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Case Report

A case of Langerhans cell histiocytosis of the skull in which preoperative methionine positron emission tomography was useful in comprehending the spreading of the lesion

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Abstract

Background: The problem with treatments against skull lesions of Langerhans cell histiocytosis (LCH) is that invasions often reach the bones and dura mater, making it difficult to accurately comprehend the range thereof prior to surgery. We herein report that ^{11}C-methionine positron emission tomography (PET) (Met-PET) carried out prior to surgery was useful in comprehending the spreading of the lesion.

Case Description: A 20-year-old female presented with swelling and dull pain on the left side of the head. A slightly heterogeneously reinforced tumor was observed inside the bone defect in the gadolinium-enhanced T1-wighted image upon magnetic resonance imaging (MRI) and the dura mater contacting the tumor was observed with an enhancing effect. Accumulation was poor in the center of the tumor upon Met-PET, and accumulation with a well-defined border was observed in the border thereof as well as the area adjacent to the brain. Surgical resection was performed; the pathological diagnosis was LCH. An invasion of tumor cells was observed in the dura mater with accumulation observed according to Met-PET. Moreover, the accumulation of tumor cells was observed in the area observed with accumulation inside the bone; however, the center part with poor accumulation lacked tumor cells, with fibrous tissue accounting for most parts.

Conclusion: Met-PET was believed to be helpful in comprehending the spreading of the tumor in the surroundings of the brain surface for skull lesions.

Key Words: Dural invasion, Langerhans cell histiocytosis, methionine positron emission tomography

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INTRODUCTION

Langerhans cell histiocytosis (LCH) is the proliferation of dendritic cells accompanying local or diffused invasion into the organs.[1] The problem with treatments against skull lesions is that invasions often reach the bones and dura mater, making it difficult to accurately comprehend the range thereof prior to surgery. To date, the utility of 18F-fluorodeoxyglucose positron emission tomography (hereinafter, referred to as FDG-PET) has

been reported regarding the spreading of lesions and treatment reactivity;[7,9] however, its uptake to the brain cortex is high and differentiation between dura mater lesions and the brain cortex is difficult, so it has not been established enough to differentiate between them. Meanwhile, ^{11}C-methionine PET (hereinafter, Met-PET) has low uptake to the brain cortex; therefore, there is a greater potential for differentiation between dura mater lesions and the brain cortex. However, as far as our research has shown, there are no reports on Met-PET regarding LCH. We herein report on our experience regarding a case of LCH of the skull accompanying dura mater invasion in which Met-PET carried out prior to surgery was useful in comprehending the spreading of the lesion.

CASE REPORT

Case
A 20-year-old female.

Past history
No major illnesses.

Present illness
The patient consulted our institute after becoming self-aware of swelling and dull pain in the left side of her head from 3 months prior.

Condition at admission
She was clearly conscious and no symptoms of neurologic deficit were observed. A soft flexible tumor was palpated in an approximately 3 × 3 cm region on the left side of her head and tenderness at that site was observed.

Blood chemistry findings
There were no inflammatory findings and the tumor marker was within the normal range.

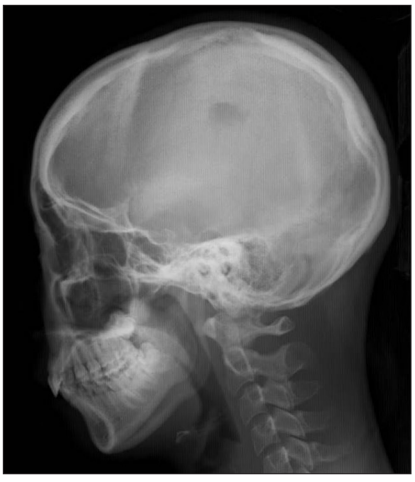

Figure 1: Skull film, lateral projection showed a rounded osteolytic lesion with a nonsclerotic rim in the left parietal bone

