Egyptian Journal of Radiology and Nuclear Medicine

CASE REPORT Open Access

Erdheim–Chester disease: with neurological manifestation and multisystem involvement: case report and radiological review

Varsha Rangankar[1], Pranav Ajmera[1*], Neha Agarwal[2], Tushar Kalekar[1], Pratiksha Yadav[1] and M. Suhas[1]

Abstract

Background: Erdheim–Chester disease (ECD) is a rarely encountered idiopathic systemic form of non-Langerhans cell histiocytosis. The clinical manifestations of ECD are highly heterogeneous, ranging from unifocal forms to life-threatening multisystem involvement. Patients with CNS involvement often do not show clinical remission.

Case presentation: We present a case of a 60-year-old male patient with worsening complaints of loss of balance, involuntary jerky movements, emotional lability and scanning speech developing over a period of 5 years. Magnetic resonance imaging of the brain at present institute revealed signal abnormalities in the midbrain, pons, cerebellar peduncles and cerebellar white matter with mineral deposition and volume loss in the bilateral basal ganglia and midbrain. Positron emission tomography–computed tomography of chest and abdomen revealed 18-fluorodeoxyglucose avid soft tissues lesion in the retroperitoneum involving bilateral perinephric spaces with intra-renal sinus extension, in pre- and paraaortic regions with enlargement of both adrenal glands. The radiographs of the long bones revealed multiple areas of sclerosis. The suspected diagnosis of ECD was confirmed on histopathology. ECD is a rare disease and has a predilection towards middle-aged males and is usually diagnosed late after the onset of initial symptoms.

Conclusions: Our case was an atypical presentation of an extremely rare disease, presenting with ataxia and choreoathetoid movements linked to ferromagnetic deposition on brain scans. Multiple other imaging feature characteristics of the disease like the hairy kidney sign, coated aorta sign, skeletal, extra-skeletal and central nervous system manifestations were noted in this single patient.

Keywords: Erdheim–Chester disease, 18-FDG PET, Hairy kidney sign, Choreoathetosis, Ataxia, Coated aorta sign

Background

Erdheim–Chester disease (ECD) is an infrequently encountered idiopathic systemic form of non-Langerhans cell histiocytosis [1, 2]. It is more frequent in middle-aged males mainly in the 5th–7th decade. However, multiple cases have been reported on extremes of the spectrum from 7 to 84 years of age [3, 4] with variable presentation. Specifically, bone pain is the most common symptom documented at initial presentation, but multisystem involvement is common, and over 50% of cases have extra-skeletal manifestations [4, 5]. The presence of constitutional symptoms like fever, loss of weight and night sweats can be misdiagnosed as tuberculosis, especially in developing nations. The diagnosis is confirmed on histopathology, which shows typical findings of infiltrates of lipid-laden macrophages and multinucleated giant cells. Immunohistochemical (IHC) staining reveals CD-68 positivity with the concurrent absence of CD1a and Birbeck granules [1, 5].

The number of published cases was barely around 500 till a decade back, though more such cases have been

*Correspondence: pranavajmera@gmail.com
[1] Department of Radiology, Dr. D.Y. Patil Medical College, Hospital and Research Center, DPU, Sant Tukaram Nagar, Pune 411018, India
Full list of author information is available at the end of the article

documented with the formation of the ECD registry [1, 6]. However, the actual frequency of encountering ECD in the clinical setting is extremely rare, and its varied multisystem presentation often leads to incorrect and delayed diagnosis. Our index case with an extremely rare presentation of ataxia and choreoathetoid movements suffered for multiple years before the diagnosis was finally established. Multiple other imaging feature characteristics of the disease like the hairy kidney sign, coated aorta sign, skeletal, extra-skeletal and central nervous system manifestations were noted in this single patient.

Case presentation

A 60-year-old male patient presented with complaints of loss of balance for 5 years and involuntary jerky movements in the last 2 years. The patient also complained of emotional lability and scanning of speech while speaking for 1.5 years. He was a known hypertensive on medication. He did not have any relevant family history of similar complaints. The previous magnetic resonance imaging (MRI) of the brain done for 4 years revealed no abnormality. The patient visited multiple hospitals where he was treated first as a case of pure cerebellar ataxia and later labelled as frontal lobe syndrome, and subsequently prescribed medications such as citicoline, angiotensin receptor blockers, carbamazepine and a selective serotonin reuptake inhibitor (SSRI) to protect against neural damage. While the patient improved and his symptoms stabilized for some time, they gradually started re-progressing for which he visited our hospital.

Informed consent was acquired from the patient to share the details.

