

CASE REPORT

“The unpredictable brain tumor”

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Abstract

Histiocytosis is a group of rare diseases with vast imaging findings, few of which are distinctive and characteristic that help to differentiate each one of them. Therefore, typical imaging appearances must be recognized to include the possibility in the differential diagnosis, whenever considered pertinent. Hereby, we present one such unique case of histiocytosis in a 26-year-old female, which involved intertwined and overlapping features of radiological findings.

Key words: Hairy kidney sign; histiocytosis; meningioma; pituitary; soft tissue thickening

Introduction

Histiocytic disorders are a group of diseases derived from macrophages and dendritic cells. They result in a wide range of clinical conditions that are restricted primarily to children but can affect adults as well. Histiocytosis is divided into-1) Langerhans cell histiocytosis. 2) Non -Langerhans cell histiocytic disorders [Figure 1].

Case History

A 26-year-old female presented in our OP department with on and off headache since the last 1 year and generalized weakness. The patient was referred for the magnetic resonance imaging (MRI) of brain. MRI brain showed a well-defined extra-axial dural-based lesion with CSF cleft in left frontal parasagittal region. The lesion appeared hypointense on T1WI, hyperintense on T2WI, and fluid-attenuated inversion recovery (FLAIR) sequences and showed intense homogenous enhancement [Figure 2]. Meningioma was given as a radiological diagnosis. Common meningioma mimickers are hemangiopericytoma, solitary fibrous tumor, dural metastases, and mucosa-associated lymphoid tissue (MALT) lymphoma of the dura.

- Hemangiopericytoma-Mostly they have a tendency to erode the adjacent bone.
- Solitary fibrous tumor-usually they show patchier low signal intensity or “yin-yang” appearance of separate areas with low-signal and high-signal intensity on T2.
- Dural metastases- history of known malignancy should be enquired.
- MALT lymphoma of the dura-is rare, difficult to distinguish from meningioma on imaging.

The patient underwent surgery and the specimen was sent for biopsy. The biopsy showed lymphocytes, histiocytes, plasma cells, and emperipolesis. Emperipolesis is a phenomenon in which histiocytes phagocytize lymphocytes, plasma cells, erythrocytes, or polymorphonuclear leukocyte. It is a characteristic feature of Rosai–Dorfman’s disease (one of the Non -Langerhans cell histiocytosis), however, it is not pathognomonic of it. Therefore, the provisional diagnosis was given as Rosai–Dorfman’s disease while immunohistochemistry report was awaited. Meanwhile, the patient was sent to positron emission tomography computed

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Cite this article as: Singh S, Raghavan B, Govindaraj J, Geethapriya S. “The unpredictable brain tumor”. Indian J Radiol Imaging 2020;30:379-82.

Received: 22-Feb-2020

Revised: 24-May-2020

Accepted: 28-May-2020

Published: 15-Oct-2020

Access this article online

Quick Response Code:



Website:
www.ijri.org

DOI:
10.4103/ijri.IJRI_66_20

tomography (PET-CT) for the evaluation of the whole spectrum of the disease.

PET-CT of the whole body showed hypermetabolic mildly enlarged pituitary gland (SUV max 6.7) [Figure 3]. No pituitary lesion was noted either on PET-CT or MRI. Mildly increased 2-fluorodeoxyglucose (FDG) uptake was noted in the soft tissue thickening within the intraconal space of the left orbit (SUV max 5.2) [Figure 4]. Mild smooth interlobular septal thickening was seen in bilateral upper lung lobes with no significant FDG uptake [Figure 4]. Mildly enhancing soft tissue thickening with low-grade FDG uptake was seen encasing the great vessels (aorta, pulmonary trunk, and inferior vena cava), around the right atrium and in the right perihilar region [Figure 5]. Prominent mediastinal and right hilar nodes with mildly increased FDG uptake were also noted [Figure 5]. Enhancing soft tissue with low-grade FDG avid was seen around renal hila, encasing the proximal ureter and perinephric soft tissue around both kidneys (left > right). This appearance is characteristically called “Hairy

kidney sign” [Figure 6]. Sclerosis with increased FDG uptake (SUVmax 4.5) was seen within both femoral heads in subarticular surface and extending along the trabeculae [Figure 7]. Similar minimal sclerosis was also noted within both humeral heads.

All these findings were pointing towards another type of Non-Langerhans cell histiocytosis i.e., Erdheim–Chester disease (while the biopsy report was suggesting Rosai–Dorfman’s disease). The final diagnosis was clinched by immunohistochemistry report, which showed positivity for S100 and CD68, which is a characteristic feature of Rosai–Dorfman disease (Erdheim–Chester disease shows positivity for CD68 but not for S100). Imaging and clinical features in favor of Rosai–Dorfman and Erdheim–Chester disease in our patient are given in Table 1.