Neuroradiological findings
An oval radiolucent line was observed in the left cranium upon simple head X-ray [Figure 1]. A slightly heterogeneously enhanced tumor in left parietal bone was observed inside the bone defect in a gadolinium-enhanced T1-weighted image upon head magnetic resonance imaging (MRI); wherein, the dura mater and subcutaneous tissue contacting the tumor widely exhibited an enhancing effect and was found to have thickened [Figure 2a and b]. Nuclide accumulation was observed in the tumor inside the bone defect upon FDG-PET; however, the brain cortex accumulation was also high in the area adjacent to the brain surface, so the border was unclear, making evaluation difficult [Figure 2c]. No significant abnormal

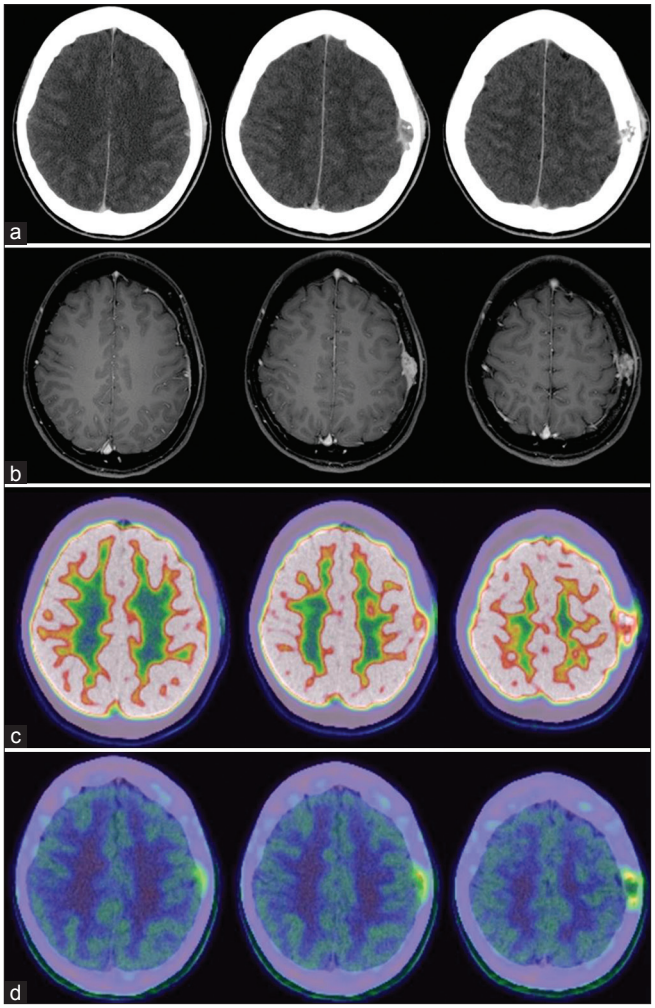

Figure 2: (a) A preoperative enhanced computed tomography revealed a bone defective part and an enhanced mass. (b) A preoperative Gadolinium enhanced T1-weighted image demonstrates the presence of a mass, which is enhanced with dural surface and subgaleal tissue. (c) Fluorodeoxyglucose positron emission tomography shows a high uptake in the osteolytic lesion obsereved on the left. (d) Methionine positron emission tomography demonstrates a high uptake in the marginal zone of the osteolytic lesion and dura matar, and a low uptake in the central zone of the osteolytic lesion

accumulations in other areas of the body were observed. Meanwhile, such accumulation was observed to be poor in the center of the tumor upon Met-PET, while accumulation with a well-defined border was observed in the border as well as the dura mater and subcutaneous tissue adjacent thereto [Figure 2d].

Accordingly, it was believed to be an extramedullary tumor such as LCH of the skull, meningioma, malignant lymphoma, etc. The tumor was symptomatic and enlarged within a relatively short period of time; therefore, surgery was carried out including pathological diagnosis.

