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**REVIEW**

- 1140 COVID-19: Gastrointestinal manifestations, liver injury and recommendations  
*Ozkurt Z, Çınar Tanrıverdi E*

**ORIGINAL ARTICLE****Retrospective Study**

- 1164 Continuous intravenous infusion of recombinant human endostatin using infusion pump plus chemotherapy in non-small cell lung cancer  
*Qin ZQ, Yang SF, Chen Y, Hong CJ, Zhao TW, Yuan GR, Yang L, Gao L, Wang X, Lu LQ*
- 1172 Sequential sagittal alignment changes in the cervical spine after occipitocervical fusion  
*Zhu C, Wang LN, Chen TY, Mao LL, Yang X, Feng GJ, Liu LM, Song YM*
- 1182 Importance of the creation of a short musculofascial tunnel in peritoneal dialysis catheter placement  
*Lee CY, Tsai MK, Chen YT, Zhan YJ, Wang ML, Chen CC*
- 1190 Clinical effect of methimazole combined with selenium in the treatment of toxic diffuse goiter in children  
*Zhang XH, Yuan GP, Chen TL*
- 1198 Clinical study on the minimally invasive percutaneous nephrolithotomy treatment of upper urinary calculi  
*Xu XJ, Zhang J, Li M, Hou JQ*

**Observational Study**

- 1206 Comparison of diagnostic validity of two autism rating scales for suspected autism in a large Chinese sample  
*Chu JH, Bian F, Yan RY, Li YL, Cui YH, Li Y*
- 1217 Doctor-led intensive diet education on health-related quality of life in patients with chronic renal failure and hyperphosphatemia  
*Feng XD, Xie X, He R, Li F, Tang GZ*

**SYSTEMATIC REVIEWS**

- 1226 What are the self-management experiences of the elderly with diabetes? A systematic review of qualitative research  
*Li TJ, Zhou J, Ma JJ, Luo HY, Ye XM*

**META-ANALYSIS**

- 1242 Comparison of the clinical performance of i-gel and Ambu laryngeal masks in anaesthetised paediatric patients: A meta-analysis  
*Bao D, Yu Y, Xiong W, Wang YX, Liang Y, Li L, Liu B, Jin X*

## CASE REPORT

- 1255** Autogenous iliotibial band enhancement combined with tendon lengthening plasty to treat patella baja: A case report  
*Tang DZ, Liu Q, Pan JK, Chen YM, Zhu WH*
- 1263** Sintilimab-induced autoimmune diabetes: A case report and review of the literature  
*Yang J, Wang Y, Tong XM*
- 1278** Unicentric Castleman disease was misdiagnosed as pancreatic mass: A case report  
*Zhai HY, Zhu XY, Zhou GM, Zhu L, Guo DD, Zhang H*
- 1286** Igaratimod in treatment of primary Sjögren's syndrome concomitant with autoimmune hemolytic anemia: A case report  
*Zhang J, Wang X, Tian JJ, Zhu R, Duo RX, Huang YC, Shen HL*
- 1291** Primary central nervous system lymphoma presenting as a single choroidal lesion mimicking metastasis: A case report  
*Jang HR, Lim KH, Lee K*
- 1296** Surgical treatment of acute cholecystitis in patients with confirmed COVID-19: Ten case reports and review of literature  
*Bozada-Gutiérrez K, Trejo-Avila M, Chávez-Hernández F, Parraguirre-Martínez S, Valenzuela-Salazar C, Herrera-Esquivel J, Moreno-Portillo M*
- 1311** Hydrogen inhalation promotes recovery of a patient in persistent vegetative state from intracerebral hemorrhage: A case report and literature review  
*Huang Y, Xiao FM, Tang WJ, Qiao J, Wei HF, Xie YY, Wei YZ*
- 1320** Ultrasound-guided needle release plus corticosteroid injection of superficial radial nerve: A case report  
*Zeng Z, Chen CX*
- 1326** Inverted Y ureteral duplication with an ectopic ureter and multiple urinary calculi: A case report  
*Ye WX, Ren LG, Chen L*
- 1333** Multiple miscarriages in a female patient with two-chambered heart and situs inversus totalis: A case report  
*Duan HZ, Liu JJ, Zhang XJ, Zhang J, Yu AY*
- 1341** Chidamide combined with traditional chemotherapy for primary cutaneous aggressive epidermotropic CD8+ cytotoxic T-cell lymphoma: A case report  
*He ZD, Yang HY, Zhou SS, Wang M, Mo QL, Huang FX, Peng ZG*
- 1349** Fatal rhabdomyolysis and disseminated intravascular coagulation after total knee arthroplasty under spinal anesthesia: A case report  
*Yun DH, Suk EH, Ju W, Seo EH, Kang H*
- 1357** Left atrial appendage occlusion in a mirror-image dextrocardia: A case report and review of literature  
*Tian B, Ma C, Su JW, Luo J, Sun HX, Su J, Ning ZP*

