Osseous lesions in Erdheim Chester disease: Just a tip of an iceberg

Award: Certificate of Merit
Poster No.: C-0414
Congress: ECR 2016
Type: Educational Exhibit
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Keywords: Inflammation, Education, MR, CT, Conventional radiography, Thorax, Musculoskeletal bone, Abdomen
DOI: 10.1594/ecr2016/C-0414

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Learning objectives

1. Review Erdheim Chester disease and illustratively describe its spectrum of involvement on radiological imaging
2. Discuss the differential diagnosis for Erdheim Chester

Background

- Erdheim Chester disease (ECD) is a rare non-Langerhans cell histiocytic disease of unknown origin, characterized by infiltration of tissues by lipid-laden foamy histiocytes. ECD histiocytes are distinct from Langerhans cells in their immunohistochemical and ultra structural characteristics.
- Langerhans macrophages contain Birbeck granules and stain positive for S-100 and CD1a. Histiocytes of ECD are constantly positive for CD68 and negative for CD1a. They are negative or only very weakly positive for S-100. Ultrastructural studies show Birbeck granules in less than 20% of ECD histiocytes.
- Demographics-Usually presents in 5th to 7th decades of life with a slight male predominance. Pediatric cases are extremely rare.
- ECD is a systemic disease with protean clinical manifestations, rendering it liable to be misdiagnosed. Bones are most commonly affected, manifesting clinically as bone pain. However, extra-osseous involvement may occur in 60% cases.
- The disease shows tropism for connective tissue, adipose tissue and perivascular tissues with diffuse xanthogranulomatous infiltration of the various organs and a tendency to extend throughout the length of aorta and (up to the heart). It can affect almost all anatomical regions such as mediastinum, lungs, retroperitoneum, abdominal viscera, CNS and orbits. Potentially life-threatening complications may result, such as respiratory failure, heart failure, tamponade, and renal failure.
- Although renal and perirenal soft tissues are the most common extra-osseous site of involvement, this is usually asymptomatic.
- Most common extra-osseous disease manifestations are diabetes insipidus and painless bilateral exophthalmos. These two manifestations together with bone pain constitutes the diagnostic triad. Other CNS manifestations include cerebellar ataxia, seizures, panhypopituitarism and/or papilledema with intracranial hypertension. Lesions of the brain, meninges, facial bones, and orbits are frequently observed and should be systematically sought on the brain MR and CT images obtained in patients with ECD, even if these patients are asymptomatic. Erdheim-Chester disease must be differentiated
from Hand-Schüller-Christian disease, which is also characterized by bone lesions, exophthalmos, and central diabetes insipidus however occurs in childhood.

- Aortic involvement is uncommon however the frequency of vascular involvement is probably underestimated because it can be asymptomatic. Aortic branch stenosis or occlusion due to disease involvement may result in end organ ischemia and complications such as cerebrovascular events, coronary insufficiency, abdominal angina and renovascular hypertension.
- Cardiac involvement carries the worst prognosis. Complications responsible for high mortality rate include cardiac tamponade, severe arrhythmias, cardiomyopathy, myocardial infarction or valve insufficiency.
- Pulmonary involvement is uncommon. However once it develops, it significantly contributes to morbidity and mortality. Radiographic evidence of the disease often precedes clinical symptoms. Clinically, dyspnea and cough are the most frequent symptoms. Pulmonary function tests show moderate restrictive defects with associated decreases in diffusion capacity. Arterial blood gases are typically normal, but may show a mild-to-moderate hypoxemia.
- Although, involvement of the breast, skeletal muscle, liver, biliary ducts, mesentery, gastrointestinal tract and testes has been described in clinical reports, it is extremely rare.
- Treatment options - oral steroids and, at advanced stages, chemotherapy, radiation therapy, or immunotherapy and surgery.
- Due to diversity and non-specificity of the clinical features, radiological and pathological correlation play a strong complimentary role in establishing the diagnosis. Diagnosis is based on the combination of radiographic features, nuclear medicine features and nearly pathognomonic immunohistochemical profile. This pictorial essay highlights the imaging features of ECD involving various organ systems from head to toe.