Workup

MRI brain of the patient revealed diffuse enlargement of pons with ill-defined T2/FLAIR hyperintense and T1 iso to hypointense signal intensities in the midbrain, pons, superior and middle cerebellar peduncles, and cerebellar white matter (Fig. 1A–E). These revealed no diffusion restriction, blooming on susceptibility-weighted images and no post-contrast enhancement. Generalized cerebral and cerebellar atrophy was seen. The pituitary gland was enlarged with sellar widening and thickened infundibulum (Fig. 1F). The patient was labelled as progressive cerebellar ataxia with chorea and autonomic involvement with possible pituitary macroadenoma. Markedly hypointense areas of signal dropouts were present in the bilateral basal ganglia, substantia nigra, red nuclei (Fig. 2A–C) and, to a lesser extent, in dorsal thalami and dentate nuclei on susceptibility-weighted images showing bright signals on filtered phase images (Fig. 2D). These were thought to be due to mineral likely iron deposition. Atrophy of the bilateral basal ganglia and dentate nuclei with a reduction in the volume of the midbrain was also present. The possibility of neurodegenerative disease, likely neurodegeneration with brain iron accumulation (NBIA), was considered. The follow-up MRI brain performed after 1 month revealed persistent signal abnormalities in basal ganglia, brainstem, cerebellum and cerebellar peduncles, though the pontine swelling had regressed. The possibility of neurodegenerative disease, likely neurodegeneration with brain iron accumulation (NBIA), was considered.

The complete blood panel revealed mild microcytic hypochromic anaemia with increased red blood cell (RBC) distribution width and elevated absolute lymphocyte and monocytes. The total iron-binding capacity and serum iron were reduced. Serum ferritin and ceruloplasmin were within normal limits. An autoimmune panel including the anti-thyroid peroxidase antibodies, anti-gliadin, anti-GAD 65, NMDA receptor antibodies was negative. A nerve conduction velocity test revealed findings of acute motor axonal neuropathy. Testing for the Huntingtin and spinocerebellar ataxia gene nucleotide repeats also was negative.

Computed tomography (CT) study of the chest and abdomen of the patient was performed. CT abdomen showed ill-defined mildly enhancing soft tissues lesions in the retroperitoneum involving bilateral perinephric spaces (hairy kidney sign), showing intra-renal sinus extension encasing renal pelvis and vessels with involvement and enlargement of both adrenal glands (Fig. 3A–C). Soft tissue lesions were also seen in pre- and paraaortic regions with focal encasement of the aorta. CT chest revealed soft tissue lesion encasing the aorta (coated aorta sign) and origin of left subclavian artery (Fig. 3D, E). The bone window revealed multiple fractures in the right 8th to 12th ribs and left 10th and 11th ribs (Fig. 3F).

Positron emission tomography (PET) (Fig. 4A–H) revealed multiple 18-fluorodeoxyglucose (FDG)–PET uptake sites throughout the body, midbrain and pons region, pituitary fossa, in superior mediastinum along with the origin and proximal portion of left subclavian artery, along the aortic arch and descending thoracic aorta (coated aorta sign), along the abdominal aorta, predominantly at the level of origin of renal arteries, at the aortic bifurcation and along the left common iliac artery. Soft tissue thickening was seen in bilateral peripheric regions and in bilateral renal sinuses with minimal FDG activity (Hairy kidney sign). Both bulky adrenal glands also showed mild diffuse FDG activity and mildly enlarged left paraaortic nodes also had minimal FDG activity.

The radiographs of the long bones revealed diffuse areas of sclerosis with loss of cortico-medullary differentiation