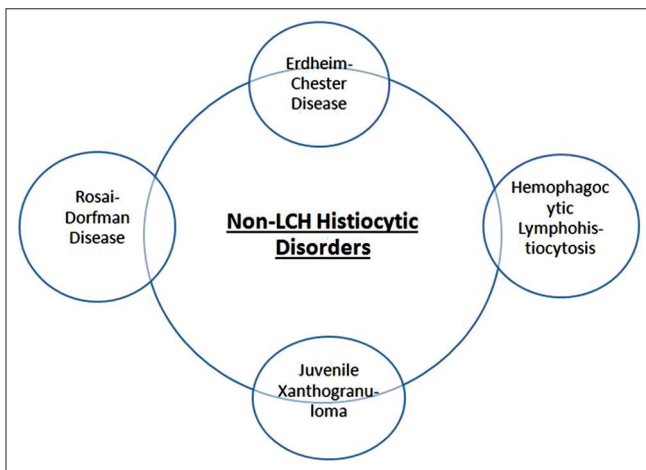


Figure 1: Different types of non-Langerhans cell histiocytosis

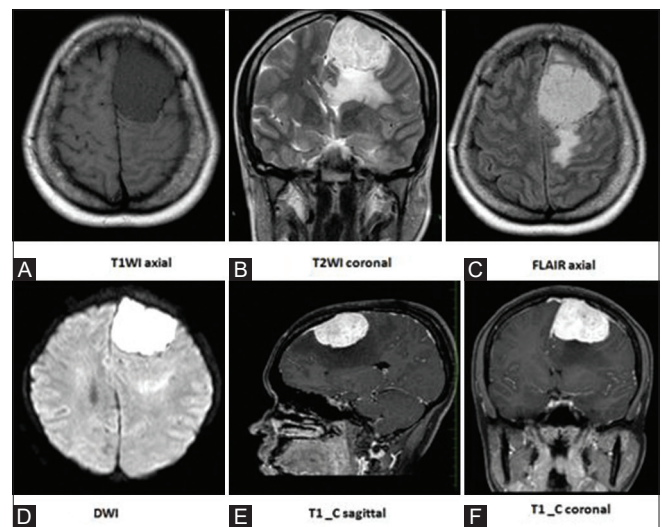


Figure 2 (A-F): MRI brain showed a well-defined extra axial dural-based T1 hypointense (A), T2 and FLAIR hyperintense (B and C) lesion with CSF cleft, showing restricted diffusion (D) and intense homogenous enhancement with dural tail (E and F) in left frontal parasagittal region

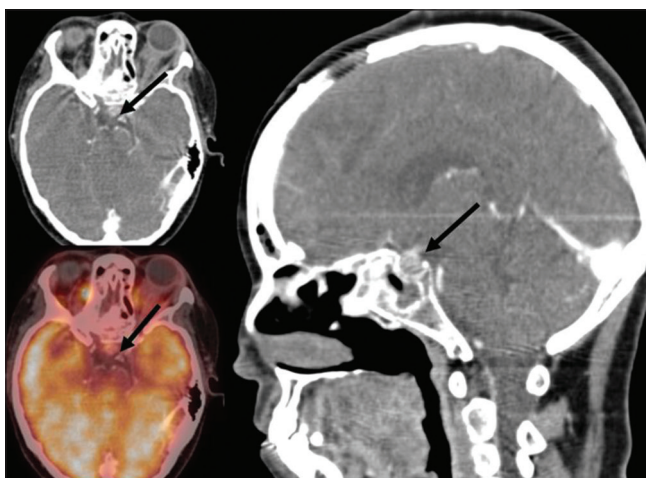


Figure 3: PET-CT of whole body showed mildly enlarged pituitary gland showing increased FDG uptake (SUV max 6.7)(arrows)

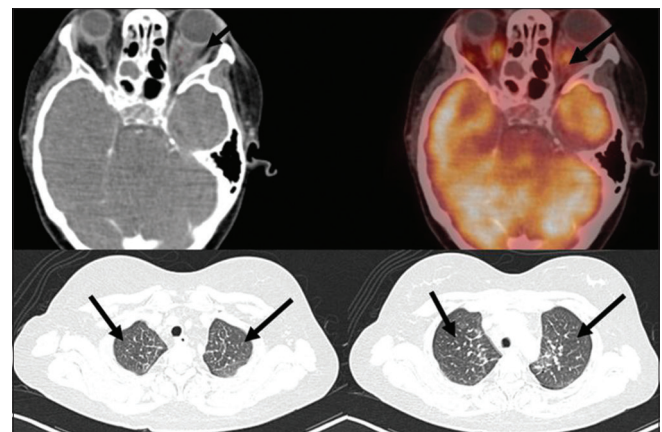


Figure 4: PET-CT of whole body showed mildly increased FDG uptake in the soft tissue thickening within intraconal space of left orbit (SUV max 5.2)(above image)(arrows). Mild smooth interlobular septal thickening was seen in bilateral upper lung lobes with no significant FDG uptake (below image)(arrows)

Table 1: Imaging and clinical features in favor of Rosai-Dorfman and Erdheim-Chester disease in our patient.