Surgical findings

A U-shaped skin incision was made on the left side of the head. When the skin flap was reversed, the subcutaneous tissue was strongly adhered to the defective part of the bone. A craniotomy was carried out leaving a distance of approximately 3 cm from the defective part of the bone such that the range observed with accumulation upon Met-PET was sufficiently included [Figure 3a-e]. When the bone flap was reversed, the tumor was observed to be yellow in color. The tumor was adhered to the dura mater and the dura mater of the attachment site was yellow in color and particularly thick. The dura mater was excised, while leaving a border of approximately 1 cm from the thickened part of the dura mater [Figure 3f-i]. Tumor cells were not observed in the stump of the excised dura mater upon immediate pathological examination. The subcutaneous tissue adhered to the defective part of the bone was also resected and the excised stump was negative. The defective part of the bone was excised upon providing a border of approximately 1 cm [Figure 3c], and this was supplemented using artificial bone Biopex®. The defective part of the dura mater was sutured using artificial dura mater (GoreTex®).

Pathological findings

The accumulation of tumor cells having a nuclear groove and constricted nucleus in the border part observed with accumulation upon Met-PET was observed inside the extracted skull [Figure 4a-e]. The tumor cells were immunohistochemically positive for CD1a, DEC205, and Langerin. The center part with poor accumulation lacked tumor cells, with fibrous tissue accounting for most parts [Figure 4d]. Accordingly, it was diagnosed as LCH. The dura mater was observed to have undergone invasion of histiocyte-like tumor cells and eosinophil similar to the tumor itself in the region corresponding to the accumulation region upon Met-PET [Figure 5a-g]. Tumor cells and inflammatory cells were not observed in the dura mater stump.

Postoperative course

The course was good and the patient was discharged without any deficiency symptoms. Relapse has not been observed as of 7 months following surgery.

DISCUSSION

LCH is a symptom of abnormal proliferation of the dendritic cells that often occurs in young patients.[10] Although it can occur in bone throughout the entire body, it particularly often occurs in the skull, with a high incidence