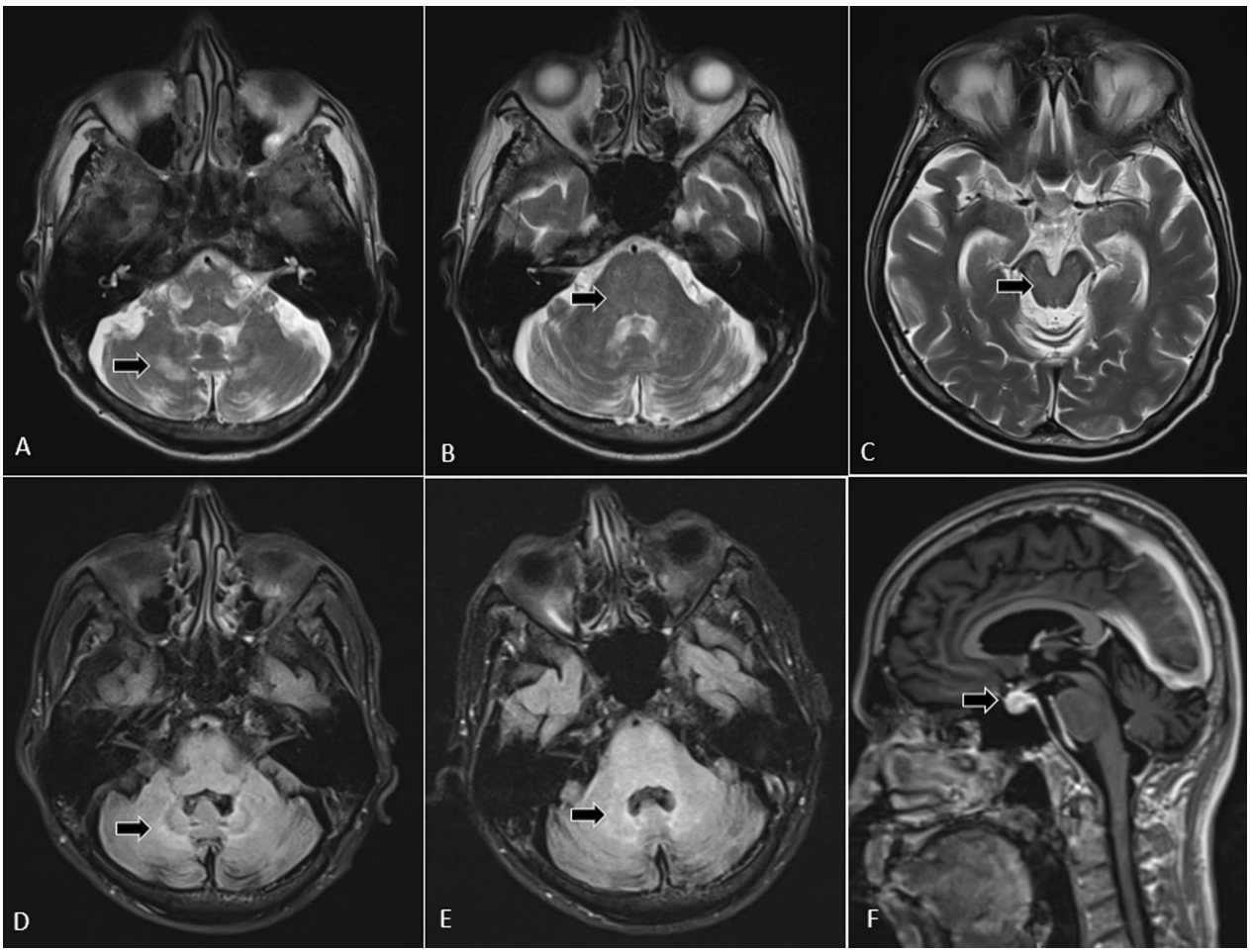

Fig. 1 Magnetic resonance imaging of brain in a 60-year-old male patient diagnosed with Erdheim–Chester disease reveals hyperintense signal in pons, midbrain, cerebellum and bilateral middle cerebellar peduncles on T2W axial images (**A**–**C**) and FLAIR images (**D**, **E**). The post-contrast T1 sagittal image (**F**) shows enlarged homogeneously enhancing pituitary gland

in diaphysis and metaphysis of femur and tibia with bone within a bone appearance (Fig. 5A–C). Diffuse sclerosis was also noted in the right fibula, humerus, radius and ulna. CT brain revealed no abnormal intra-cranial calcifications; however, ill-defined sclerotic foci were present in the calvarium (Fig. 5D, E).

The detailed case history and the radiological findings with a multisystem pattern of involvement and typical pattern of 18-FDG uptake were in favour of Erdheim–Chester disease (ECD) as a potential diagnosis. The MRI brain findings were likely neurodegenerative in nature with iron accumulation in the brain, which is an infrequent presentation of CNS involvement in ECD. The diagnosis was confirmed conclusively by a subsequent ultrasound-guided biopsy from the perinephric soft tissue lesions. The histopathology report revealed aggregates of foamy macrophages, lymphocytes and plasma cells with the tissue staining positive for CD-68 (Fig. 6A, B). The clinical symptomatology alongside the typical histopathological findings along with atypical 18-FDG–PET uptake at multiple sites, including the brain, was diagnostic of ECD with neurological manifestations. The extensive testing was succinct to rule out the possibility of ECD with an alternate concurrent neurodegenerative aetiology, and hence, a separate brain biopsy was advised against.

Treatment and follow-up

Mutational testing for BRAF V-600E was negative, and the patient was started on interferon-alpha 2b, subcutaneously thrice weekly (3 million IU) and injection methylprednisolone 1 g IV. He was discharged with the continuation of tablets tetrabenazine, haloperidol, vitamin complex. The 2- and 4-month follow-ups have not