- 1366 Imaging presentation of biliary adenofibroma: A case report  
*Li SP, Wang P, Deng KX*
- 1373 Multiple gouty tophi in the head and neck with normal serum uric acid: A case report and review of literatures  
*Song Y, Kang ZW, Liu Y*
- 1381 Toxic epidermal necrolysis induced by ritodrine in pregnancy: A case report  
*Liu WY, Zhang JR, Xu XM, Ye TY*
- 1388 Direct antiglobulin test-negative autoimmune hemolytic anemia in a patient with  $\beta$ -thalassemia minor during pregnancy: A case report  
*Zhou Y, Ding YL, Zhang LJ, Peng M, Huang J*
- 1394 External penetrating laryngeal trauma caused by a metal fragment: A Case Report  
*Qiu ZH, Zeng J, Zuo Q, Liu ZQ*
- 1401 Antegrade in situ laser fenestration of aortic stent graft during endovascular aortic repair: A case report  
*Wang ZW, Qiao ZT, Li MX, Bai HL, Liu YF, Bai T*
- 1410 Hoffa's fracture in an adolescent treated with an innovative surgical procedure: A case report  
*Jiang ZX, Wang P, Ye SX, Xie XP, Wang CX, Wang Y*
- 1417 Hemizygous deletion in the *OTC* gene results in ornithine transcarbamylase deficiency: A case report  
*Wang LP, Luo HZ, Song M, Yang ZZ, Yang F, Cao YT, Chen J*
- 1423 Langerhans cell histiocytosis presenting as an isolated brain tumour: A case report  
*Liang HX, Yang YL, Zhang Q, Xie Z, Liu ET, Wang SX*
- 1432 Inflammatory myofibroblastic tumor after breast prosthesis: A case report and literature review  
*Zhou P, Chen YH, Lu JH, Jin CC, Xu XH, Gong XH*
- 1441 Eustachian tube involvement in a patient with relapsing polychondritis detected by magnetic resonance imaging: A case report  
*Yunaiyama D, Aoki A, Kobayashi H, Someya M, Okubo M, Saito K*
- 1447 Endoscopic clipping for the secondary prophylaxis of bleeding gastric varices in a patient with cirrhosis: A case report  
*Yang GC, Mo YX, Zhang WH, Zhou LB, Huang XM, Cao LM*

**LETTER TO THE EDITOR**

- 1454 Rituximab as a treatment for human immunodeficiency virus-associated nemaline myopathy: What does the literature have to tell us?  
*Gonçalves Júnior J, Shinjo SK*

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## Langerhans cell histiocytosis presenting as an isolated brain tumour: A case report

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### Abstract

#### BACKGROUND

Langerhans cell histiocytosis (LCH) is a rare proliferative histiocyte disorder. It can affect any organ or system, especially the bone, skin, lung, and central nervous system (CNS). In the CNS, the hypothalamic-pituitary is predominantly affected, whereas the brain parenchyma is rarely affected. LCH occurring in the brain parenchyma can be easily confused with glioblastoma or brain metastases. Thus, multimodal imaging is useful for the differential diagnosis of these intracerebral lesions and detection of lesions in the other organs.

#### CASE SUMMARY

A 47-year-old man presented with a headache for one week and sudden syncope. Brain computed tomography (CT) and magnetic resonance imaging showed an irregularly shaped nodule with heterogeneous enhancement. On <sup>18</sup>F-fluorodeoxyglucose (<sup>18</sup>F-FDG) positron emission tomography/CT, a nodule with <sup>18</sup>F-FDG uptake and multiple cysts in the upper lobes of both lungs were noted, which was also confirmed by high-resolution CT. Thus, the patient underwent surgical resection of the brain lesion for further examination. Postoperative pathology confirmed LCH. The patient received chemotherapy after surgery. No recurrence was observed in the brain at the 12-mo follow-up.

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## CONCLUSION

Multimodal imaging is useful for evaluating the systemic condition of LCH, developing treatment plans, and designing post-treatment strategies.