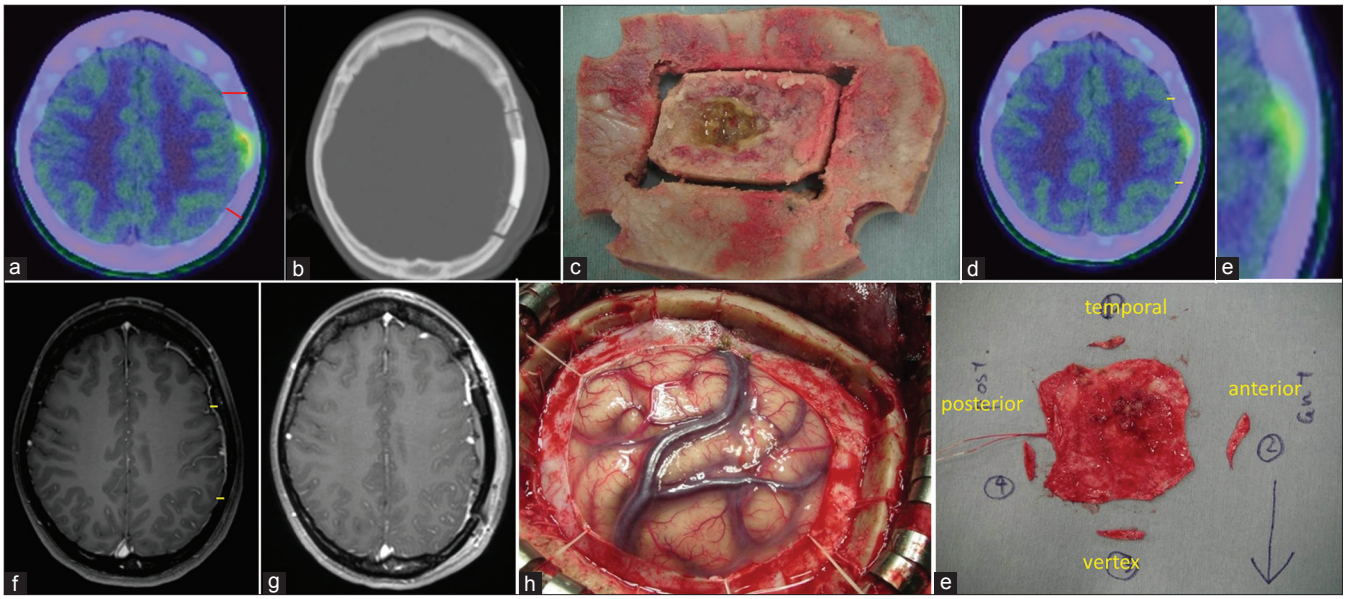

Figure 3: (a) Preoperative methionine PET. The red line shows the range of the bone excision. (b) Postoperative CT scan (bone image) (c) Intraoperative photograph of the bone. We performed bone excision and established a border, which was located about 1 cm from the tumor location. (d) Preoperative methionine PET. The yellow line shows the range of the dura excision. (e) Magnification image of the methionine PET (f) Preoperative enhanced T1-weighted image. The yellow line shows the range of the dura excision. (g) Postoperative enhanced T1-weighted image. (h) Intraoperative photograph of the brain surface. No invasion to the brain was observed. (i) Photograph of the dura mater. No tumor cells were observed in all excised specimens

of occurrence in the parietal bone and frontal bone.[11] It is characterized by the observance of a well-defined radiolucent line (punched out lesion) that does not accompany osteosclerosis upon simple skull X-ray.[10]

Although reports on MRI findings have occasionally been found in recent years, it is regarded that tumors exhibit iso to low signals upon T1-weighted imaging, high signals upon T2-weighted imaging, and enterogenous enhancing effects upon gadolinium-enhanced T1-weighted imaging.[2,3,5,8] Although there are few reports in which the enhancing effect of the dura mater and/or subcutaneous tissue is observed, regarding the pathological significance thereof, there are some cases that show dura mater invasion of the tumor and some cases that do not. Yunoki *et al*. reported a case in which the dura mater in the vicinity of the tumor was widely reinforced due to imaging MRI; however, because no tumor cells were observed in the dura mater, they reported that there is a high possibility that this is an inflammatory change and not an invasion of the tumor.[11] In contrast, Takeuchi *et al*. reported on a case in which the surrounding dura mater is reinforced in the same manner, but reported this as dura mater invasion.[10] Regarding the enhancing effect of subcutaneous soft tissue as well, Keyaki *et al*. reported that this was a tumor invasion from the histopathology.[4] However, there are reports mentioning that this was a change in reactivity,[6] and a consistent consensus has not been reached. That is, at the present stage, it is difficult to preoperatively predict the presence of a tumor invasion of the surrounding tissues from studying images of the imaging MRI.

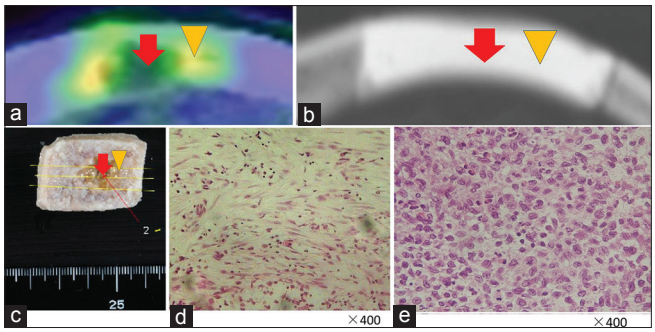

Figure 4: (a) Preoperative methionine PET. Arrow (D). Arrow head (e) (b) Postoperative CT scan (bone image). Arrow (D). Arrow head (E) (c) Photograph of the surgical specimen. Arrow (D). Arrow head (E) (d) Photomicrograph of the section D demonstrates fibrotic cell and less giant cell (hematoxylin and eosin stain, ×400) (e) Photomicrograph of the section E demonstrates multicleated giant cell (hematoxylin and eosin stain, ×400)

FDG PET has been determined to be useful for accurately diagnosing the spreading of LCH lesions.[7] The present case was observed as having nuclide accumulation in the tumor of the skull upon carrying out preoperative FDG PET. No other abnormal accumulations in the entire body were observed, so a diagnosis was made of a discrete lesion of the skull. Upon detailed investigation into the accumulation region, the dura mater and subcutaneous tissue approaching the brain surface had an indistinct border because accumulation of the brain cortex was fundamentally high and evaluating the spreading to the dura mater was difficult.