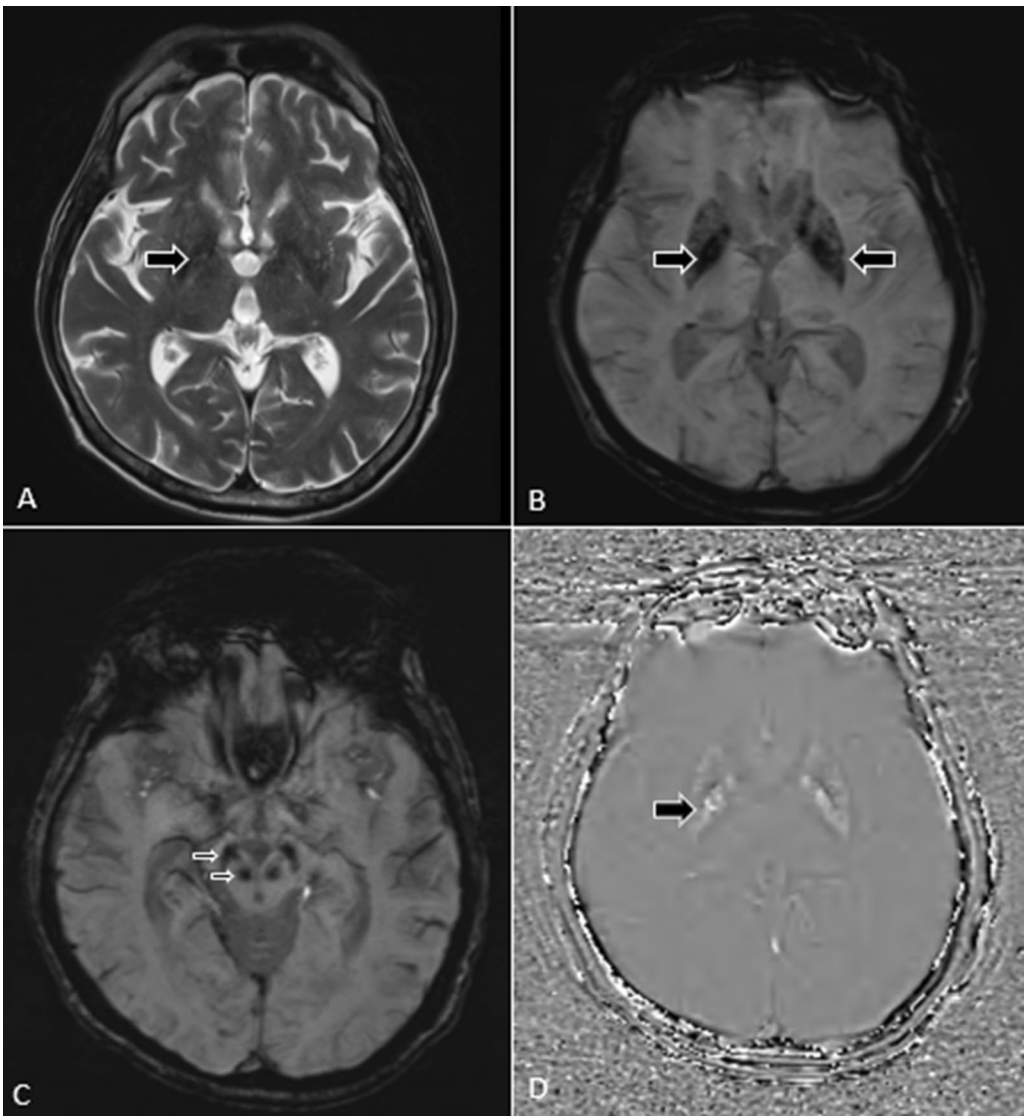

Fig. 2 Magnetic resonance imaging of brain reveals hypointense signals in basal ganglia on T2-weighted image (**A**). Susceptibility-weighted imaging reveals areas of signal dropouts in bilateral basal ganglia (**B**), substantia nigra and red nuclei (**C**) with bright signals in basal ganglia in filtered phase image (**D**). These findings are consistent with mineral deposition

revealed much improvement in symptomatology, and the patient still continues to suffer. He is planned for a follow-up evaluation of blood and radiological parameters after 6 months, and till then, the therapy will continue with close observation.

Discussion

The first reported case of Erdheim–Chester disease (ECD) was by two pathologists Jacob Erdheim and William Chester [7], in 1930, who called it lipoid granulomatosis, and the term ECD was coined only in the 1970s. In the meantime, from symptoms onset to diagnosis is reportedly a protracted 2.7 years, ranging from a couple of months to 25 years, mainly due to mislabelling and treatment as a different pathology before correct diagnosis [1]. In our case, the duration for correct diagnosis was of nearly 5 years since the onset of symptoms and 4.5 years since the first visit to the hospital.

Due to the systemic nature of the disease and multisystem involvement, a thorough multisite evaluation by CT, MRI, nuclear imaging is useful to ascertain the exact extent of the disease. Skeletal involvement is seen in up to 96% of cases and presents with bone pain, particularly involving the long bones of the appendicular skeleton