**Key Words:** Langerhans cell histiocytosis; Brain neoplasms; Lung; Computed tomography; Magnetic resonance imaging; Positron emission tomography/computed tomography; Case report

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**Core Tip:** Langerhans cell histiocytosis (LCH) is a rare hematological disease characterized by a clonal proliferation of abnormal langerhans cells. It can affect any organ or system, especially the bone, skin, lung, and central nervous system (CNS). In the CNS, the hypothalamic-pituitary is predominantly affected, whereas the brain parenchyma is rarely affected. Cases of LCH involving the brain parenchyma and presenting as an isolated brain tumour have been reported, but all the reports lack complete multimodal imaging. In this manuscript, we have reported a case of LCH involving the brain parenchyma and bilateral lungs, which was assessed using computed tomography (CT), high-resolution CT, magnetic resonance imaging, and <sup>18</sup>F-fluorodeoxyglucose positron emission tomography/CT. Furthermore, we have reviewed the relevant literature.

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## INTRODUCTION

Langerhans cell histiocytosis (LCH) is an uncommon disease characterized by clonal proliferation of myeloid precursors that differentiate into cluster of differentiation (CD)1a<sup>+</sup>/CD207<sup>+</sup> (Langerin) cells in lesions[1,2]. It mainly affects children, with a reported incidence of 4-5 cases *per million children* aged < 15 years *per year*, while its incidence in adults is uncertain[3]. LCH may affect any organ or system, but it most frequently affects the bone, skin, pituitary, liver, spleen, hematopoietic system, lung, lymph nodes, and central nervous system (CNS)[4,5]. Cases of LCH involving the brain parenchyma and presenting as an isolated brain tumour have been reported, but all the reports lack complete multimodal imaging. Herein, we have reported a case of LCH involving the brain parenchyma and bilateral lungs, which was assessed using computed tomography (CT), high-resolution CT (HRCT), magnetic resonance imaging (MRI), and <sup>18</sup>F-fluorodeoxyglucose (<sup>18</sup>F-FDG) positron emission tomography (PET)/CT. Furthermore, we have reviewed the relevant literature.

## CASE PRESENTATION

### Chief complaints

A 47-year-old man was referred to our hospital with a headache for one week and sudden syncope in the morning.

### History of present illness

A 47-year-old man was referred to our hospital with a headache for one week and sudden syncope in the morning.

### History of past illness

The patient had no history of polyuria or polydipsia. No other illnesses were observed.



### **Personal and family history**

The patient had no known comorbidities or family history, but had a 15-year smoking history.

### **Physical examination**

No rash or positive neurological signs were found on physical examination.

### **Laboratory examinations**

Laboratory tests results showed increased in carcinoembryonic antigen (CEA) levels (7.03 ng/mL, reference: 0-5 ng/mL), with no other abnormal findings.

### **Imaging examinations**

Non-contrast brain CT showed irregularly shaped nodular foci with isodensity at the left frontal corticomedullary junction. Large patches of hypodense edema were noted in the adjacent white matter (Figure 1A and B represent the lateral ventricular and basal ganglia levels, respectively). Contrast-enhanced brain CT showed significant heterogeneous enhancement in the left frontal foci (Figure 1C and D, the same level as the Figure 1A and B). Coronal and sagittal views of contrast-enhanced CT images showed irregular morphology of the lesion and poor demarcation with the adjacent skull (Figure 1E and F). Bone window CT showed no abnormalities in the adjacent skull (Figure 1G and H, the same level as the Figure 1E and F). Subsequently, the patient underwent a brain MRI. Axial T1-weighted images (T1WI) showed heterogeneous hypointense lesions in the left frontal lobe (Figure 2A). Axial T2-weighted images (T2WI) showed a heterogeneously mixed hyperintensity signals with hypointense areas in the left frontal lobe lesion (Figure 2B). After administration of gadolinium, the lesion showed heterogeneous enhancement on axial (Figure 2C), coronal (Figure 2D), and sagittal T1WI (Figure 2E). No abnormalities were found on sagittal T1WI of the sellar region (Figure 2F).

