial changes (hypothalamic-pituitary region, meninges, circumventricular organs);
3) Intracranial intra-axial changes (white matter and gray matter);
4) Cerebral atrophy.

The progression of the condition varies greatly, ranging from spontaneous regression to rapid progression, with or without recurrence and long-lasting sequelae.

The diagnosis of LCH is based on characteristic histopathological features. Under light microscopy, tumor cells present as large histiocytes with grooved nuclei, and mononuclear histiocytes and multinucleated giant cells are intermixed with eosinophils, lymphocytes, plasma cells, and neutrophil polymorphs. Immunohistochemistry shows S-100 protein and CD1a expression in Langerhans cells.

Electron microscopy may also reveal “tennis racket” shaped cytoplasmic inclusions within histiocytes called Birbeck granules, which is the gold standard for LCH diagnosis. Imaging often shows a round or elliptical transparent bone defect. The edge of the defect may be clear or unclear, and the defect can involve inner and outer areas of cranial bones. Cranial and intracranial changes measured by MRI include:

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CASE REPORT

A 13-year-old female patient sought treatment complaining of chronic headaches and dizziness that she had been experiencing for the preceding year. Head CT results showed a high density mass on the right calvaria with calcification spots and an adjacent skull defect with uneven borders (Fig. 1A, B). Non-contrast MRI revealed a heterogeneous area of signal intensity in T1WI and a high-intensity mass with low-intensity shadows within it in T2WI. Contrast MRI showed an intensified mass of 3.5×3.8×2.0 cm with a heterogeneous signal. The lesion was connected with the dura mater through the wide base, showing a round or elliptical transparent bone defect. The edge of the defect may be clear or unclear, and the defect can involve inner and outer areas of cranial bones.

The dural origin of the lesion was verified upon surgical dissection. There are no previous reports in the literature describing LCH of dural origin presenting in young patients with typical dural tail signs and meningeoma-like imaging findings. The current case report underscores the need for thorough histological and immunocytochemical examinations in LCH differential diagnosis.

Key Words: CT · MRI · Meningioma · Langerhans cell histiocytosis.
a dural tail sign and pressing downward on adjacent brain tissue. A strip-shape intensified signal was observed in the adjacent skull (Fig. 1C, D). Low-signal areas in the T2WI nd areas without intensification demonstrated by contrast MRI were identical to calcified areas in CT result.

The lesion was diagnosed initially as a meningioma and the patient underwent a craniotomy. During the surgery, it was revealed that the lesion originated in the dura mater and had

Fig. 1. A : Plain CT scan. Soft-tissue window, high-density mass on the right calvaria with internal calcification spots. B : Plain CT scan. Bone window and adjacent bone deficit with unclear edges. C and D : Enhanced MRI revealing an irregularly shaped tumor with clear boundaries and a wide base attached to the meninges (sagittal and coronal planes, respectively).

Fig. 2. Histopathological demonstration of LCH. Hematoxylin-eosin staining of the resected mass revealed a large number of Langerhans cells, multinucleated giant cells, and diffuse eosinophil lymphocyte infiltration (A). Immunohistochemistry revealed the presence of membranous CD1a (B), cytoplasmic CD68 (C), and nuclear S-100 (D). Original magnification : ×200.
grown downward, pressing on the brain tissue and invading the inner skull. The dura around the tumor was thickened. The tumor was elastic and had a rich blood supply, indicating endogenous angiogenesis. The tumor and surrounding dura were removed, and artificial dura material was used to fill the resultant defect in the dura mater. Histopathological examination revealed a grey-white irregular tissue with a dark brown cross-section. Microscopy revealed a large number of proliferated Langerhans cells and multinucleated giant cells, as well as infiltration of eosinophils and lymphocytes (Fig. 2). Immunohistochemical examination indicated that the tumor tissue was CD1a-, CD68-, and S-100-positive. Therefore, a final diagnosis of LCH was confirmed.

DISCUSSION

Here we reported the case of a 13-year-old female patient with a unifocal LCH of dural origin that was initially misdiagnosed as a meningioma based on imaging findings, but was later determined by the pathological examination to be LCH. To our knowledge, there have been no prior reports of teenagers having dura-originating calvarial LCHs that exhibit the typical dural tail sign and an imaging profile consistent with meningioma.

There have been prior reports of LCHs originating in the dura and the tentorium of the cerebellum. Cranial and intracranial changes associated with LCH include: 1) lesions of the craniofacial bone and skull base, with or without soft-tissue extension; 2) intracranial and extra-axial changes in the thalamic-pituitary region, meninges, and other circumventricular organs, including the pineal gland, choroid plexus, and ep- endyma; 3) intra-axial parenchymal disease in the gray matter or white matter, with striking symmetric lesions and a predominant neurodegenerative pattern in the cerebellum and basal ganglia; and 4) localized or diffused cerebral atrophy.

Meningiomas are relatively common tumors that occur most frequently on intracranial meninges. The World Health Organization (WHO) classification of central nervous system tumors defines various types of meningiomas. Most are slow-growing (WHO grade I), including some rare subtypes such as microcystic meningioma, secretory meningioma, and metaplastic meningioma. Brain-invasive (WHO grade II), atypical (WHO grade II), and anaplastic (WHO grade III) meningiomas are more aggressive. Specifically, osseous destruction can occur in atypical malignant meningiomas or hyperostosis-associated benign meningiomas, and approximately 25% of meningiomas show psammomatous calcification. Typically, benign meningiomas appear as round or elongated extra-axial masses with a broad dural attachment and have a density similar to that of the cerebrum in CT images. Enhanced scans of meningiomas show a dural tail sign, which indicates the presence of neoplastic dural infiltration and/or reactive vascularization of the adjacent dura. Low-intensity signals produced by calcification or vascular flow voids may be observed within the tumor. In addition to the typical meningioma signs, the LCH mass in the present case also presented with a slight defect of the adjacent intracranial skull consistent with atypical meningioma.

CONCLUSION

This report presents a case of dura-originating calvarial LCH that mimicked the imaging characteristics of a meningioma, including the typical dural tail sign. This case underscores the importance of a thorough histological biopsy and immunohistochemical examination in head tumor differential diagnosis.

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