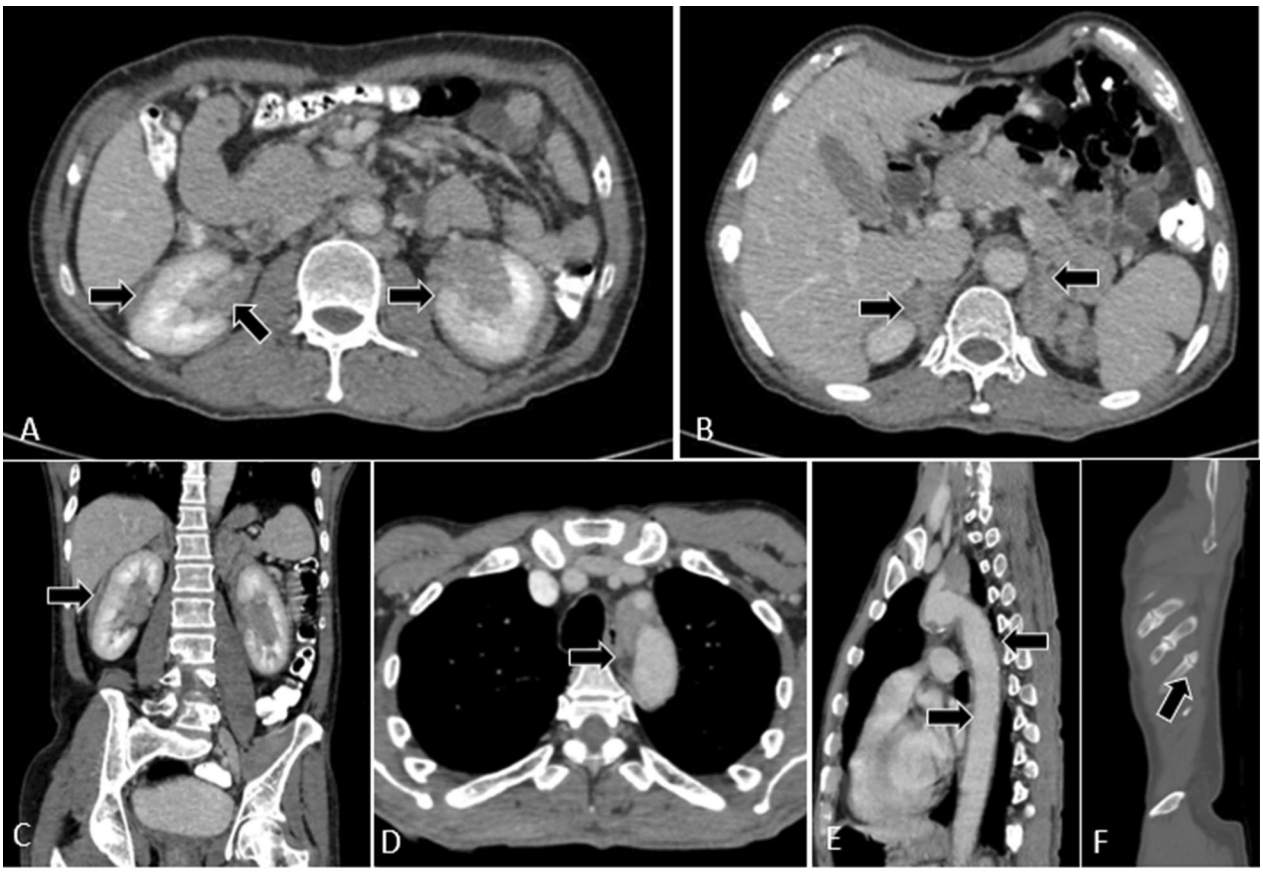

Fig. 3 Contrast-enhanced computed tomography of a 60-year-old patient diagnosed with Erdheim–Chester disease reveals ill-defined enhancing soft tissue lesions involving bilateral perinephric spaces (hairy kidney sign) with intra-renal sinus extension encasing renal pelvis and vessels (**A**, **C**) with involvement and enlargement of both adrenal glands (**B**). Computed tomography study of the chest revealed soft tissue lesion encasing aorta (coated aorta sign) and origin of left subclavian artery (**D**, **E**). Old fracture of the ribs also noted (**F**)

[4]. On plain radiography, there is typical symmetrical bilateral cortical sclerosis affecting meta-diaphysis and involvement of medulla, leading to complete loss of cortico-medullary differentiation with continued progression. Sclerotic changes in axial skeleton and epiphysis and also lytic lesion are seen in a minority of cases [3, 4]. In the present case, the involvement of the long bones was giving a "bone within bone" appearance, which has not been previously described with ECD. The skeletal lesions show FDG avidity on PET-CT, which has much better resolution than Technetium 99 m scintigraphy and also provides information about extra-skeletal involvement. Differentials for a skeletal appearance on radiographs include other osteosclerotic conditions, while symmetrical radiotracer uptake on PET is a key point in favour of ECD.

ECD tends to affect the CNS in nearly 50% of cases and is an independent predictor of death, which has been reported in almost one-third of cases [7]. The lesions are mostly extra-axial and intra-axial involvement, as is a rare presentation. The extra-axial disease process involves the pituitary gland and infundibulum presenting with central Diabetes insipidus. It may diffusely infiltrate the dural and meningeal layers leading to the formation of enhancing masses which nay mimic meningioma. Osteosclerosis of calvarial bones may also be seen. Perivascular involvement by the infiltrates can lead to stroke [8]. The intra-axial involvement is rare and can be seen as an area of altered signal intensity, usually in the posterior fossa structures with symmetrical involvement of the cerebellum and occasionally middle cerebellar peduncles and pons [9]. This neurodegenerative presentation is due to demyelination and is associated with cognitive deterioration and also cerebellar or brainstem syndromes with symptoms like ataxia, chorea and loss of balance, as seen in our case. The disease can also present as space-occupying lesions due to focal parenchymal deposits usually seen in the posterior fossa [4]. The neuroimaging differentials of ECD include Langerhans cell histiocytosis (LCH), sarcoidosis and meningiomas and