Considering the elevated CEA levels and CT and MRI manifestations, further investigation was required to rule out brain metastases. Therefore, <sup>18</sup>F-FDG PET/CT was performed. Maximum-intensity-projection imaging showed a focal increase in <sup>18</sup>F-FDG uptake in the right maxillary sinus and multiple foci of increased <sup>18</sup>F-FDG uptake in the bilateral lung fields (Figure 3A). Axial (Figure 3B-D) and coronal (Figure 3E-G) views of the selected PET, non-enhanced CT (NE-CT), and fused PET/CT images showed moderately increased <sup>18</sup>F-FDG uptake in the left frontal nodule [the maximum standardized uptake value (SUVmax) of the lesions and surrounding tissues are shown in Supplementary Figure 1]. No abnormal <sup>18</sup>F-FDG uptake was observed in the sellar region (Supplementary Figure 2). Axial (Figure 3H-J) views of the selected PET, NE-CT, and fused PET/CT images showed multiple cysts with peripheral exudation in the upper lobes of bilateral lungs, with slightly increased <sup>18</sup>F-FDG uptake. HRCT was performed to further evaluate the pulmonary lesions. Axial (Figure 4A), coronal (Figure 4B), and sagittal (Figure 4C and D, left and right lungs, respectively) views of HRCT images showed multiple scattered small thick-walled irregular cysts and small nodules. Sinusitis was diagnosed in the right maxillary sinus. Bilateral lung manifestations should be differentiated from pulmonary LCH, but brain nodules are more difficult to diagnose and should be differentiated from gliomas.

### **Further diagnostic work-up**

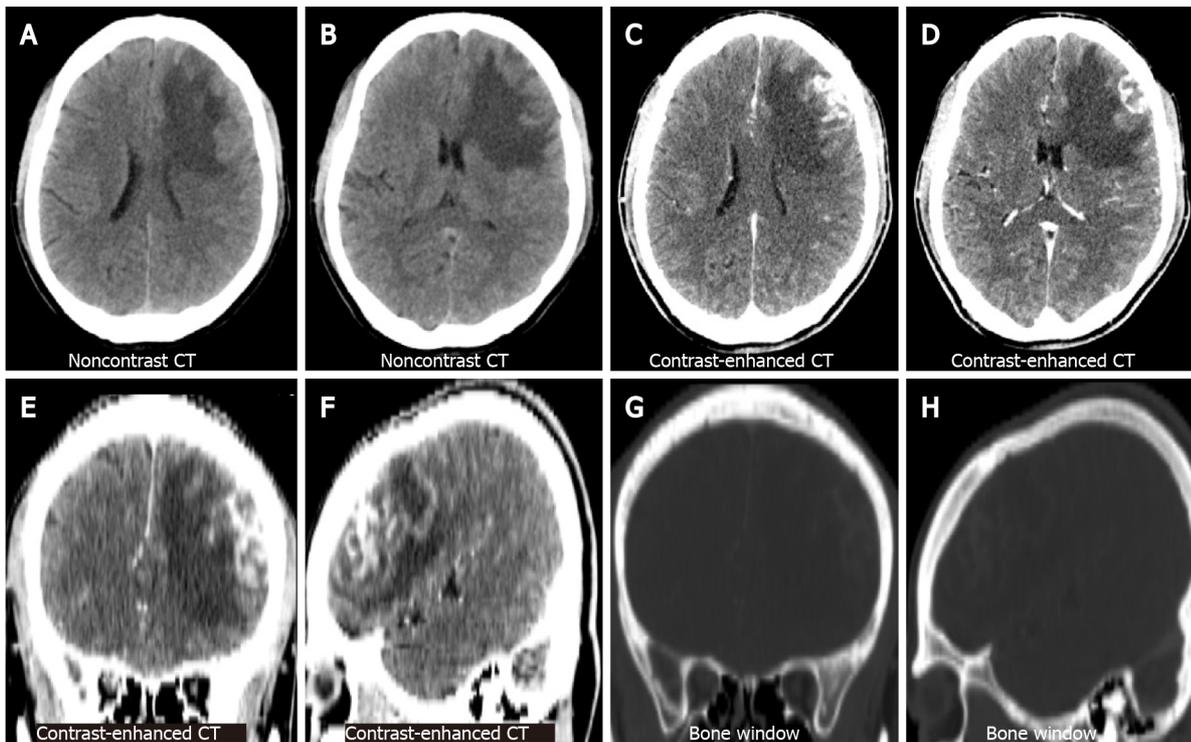
The patient underwent brain tumour resection. Gross examination showed that the specimen was a grey-brown solid tumour (Figure 5A) and the cut surface was grey-brown and grey-white (Figure 5B). Histopathological examination revealed mononucleated and multinucleated histocytes with abundant cytoplasm and slight staining (haematoxylin and eosin, magnification, × 200; Figure 5C). On immunohistochemistry, the specimen stained positive for S100, CD207 (Langerin), CD4, and CD1a, and negative for CD3 and CD20. Ki67 (MIB-1) index was slightly > 30%.