Points in favor of Rosai-Dorfman Disease

1. More common in children and young adults.
2. The epidural and subdural compartments are more commonly seen than Erdheim-Chester disease. Frequent locations are cerebral convexities, parasagittal, and petroclival regions.
3. Prominent lymph nodes, anaemia.
4. No cardiac involvement or diabetes insipidus.
5. Mild smooth interlobular septal thickening in lungs.
6. Orbit involvement.
7. Emperipolesis.
8. Positivity for both S100 and CD68.

Points in favor of Erdheim-Chester Disease

- More common in adults.
- Most commonly involved region in Central nervous system is Sellar region. Diabetes insipidus is the most common presenting symptom. The epidural and subdural compartments are less commonly involved than Rosai-Dorfman disease.
- Rind of tissue surrounding kidneys and hilum-*Hairy kidney Sign*.
 - Soft tissue thickening encasing the mediastinal great vessels and heart.
 - Pituitary gland is enlarged.
 - Orbit involvement.
 - Positivity for CD68 but not for S100.
 - Sclerosis within both femoral heads.

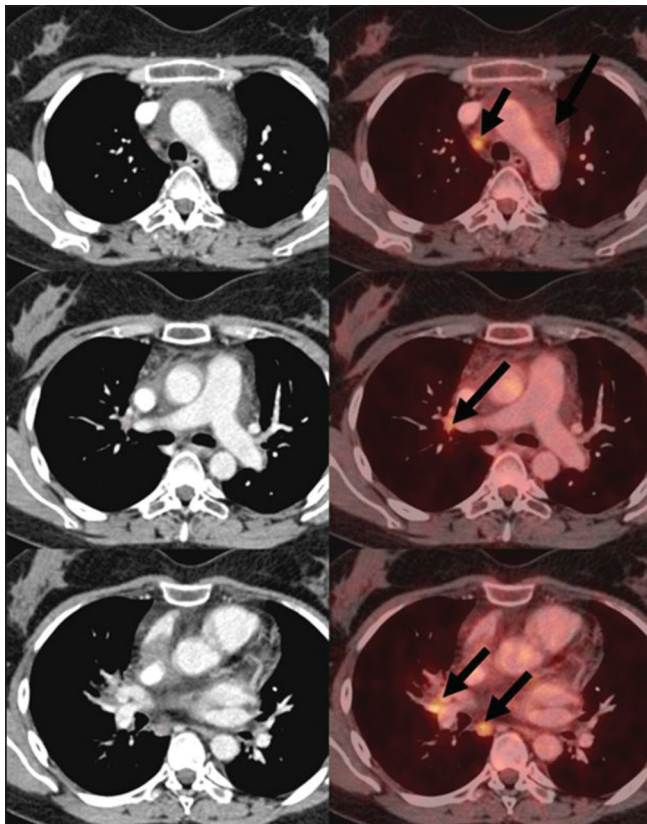


Figure 5: PET-CT of whole body showed mildly enhancing mediastinal soft tissue thickening with low-grade FDG uptake encasing the great vessels (aorta, pulmonary trunk, and inferior vena cava), around the right atrium and in right perihilar region. Prominent mediastinal and right hilar nodes with mildly increased FDG uptake were also noted (arrows)

Discussion

There are four types of Non -Langerhans cell histiocytic disorders:

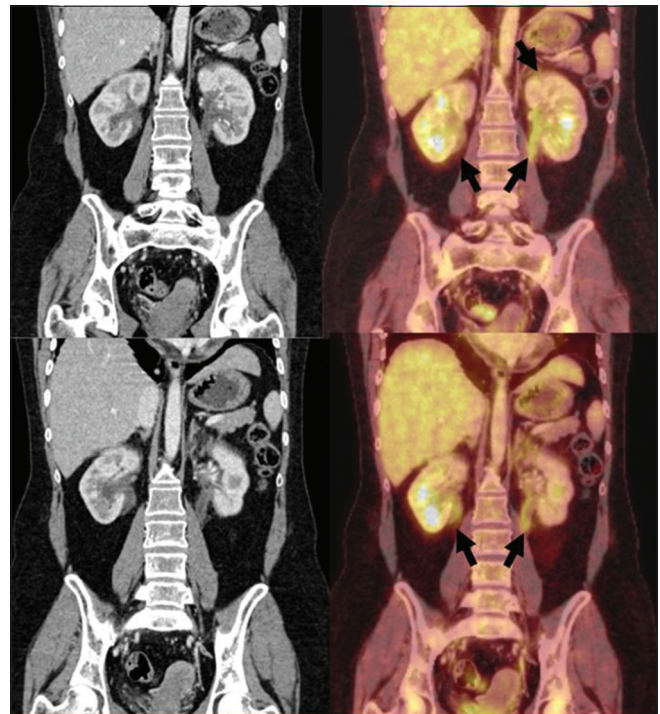


Figure 6: Enhancing soft tissue within renal hila, encasing the proximal ureter and perinephric soft tissue around both kidneys-*"Hairy kidney sign"*(arrows)