**Findings and procedure details**

Different imaging techniques such as radiographs, computed tomography (CT), magnetic resonance imaging (MRI and nuclear medicine can be used to demonstrate various osseous and extra-osseous lesions.

- Conventional radiography is necessary to demonstrate bilateral osteosclerosis of the major long bones which is a typical finding in ECD.
• Bone scintigraphy allows a global evaluation of skeletal abnormalities, which helps to detect radiographically and clinically silent bone involvement.
• Cross sectional imaging is used to evaluate for the presence of any visceral involvement in an established case of ECD.
• CT is useful for assessment of abdominal visceral, pleural, mediastinal and aortic abnormalities and for image guided biopsy. HRCT is useful for better detection and characterization of pulmonary involvement.
• Magnetic resonance (MR) imaging is more sensitive for cardiovascular lesions.
• FDG PET/CT has the advantage of simultaneously evaluating the extent of the skeletal and extraskeletal disease, detecting occult visceral and vascular involvement and following its evolution and the response to treatment.

**Muskuloskeletal system:**

• Bones are almost always involved.
• Typical radiographic finding is bilateral symmetric osteosclerosis of metaphysis and diaphysis of long tubular bones, most commonly lower limbs with sparing of epiphysis Fig. 1 on page 8 Fig. 2 on page 10 Fig. 3 on page 12 Fig. 4 on page 13 (Fig 1-4). It usually causes medullary sclerosis however patchy areas of increased density with coarse trabeculae and cortical thickening may be seen. Sometimes, there may be associated periostitis and endosteal thickening.
• Rarely, the lesions may appear as mixed lytic-sclerotic and purely lytic lesions with sclerotic margins.
• Axial skeleton is usually spared. When involved, it is almost always associated with osteosclerosis of appendicular skeleton Fig. 5 on page 15 (Fig 5). However, isolated involvement of axial skeleton has been reported in literature.
• On technetium-99m bone scintigraphy also, ECD has pathognomonic features, namely the intense symmetric activity of the appendicular skeleton without reaching the epiphyseal regions.
• FDG PET/CT scan also reveals typical bilateral symmetric FDG uptake in the long bones Fig. 6 on page 15 Fig. 7 on page 17 (Fig 6,7).

**Differential diagnosis**

1. Langerhans cell histiocytosis (LCH)- younger age group. Sharply defined lytic lesions in the axial skeleton, namely in the spine, skull and mandible, is characteristic,
2. Paget's disease-appendicular skeleton is often asymmetric. Axial skeleton frequently involved.
4. Progressive diaphyseal dysplasia, characterised by endosteal and periosteal thickening and narrowing of the marrow cavity, and intramedullary osteosclerosis, which shows unilateral or bilateral asymmetric endosteal thickening confined to the diaphysis of the long bones. However usually detected in young people.

CNS and orbits:

- Intracranial lesions in ECD include intraaxial, meningeal, perivascular, and infundibular stalk lesions.
- The hypothalamic-pituitary axis involvement is frequent and is seen as loss of posterior pituitary bright spot on T1-weighted MR images or infundibular stalk mass with homogeneous intense enhancement after gadolinium-based contrast material administration Fig. 8 on page 17 Fig. 9 on page 18 (Fig 8,9).
- Meningeal involvement may be seen as single or multiple dural masses or diffuse pachymeningeal thickening.
- Intra-axial involvement may be seen as non-specific enhancing intra-axial masses (brain stem, cerebellum, and middle cerebellar peduncles, cerebral hemispheres and the basal ganglia) or non-enhancing bilateral symmetric high signal intensity in the dentate nucleus areas on T2-weighted MR images with corresponding low signal intensity on T1-weighted images.
- The intra-axial and dural masses show an iso- or hypointense signal on T2-weighted images, an isointense signal on T1-weighted images, and homogeneous intense enhancement on gadolinium enhanced T1-weighted images, the latter simulating meningiomas.
- Vascular involvement is seen as intracranial periarterial enhancing infiltration or venous sinus lesions Fig. 10 on page 19 Fig. 11 on page 20 Fig. 12 on page 21 (Fig 10,11,12).
- Orbital involvement is seen as intraconal or extraconal masses, causing proptosis. These may be seen either as enhancing well-defined retro-ocular masses sheathing the optic nerves or diffuse infiltration of the retro-ocular fat Fig. 12 on page 21 Fig. 13 on page 22 Fig. 14 on page 24 (Fig 12,13,14). For differential diagnosis and therapeutic purposes, it is important to localize the specific orbital compartment affected by the disease using MRI.
- Intracranial lesions in ECD are rarely isolated and are almost always found to have either osteosclerosis of the sinus walls or orbital masses. Hence, osteosclerosis of facial or skull bones associated with meningeal and orbital masses or with orbital and infundibular stalk masses have been considered highly suggestive of a diagnosis of ECD Fig. 15 on page 25 (Fig 15).
Differential diagnosis

1. Central diabetes insipidus - acquired causes due to infiltration of the hypothalamic-pituitary axis by granulomatous disease, such as tuberculosis, sarcoidosis, and Langerhans cell disease, or by metastases via hematogenous (breast carcinoma, lung carcinoma, and lymphoma) or cerebrospinal fluid (ependymoma and germinoma) spread.

2. Orbital lesions

Grave disease- Enlarged extraocular muscle bellies with sparing of tendons.

Sarcoid and leukemia - Bilateral exophthalmos with infiltration of multiple compartments

Orbital pseudotumor- Infiltration of the intraconal and extraconal space, particularly of the retrobulbar fat or of muscles with involvement of both bellies and tendons.

Lung parenchyma:

- Pulmonary involvement most commonly manifests as an interstitial process characterized by smooth interlobular septal thickening and centrilobular nodular opacities, fissural thickening, and pleural effusions Fig. 16 on page 25 Fig. 17 on page 26 Fig. 18 on page 27 Fig. 19 on page 28 Fig. 20 on page 29 (Fig 16-20).
- Although each of these findings is nonspecific, the constellation (though not pathognomonic for Erdheim-Chester disease with pulmonary involvement) is strongly suggestive, especially when observed with the typical skeletal findings.

Differential diagnosis

1. Early pulmonary Langerhans cell histiocytosis - Typical evolution and distribution of LCH, from reticulo-nodular opacities to bizarrely shaped cysts, which tend to be most numerous in the upper and middle lung zones and spare the lung bases.

2. Cardiogenic interstitial edema - Smooth interlobular septal thickening with occasional ground-glass opacification is typical of, but centrilobular nodular opacities are unusual.

3. Sarcoidosis and lymphangitic carcinomatosis-usually nodular not smooth septal thickening. Lymphangitic carcinomatosis has a multifocal rather than diffuse tendency.

4. Pulmonary venoocclusive disease-similar smooth septal thickening, associated however with signs of pulmonary hypertension, such as enlargement of the central pulmonary arteries. Centrilobular nodular opacities are infrequent.
5. Diffuse pulmonary lymphangiomatosis - diffuse smooth thickening of the interlobular septa accompany occasional patchy ground-glass attenuation; however, the thickening also affects bronchovascular bundles and diffuse infiltration of the mediastinal fat occurs. The distribution also tends to be central and perihilar.

6. Alveolar proteinosis - thickened interlobular septa with ground glass opacities (crazy paving); Centrilobular nodular opacities are not expected.