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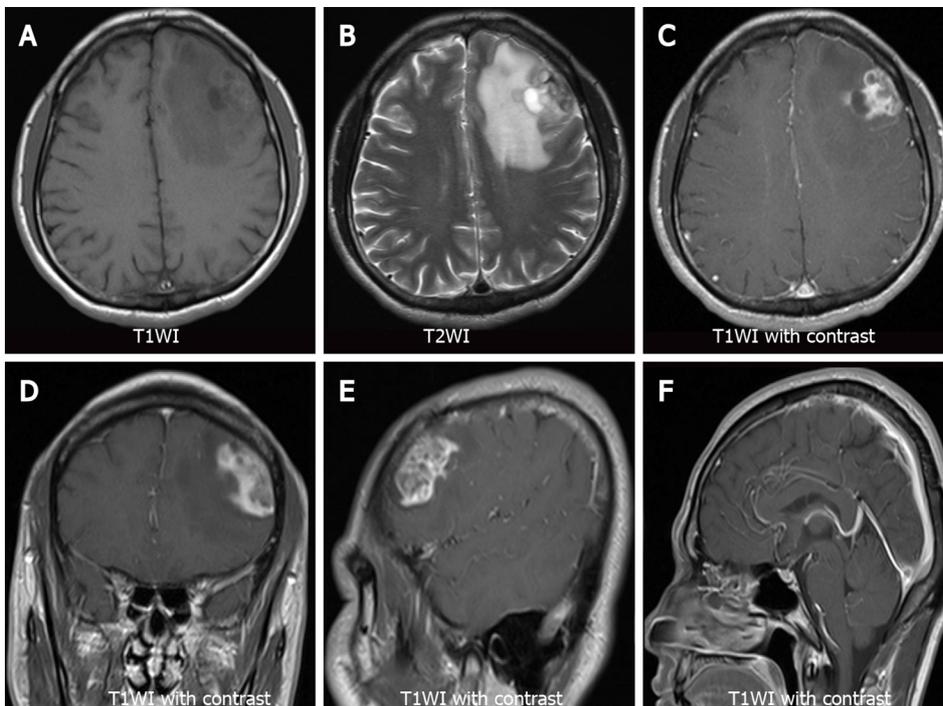
## **FINAL DIAGNOSIS**

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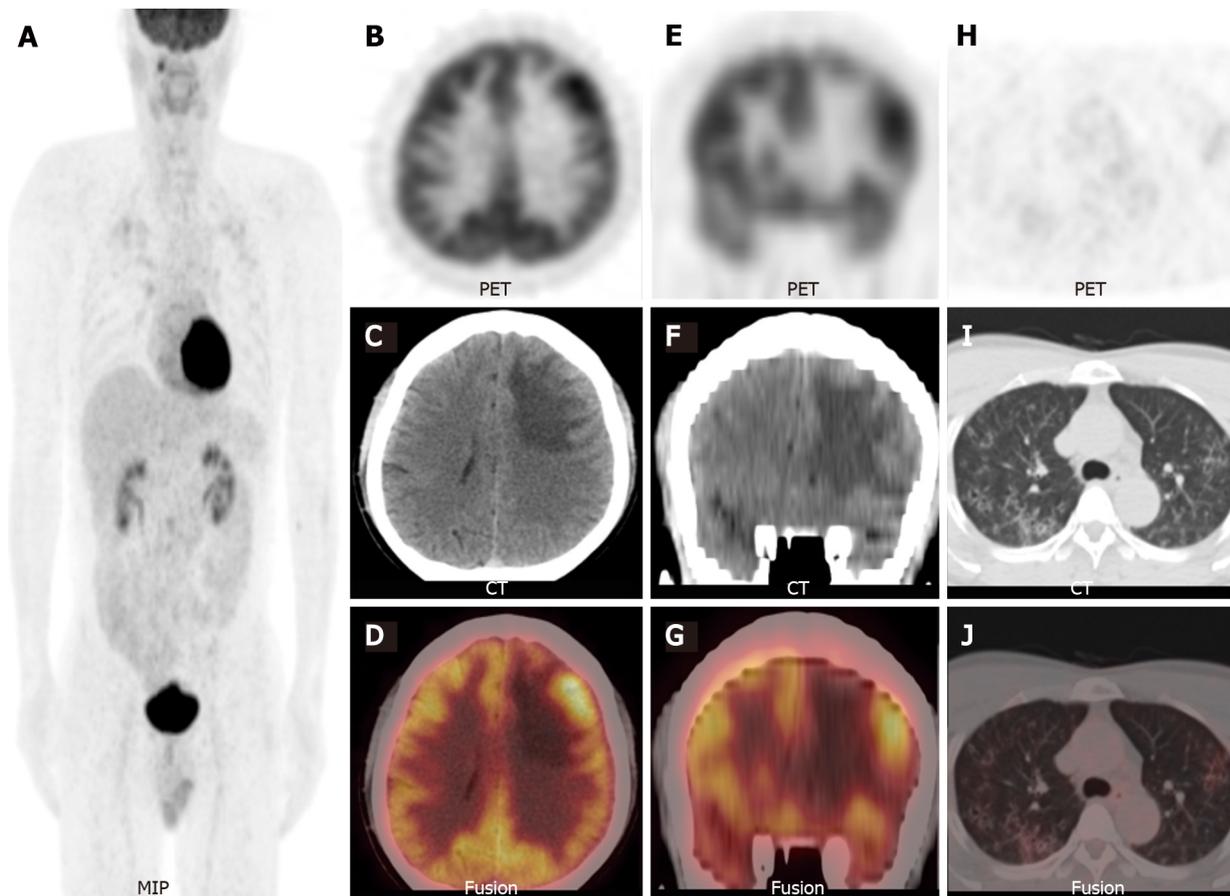
The final histological diagnosis was LCH.