Meanwhile, regarding Met-PET, although the center of the tumor has poor nuclide accumulation, accumulation with a well-defined border was observed in the border thereof as well as in the dura mater adjacent to the brain. When findings from the pathology specimen and Met-PET were compared, the center part with poor accumulation lacked tumor cells, with fibrous tissue accounting for most parts. Accumulation of tumor cells was observed in the tumor periphery observed with accumulation. Moreover, further regarding the dura mater, accumulation of tumor cells

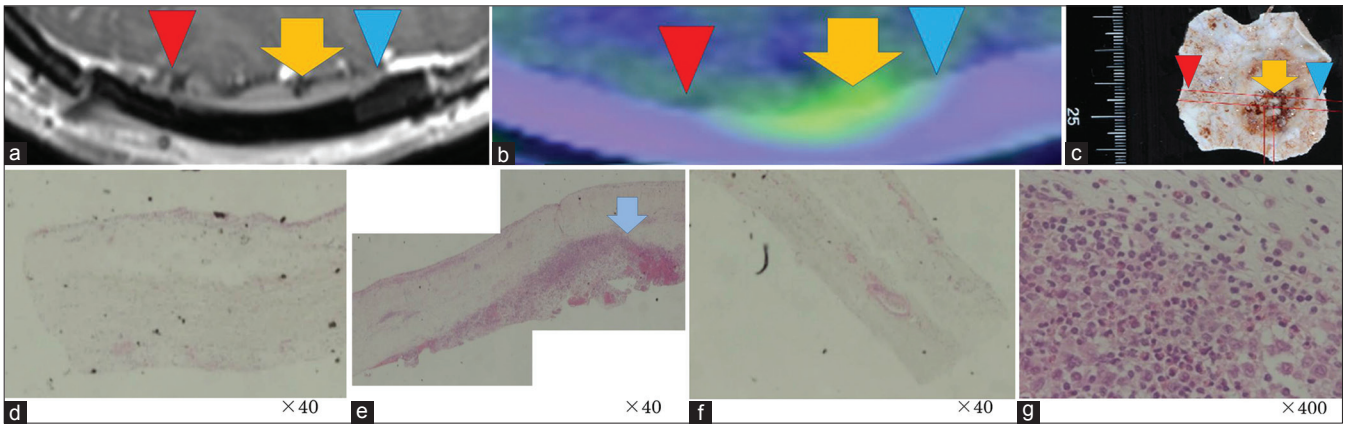

Figure 5: (a) Postoperative enhanced T1-weighted image. Red arrowhead (d). Yellow arrow (E). Blue arrowhead (F). (b) Preoperative methionine PET. Red arrowhead (D). Yellow arrow (E). Blue arrowhead (F). (c) A photograph of the surgical specimen. Red arrowhead (D). Yellow arrow (E). Blue arrowhead (F). (d) A photomicrograph of section D shows no tumor cells (hematoxylin and eosin stain, ×40) Blue arrow (G). (e) A photomicrograph of section E demonstrates tumor infiltration into the dura mater (H and E, ×40). (f) A photomicrograph of section F reveals no tumor cells (H and E, ×40). (g) Photomicrograph of section G demonstrates tumor infiltration into the dura mater (H and E, ×400)

was also substantially correspondingly observed in the part observed with nuclide accumulation.

Regarding LCH, as far as our research was able to show, there are no reports on Met-PET. Although resection was carried out in the present case including the dura mater observed with accumulation in Met-PET and subcutaneous tissues, the accumulation region and spreading of tumor cells were proximate. Because accumulation in the brain is poor in Met-PET compared with FDG-PET, regarding skull lesions such as the present case, it may be helpful in comprehending the spreading of tumor cells to the dura mater. Regarding treatment, resection has been determined as the standard for isolated lesions.[6] Chemotherapy and/or radiation therapy are recommended for cases of relapse following surgery and residual tumors.[6] When wide-ranging dura mater enhancing effects such as the present case are observed, the range of resection becomes problematic; however, it was believed that a further appropriate range may be excised by combining findings from Met-PET.

CONCLUSION

We experienced a surgical case of LCH of the skull in which an enhancing effect was observed in the dura mater upon MRI. Regarding cranial lesions, tumor cell invasion into the dura mater and subcutaneous tissue may be understood in detail by combining Met-PET.

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