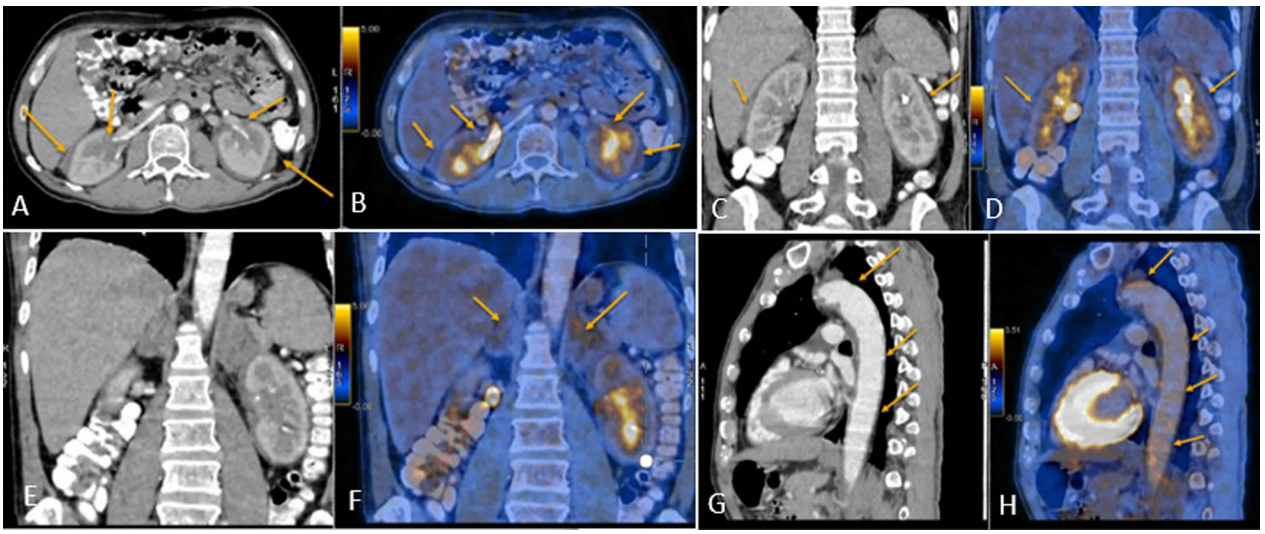

Fig. 4 Positron emission tomography scan of a 60-year-old male patient diagnosed with Erdheim–Chester disease reveals a perinephric rim of soft tissue thickening extending in bilateral renal sinuses bilaterally (**A**, **C**), and corresponding uptake images reveal minimal FDG activity (SUV max. 3.3) in this region (**B**, **D**). Both adrenal glands appear bulky (**E**) and show mild diffuse increased FDG activity (SUV max. 3.8) (**F**). Soft tissue thickening is seen along aorta (**G**) which demonstrates mildly FDG avid (SUV max. 3.4) wall uptake (**H**)

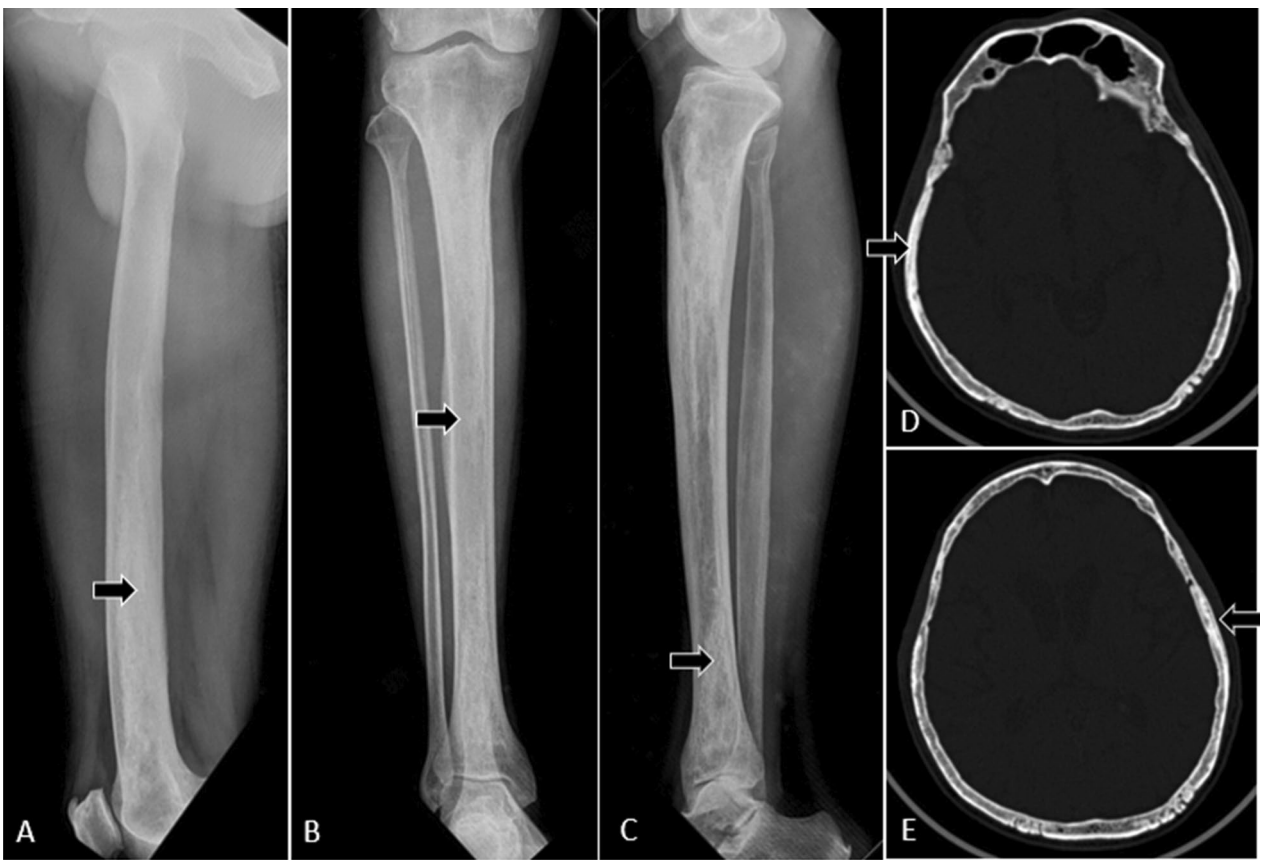

Fig. 5 Radiographs of a 60-year-old male patient diagnosed with Erdheim–Chester disease reveals regions of diffuse sclerosis with loss of cortico-medullary differentiation in diaphysis and metaphysis of right femur (**A**, **B**) and tibia (**C**) with bone within bone appearance. Computed tomography of the brain reveals ill-defined areas of sclerosis in the calvarium (**D**, **E**)