**Figure 1 Brain computed tomography.** A and B: Represent the lateral ventricular and basal ganglia levels on non-contrast computed tomography (CT), respectively. An irregularly shaped nodule is observed in the left frontal lobe with large perifocal low-density oedema; C and D: Represent the same level as the former on contrast-enhanced CT. The nodules are significantly enhanced heterogeneously; E and F: Represent the coronal and sagittal views of the contrast-enhanced CT; G and H: Represent the same level as the former, with no abnormalities in the adjacent skull. CT: Computed tomography.



**Figure 2 Brain magnetic resonance imaging.** A: Axial T1-weighted images (T1WI) show heterogeneous hypo-intensity of the left frontal lobe lesion; B: Axial T2-weighted images (T2WI) show heterogeneously mixed signal of hyperintensity with hypointense areas of the left frontal lobe lesion; C-E: Axial, coronal, and sagittal views of T1WI with contrast agent administration show heterogeneous enhancement of the lesion; F: Sagittal T1WI show no abnormality in the sellar region. T1WI: T1-weighted images; T2WI: T2-weighted images.



**Figure 3**  $^{18}\text{F}$ -fluorodeoxyglucose positron emission tomography/computed tomography. A: Maximum-intensity-projection show a focal  $^{18}\text{F}$ -fluorodeoxyglucose ( $^{18}\text{F}$ -FDG) uptake lesion in the right maxillary sinus and multiple foci with  $^{18}\text{F}$ -FDG uptake in the bilateral lung field; B-G: Axial and coronal views of the selected positron emission tomography (PET), non-enhanced computed tomography (NE-CT), and fused PET/CT images show the left frontal lesion with  $^{18}\text{F}$ -FDG uptake (SUVmax 9.5, arrowheads); H-J: Axial views of the selected PET, NE-CT, and fused PET/CT images show multiple cysts with peripheral exudation in the upper lobes of bilateral lungs, with slightly increased  $^{18}\text{F}$ -FDG uptake (SUVmax 3.2). MIP: Maximum-intensity-projection; PET: Positron emission tomography; CT: Computed tomography.

## TREATMENT

The patient received chemotherapy (vindesine and prednisone acetate) after surgery.

## OUTCOME AND FOLLOW-UP

No recurrence was observed on brain MRI at the 12-mo follow-up ([Supplementary Figure 3](#)).

## DISCUSSION

LCH involving the hypothalamic-pituitary or skull is not uncommon, but involvement of the brain parenchyma, such as the frontotemporal lobe, is rare. As of January 2019, fewer than 30 cases have been reported in the PubMed database ([Table 1](#))[6-8]. We reviewed the relevant PubMed literature from 1990 to May 2021 and found 16 cases of brain parenchymal LCH with imaging data. The mean age was 31 years (95% confidence interval: 21.5-41.2). The male-to-female ratio was 14:2, which is consistent with that reported in previous literature reviews of intracerebral LCH, but higher than that in children with LCH[3,9,10]. The lesions were mostly located in the frontotemporal lobe (14 cases), particularly in the frontal lobe. The clinical presentation of LCH is non-characteristic and varies depending on the site. Most cases showed non-specific symptoms of mass effect such as headache, seizures, hemiparesis, and/or sensory disturbances. MRI findings without contrast were also largely non-specific.