- A.) Juvenile Xanthogranuloma: Dermal manifestations are most common (majority present <1 year of age); diagnosis is usually made clinically.
- B.) Hemophagocytic Lymphohistiocytosis: presents with infections or prolonged immunosuppression. Diagnosis is confirmed when 5 of 8 criteria are present:-fever, splenomegaly, cytopenia (at least 2 cell lineages), increased TG/fibrinogen, increased ferritin, hemophagocytosis, decreased natural killer cells, and increased IL-2 receptor cells.

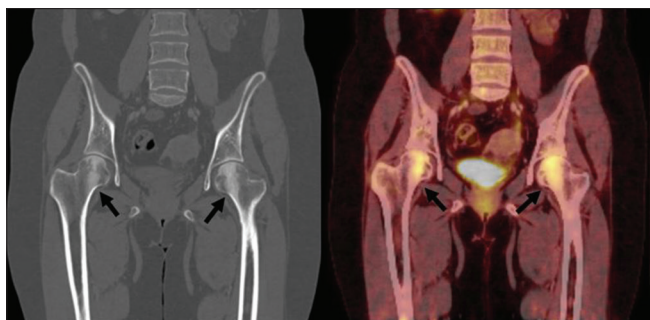


Figure 7: Increased FDG uptake (SUVmax 4.5) was seen within the sclerosis in both the femoral heads in subarticular surface and extending along the trabeculae (arrows)

C.) Erdheim–Chester Disease: more common in adults. The most common region involved in central nervous system is the sellar region. Diabetes insipidus is the most common presenting symptom in CNS involvement.^[1] The epidural and subdural compartments are less commonly involved than Rosai–Dorfman disease.

Bilateral and symmetrical osteosclerosis is seen in long bones as diffuse/patchy opacities, medullary sclerosis, and cortical thickening.^[2]

The homogeneous rind of tissue is commonly seen surrounding kidneys and renal hilum, the appearance known as Hairy kidney Sign.

Mediastinal soft tissue infiltration is observed frequently. Pulmonary centrilobular nodules and cysts are often seen. Cardiac involvement is noted in the form of fibrosis, pericardial thickening or effusion, and cardiomyopathy.^[3]

Orbit's involvement is also commonly noted. Histopathological features include a mononuclear infiltrate consisting of lipid-laden, foamy histiocytes, showing CD68 positivity but not S100.^[1]

D.) Rosai–Dorfman disease: More common in children and young adults. Fever, elevated erythrocyte sedimentation rate, and anemia are the common symptoms.^[2]

The epidural and subdural compartments are more commonly involved than Erdheim–Chester disease. Frequent locations are cerebral convexities, parasagittal, and petroclival regions.^[2] Interestingly, the majority of patients with CNS disease do not have lymphadenopathy. Multicentric lytic lesions are commonly seen. Commonly involved regions are skull, tibia, and femur.

In the abdomen, less commonly retroperitoneal and inguinal nodes are involved.

Thoracic manifestations are in the form of pulmonary nodules, septal, and interstitial thickening. Cardiac involvement is not so common. Painless lymphadenopathy is frequently seen.

The most commonly involved nodes are cervical lymph nodes followed by mediastinal and axillary lymph

nodes.^[2] The orbits are also a well-documented site of involvement.

Histopathological analysis reveals an infiltrate of lymphocytes, histiocytes, plasma cells, and emperipolesis. It is a phenomenon in which histiocytes phagocytize lymphocytes, plasma cells, erythrocytes, or leukocyte. This is a characteristic but not pathognomonic of Rosai–Dorfman's disease. Immunohistochemistry shows positivity for both S100 and CD68.^[4]

Conclusion

Histiocytosis is a group of rare diseases with some peculiarities that help to differentiate each one of them. They have a wide spectrum of clinical manifestations and radiologic appearances. Sometimes, imaging pictures can be similar. Radiology plays a large role in the diagnosis of the full spectrum of the disease and follow-up. Therefore, typical imaging appearances must be recognized to include the possibility in the differential diagnosis, whenever considered pertinent. However, the key to successful therapy is accurate identification at the tissue level and appropriate staging i.e., pathological, radiological, and clinical correlation is paramount.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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