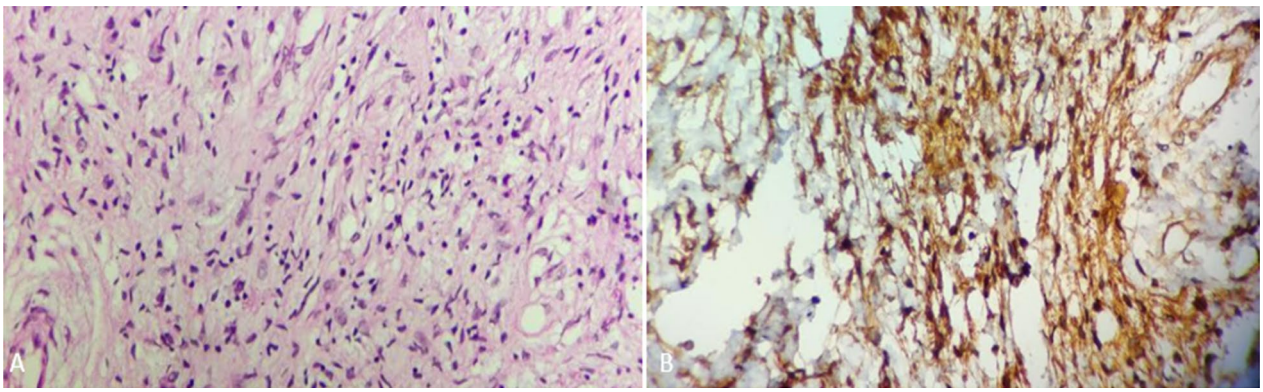

Fig. 6 Photomicrograph of the biopsy sample performed from the perinephric soft tissue infiltrates in a 60-year-old male patient with Erdheim–Chester disease reveals collection of histiocytes with abundant foamy cytoplasm and associated with minimal lympho-plasmacytic infiltrates on high power (100×) (**A**). Immunohistochemistry staining for CD-68 reveals histiocytes positive for CD-68 cells (**B**)

a key differentiator is the concurrent orbital masses and osteosclerotic lesions of bones [4].

The deposition of ferromagnetic substances in the basal ganglia, red nucleus, substantia nigra and dentate nuclei along with striatal and cerebellar atrophy has been reported in the ECD with clinical presentation of choreoathetosis and ataxia [10, 11]. Miron et al. [11] suggested the possibility of the disruption of motor pathway circuits due to basal ganglia lesions as a cause for these symptoms. However, the basal ganglia involvement is a very non-specific feature and more common in the Langerhans cell histiocytosis (LCH), which also presents with chorea, tremor and cognitive decline. The supra- and infratentorial involvement with the presence of ferromagnetic substances deposition presenting with choreoathetoid movements seen in the present case of ECD is extremely rare.

The involvement of vasculature is a poor prognostic sign as the disease process culminated in fibrosis and consequent persistent and progressive ischemic manifestations. The vasculature associated with the aorta is most frequently involved, and the disease course involves the formation of soft tissue infiltrates in a circumferential or non-circumferential pattern of segmental involvement. On CT, as was seen in our case, hypodense plaque-like tissue infiltrates circumferentially involving the aorta (coated aorta sign) are seen. This needs to be differentiated from Takayasu's arteritis, which also involves the vasculature. However, the involvement of the vessel wall is transmural in nature in Takayasu's while it is only adventitial in ECD, and vessels wall MR imaging would be useful in such cases [12, 13].

Retroperitoneum is a common site in ECD after bones and is characterized by the involvement of kidneys seen as perirenal soft tissue infiltrates extending into the posterior pararenal spaces. This extrinsic development of soft tissue causes a compressive mass effect on the kidney. The spiculated appearance of these infiltrates on close inspection begets the use of the term "hairy kidney sign". These infiltrates may involve bilateral adrenal glands without any adrenal insufficiency. This has to be differentiated from retroperitoneal fibrosis. The initiation of the disease from aortic bifurcation with the involvement of distal ureters and sparing the perirenal space are features of retroperitoneal fibrosis. The ECD is centred at the level of the renal hilum and tends to involve the proximal ureters. Typically, the disease does not involve solid visceral organs, but when it does, the prognosis is very poor [3, 4, 14].