**Table 1** Summary of 16 cases with brain parenchymal langerhans cell histiocytosis with imaging data

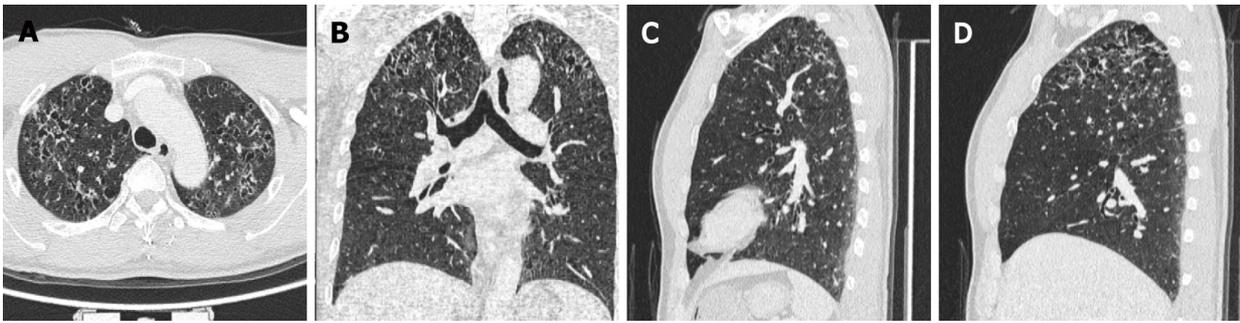
| Ref.                                      | Age/sex   | Diameter (cm) <sup>1</sup> | Location (Lobe)                                                       | MRI Finding        |                                       |                                             | <sup>18</sup> F-FDG PET/CT finding             |
|-------------------------------------------|-----------|----------------------------|-----------------------------------------------------------------------|--------------------|---------------------------------------|---------------------------------------------|------------------------------------------------|
|                                           |           |                            |                                                                       | T1WI               | T2WI                                  | T1WI with contrast                          |                                                |
| Caresio <i>et al</i> [20], 1991           | 29/Male   | 3.0                        | Right temporal lobe                                                   | Hypointense        | Hyperintense                          | Ringlike enhancement                        | NA                                             |
| Itoh <i>et al</i> [21], 1992              | 7/Male    | NA                         | Right frontal lobe                                                    | Hypo-/iso-intense  | Hyperintense                          | Intense enhancement                         | NA                                             |
| Bogaert <i>et al</i> [22], 1994           | 40/Female | NA                         | Left parietal lobe                                                    | Hypointense        | Hyperintense                          | Intense enhancement                         | NA                                             |
| Vital <i>et al</i> [23], 1996             | 32/Female | NA                         | Right insula lobe                                                     | NA                 | Hyperintense                          | Intense enhancement                         | NA                                             |
| Grant <i>et al</i> [24], 1999             | 20/Male   | 3.5                        | Right temporal lobe                                                   | NA                 | NA                                    | Intense enhancement                         | NA                                             |
| Katati <i>et al</i> [25], 2002            | 36/Male   | NA                         | Left temporal lobe                                                    | Hypointense        | NA                                    | Intense enhancement                         | NA                                             |
| Cagli <i>et al</i> [9], 2004              | 24/Male   | 1.5                        | Left temporal lobe                                                    | Hypointense        | NA                                    | Intense enhancement                         | NA                                             |
| Yamaguchi <i>et al</i> [26], 2004         | 2/Male    | NA                         | Multiple lesions/bilateral frontal and temporal lobes                 | NA                 | NA                                    | Intense enhancement                         | NA                                             |
| Rodríguez-Pereira <i>et al</i> [10], 2005 | 30/Male   | 5.0                        | Left frontal lobe                                                     | NA                 | NA                                    | Gyral enhancement                           | NA                                             |
| Rodríguez-Pereira <i>et al</i> [10], 2005 | 65/Male   | 2.5                        | Left parietal lobe                                                    | NA                 | NA                                    | Peripheral enhancement                      | NA                                             |
| Dieter[27], 2017                          | 4/Male    | 2.0 <sup>1</sup>           | Multiple lesions, right frontal, and parietal lobe                    | Iso-/hyper-intense | Hypointense                           | Uniform enhancement                         | NA                                             |
| Cai <i>et al</i> [6], 2014                | 23/Male   | 4.1                        | Right frontal lobe                                                    | Hypo-/iso-intense  | Iso-/hyper-intense                    | Moderate to intense homogeneous enhancement | NA                                             |
| Dardis <i>et al</i> [28], 2015            | 64/Male   | NA                         | Multiple lesions, left frontal and right temporal lobe, and brainstem | NA                 | Hyperintense                          | Patchy enhancement                          | NA                                             |
| Kim <i>et al</i> [7], 2018                | 36/Male   | 3.0                        | Right frontal lobe                                                    | Isointense         | Hyperintense                          | Heterogeneous enhancement                   | NA                                             |
| Bärtschi <i>et al</i> [8], 2019           | 42/Male   | NA                         | Right insular lobe                                                    | NA                 | Hyperintense                          | Intense enhancement                         | NA                                             |
| Current case                              | 47/Male   | 3.6                        | Left frontal lobe                                                     | Hypointense        | Hyperintensity with hypointense areas | Heterogeneous enhancement                   | Lesion SUVmax 9.5 and contralateral SUVmax 8.1 |