The last decade saw considerable breakthroughs in understanding the molecular pathways of histiocytic disorders, with the discovery of BRAF-V600E mutation in 2010, followed by deep insights into the kinase mutations and the role of RAS–RAF–MEK–ERK pathways. This culminated in reassigning Langerhans cell histiocytosis (LCH) and ECD to the "L" group of revised 2016 histiocytosis classification [1, 15, 16]. As per WHO, the histiocytic disorders are to be classified either as class 1, with LCH or class 2, encompassing all non-LCH conditions like ECD, juvenile xanthogranuloma and Rosai–Dorfman disease or as class 3 which includes malignant histiocytic disorders. While ECD and LCH overlap in a certain minority of cases, IHC for S-100 protein and electron microscopic assessment of Birbeck granules are key differentiators. BRAF mutation is usually positive in over half of all cases of ECD [3]. However, Parks et al. [9] found that $BRAF^{V600E}$ mutation status was not associated with an increased risk of radiographic CNS involvement.

Our index case highlights the typical dilemma inpatient of ECD, right from protracted diagnostic time to

initiation of therapy. The biggest contributory factor is the low disease incidence and a varied presentation both clinically and radiologically. To diagnose ECD, one must have it as a differential, especially in all those patients who presented with symmetric meta-diaphyseal involvement of bones or diffuse involvement of regions of cerebellar peduncles and dentate nucleus. A detailed multimodality and multiorgan evaluation have to be performed in these cases, including thoracic and abdominal CT, MRI brain and radiographs of the long bones. The role of PET-CT cannot be overstated as the regions of involvement show FDG uptake and thus help in collaborating the diagnosis. Arnaud et al. [17] reported the specificity of PET-CT at initial and follow-up scans for diagnosis of CNS disease as 92.3% and 100%, respectively. PET-CT also allows for monitoring of the number of lesions based on SUVmax of the lesions, which correlates with the metabolic activity. PET scanning is useful in assessing CNS involvement in the disease, which is a major prognostic factor in ECD. PET scans have a moderate sensitivity but a decent specificity for the evaluation of large vessel involvement when compared to CT scans. The sensitivity of PET scans to detect involvement of the orbit, paranasal sinuses and retroperitoneum is very low [17].

There are only limited cases in the literature showcasing multiple typical ECD features on the initial PET scan. Our case had FDG avid regions in CNS—midbrain, pons, pituitary; mediastinum—along the left subclavian artery, descending aorta (coating of the aorta); abdomen—along with bilateral renal sinuses (hairy kidney sign), both adrenal glands, abdominal aorta at the level of origin of renal arteries and even paraaortic nodes; and musculoskeletal multiple ribs.

The extent of extra-osseous involvement affects the prognosis in ECD similar to in LCH. The affliction of certain organ systems like the central nervous and cardiovascular are particularly associated with a poor response to chemotherapy. While the morbidity because of ECD has significantly decreased as a result of therapeutic advances, the mortality is still significant, with a 5-year survival of only 68% [4].

Conclusions

Isolated neurological manifestations without concurrent changes in the brain on imaging may be an early presentation of Erdheim–Chester disease. The disease process in the discusses case is diffuse with multisystem involvement. Typical osseous involvement of the meta-diaphyseal region of long bones showing characteristic symmetric 18-FDG radiotracer uptake is the most common sign. CNS and CVS are the most common non-osseous sites of involvement. The pattern of involvement of CNS is much more likely to be extra-axial than intra-axial. The intra-axial disease is mostly visualized as a diffuse alteration in signal intensity in posterior fossa structures than a space-occupying lesion. The typical pattern of involvement of the kidneys and vasculature along with the characteristic "hairy kidney sign" and "coated aorta sign", while rare in a single case, is highly specific for Erdheim–Chester disease. 18-FDG uptake on PET-CT even with variable site-specific sensitivity must be performed whenever possible as it is highly specific for the disease, and the SUVmax establishes the baseline of the disease process, which can be subsequently used for monitoring of treatment response.

Abbreviations

ECD: Erdheim–Chester disease; CT: Computed tomography; MRI: Magnetic resonance imaging; SSRI: Selective serotonin reuptake inhibitor; NBIA: Neurodegeneration with brain iron accumulation; RBC: Red blood cell; PET: Positron emission tomography; FDG: Fluorodeoxyglucose; LCH: Langerhans cell histiocytosis.

Acknowledgements

Not applicable.

Authors' contributions

VR conceived the original idea and supervised the project. PA wrote the manuscript with support from VR. VR, TK and PY helped perform the experiments (reported the case). PA worked out most of the technical details with support from SM. All authors have read and approved the manuscript.

Funding

The authors received no financial support for the research, authorship and/or publication of this article.

Availability of data and materials

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Declarations

Ethical approval and consent to participate

Ethical clearance was waived by the Institutional Review Board, in view of the article being a case report. The board recommended only patient consent.

Consent for publication

Informed, full, free and voluntary verbal and written consent was provided by the patient for the entire process of publication and sharing of images and data.

Competing interests

None of the authors have any conflict of interest to declare.

Author details

[1]Department of Radiology, Dr. D.Y. Patil Medical College, Hospital and Research Center, DPU, Sant Tukaram Nagar, Pune 411018, India. [2]Department of Pathology, Dr. D.Y. Patil Medical College, Hospital and Research Center, DPU, Sant Tukaram Nagar, Pune 411018, India.

Received: 24 July 2021 Accepted: 8 October 2021
Published online: 22 October 2021

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