<sup>1</sup>The largest lesion.

NA: Not available; PET: Positron emission tomography; CT: Computed tomography; T1WI: T1-weighted images; T2WI: T2-weighted images; MRI: Magnetic resonance imaging.

Nonetheless, MRI showed hypointensity on T1WI and hyperintensity on T2WI in most cases. After administration of gadolinium, most cases showed intense homogeneous or heterogeneous enhancement. Another characteristic feature is sulcal enhancement around the lesion[6,7]. MRI images showed leptomeningeal involvement near the lesions in several cases, as reported by Kim *et al*[7]. This may be a characteristic sign of brain parenchymal LCH, but it needs to be confirmed in more cases.

There are no previous reports of <sup>18</sup>F-FDG PET/CT for assessing the metabolic activity of brain parenchymal LCH. To our knowledge, our case report is the first with a PET/CT description. The SUVmax of the brain lesion was approximately 9.5, which was similar to the SUVmax of LCH lesions involving other regions reported in the literature[11]. Additional bilateral lung lesions were found, and pulmonary manifestations were decisive for diagnosis[12]. As 30% patients with LCH present with multi-organ system involvement, it is important to detect involvement of other tissues (such as the bone, soft tissue, the CNS, or the lungs)[3,13]. Single or isolated brain lesions



**Figure 4 High-resolution computed tomography of the lung.** Axial, coronal, and sagittal (the left and right lungs, respectively) views of high-resolution computed tomography images show multiple scattered small thick-walled irregular cysts as well as small nodules. A: Axial; B: Coronal; C: Left lungs; D: Right lungs.



**Figure 5 Histopathological images.** A: The specimen is a greyish brown and greyish dark solid tumour; B: The cut surface is greyish brown and greyish white; C: Histopathological examination reveal mononucleated and multinucleated histiocytes with abundant cytoplasm and light staining (haematoxylin and eosin, magnification,  $\times 400$ ).

have previously been reported based on only brain CT or MRI, without whole-body scans[6,7]. Without whole-body evaluation, reports of isolated brain lesions may be non-rigorous or biased. More recent studies that performed whole-body evaluations have identified a higher rate of focal LCH lesions than that previously reported[14,15]. Therefore, PET/CT or PET/MRI seems to be more appropriate for evaluating this disease[16]. This is especially true for combined bone and lung lesions as some case without obvious symptoms are incidentally detected; they may be missed by relying solely on radiography or CT[8]. Several studies have confirmed the diagnostic value of systemic scans, such as PET/CT or PET/MRI for LCH[14,15,17,18]. The diagnostic evaluation of LCH plays a crucial role in treatment planning. PET/CT or PET/MRI can be used to assess multiple foci throughout the body, guide biopsy sites, and assist with post-treatment strategies.

Based on prospective trials, the combination of vinblastine plus prednisolone is the most commonly used induction chemotherapy regimen and is administered over six weeks[19].

## CONCLUSION

As a systemic disease, LCH has the potential to involve the brain parenchyma, and its diagnosis is extremely challenging. The use of multimodal imaging or whole-body imaging, combined with the manifestation of lesions at other sites, can be helpful in the diagnosis of this disease. Moreover, multimodality imaging is useful for assessing the systemic status of LCH, developing treatment plans, and evaluating post-treatment strategies.

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