7. Amyloidosis - interlobular septal thickening or irregular lines; however, associated nodules, honeycombing, or both appear, not centrilobular nodular opacities

Heart and pericardium

- Cardiac involvement can be endocardial, myocardial, or pericardial.
- Pericardial involvement is the most frequent, seen as pericardial effusion or thickening Fig. 21 on page 30 Fig. 22 on page 31 (Fig 21,22).
- Endocardial and myocardial involvement may result in a restrictive pattern Fig. 23 on page 32 Fig. 24 on page 33 Fig. 25 on page 34 Fig. 26 on page 35 (Fig 23-26).

Differential diagnosis

1. Sarcoidosis
2. Amyloidosis

Aorta and aortic branches

- "Coated aorta"-Diffuse aortic involvement is seen as a regular circumferential periaortic infiltration, extending from the ascending aorta to the iliac junction Fig. 27 on page 36 Fig. 28 on page 38 Fig. 28 on page 38 Fig. 29 on page 49 Fig. 30 on page 40 (Fig 27-30).
- Involvement may be asymmetric or isolated to a smaller segment.
- Soft tissue is isodense to the muscle on CT and isointense on MRI, with homogeneous and weak enhancement after contrast injection.
- Preferentially periadventitial infiltration rather than parietal.
- May extend to the aortic branches, involving ostial segments of the supra-aortic, intercostal and coronary arteries in the thorax and celiac trunk, renal and mesenteric arteries in the abdomen Fig. 28 on page 38 Fig. 29 on page 49 Fig. 30 on page 40 Fig. 31 on page 40 (Fig 28-31).
- Atypically, isolated nonostial vascular involvement may occur.

Differential diagnosis
1. Retroperitoneal fibrosis - periadventitial infiltrate
2. Takayasu's disease - parietal thickening
3. Horton's disease - parietal thickening
4. Infectious aortitis - parietal thickening

Pulmonary arteries and superior vena cava

- Abnormal periarterial soft tissue infiltration appears to respect venous structures, at least in the early stages of the disease.
- Abnormal soft tissue infiltration of the mediastinum occurs and in advanced cases, may result in narrowing of pulmonary arteries and superior vena cava Fig. 32 on page 41 (Fig 32).

Renal and retroperitoneal involvement:

- Classically a hypoattenuated poorly enhancing homogeneous tissue infiltration in the renal fossae which may extend to the fat of the pararenal spaces or adrenal fossae. On MRI, the perinephric fat infiltration is isointense to muscle on T1 and T2 weighted images with mild homogeneous enhancement on post gadolinium images.
- "Hairy kidney" appearance, a symmetric and bilateral irregular soft-tissue infiltration of both the perirenal and posterior pararenal space is highly suggestive of this condition Fig. 33 on page 42 Fig. 34 on page 43 Fig. 35 on page 44 (Fig 33, 34,35).
- May extend into the renal sinuses and around proximal ureters, resulting in upper urinary tract obstruction.
- Infiltration of the adrenal fossae is seen on the imaging as bilateral, symmetric and diffuse thickening of the adrenal glands associated with infiltration of the adjacent fat however rarely causes adrenal insufficiency Fig. 36 on page 45 (Fig 36).

Differential diagnosis

1. Retroperitoneal fibrosis
2. Lymphoma

Other rare locations - skeletal muscle Fig. 37 on page 46 (Fig 37), Scrotum Fig. 38 on page 47 Fig. 39 on page 48 (Fig 38,39)

Images for this section:
**Fig. 1:** Figure 1,2: AP and lateral radiographs of right and left leg respectively show typical bilaterally symmetric involvement with heterogeneous osteosclerosis of diaphyses and metaphyses with sparing of epiphyses.

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diaphysis. The marrow cavity is partially obliterated. Periostitis is visible as a wavy contour
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**Fig. 5:** Figure 5: PET-CT in an established case of Erdheim Chester shows vertebral sclerosis with increased FDG uptake. Axial involvement is very rare.

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Fig. 6: Whole body PET image shows bilateral and symmetric elevated metabolic activity in the long bones of the lower limbs, more exuberant in the femora and in bilateral proximal humeri. An extra-skeletal site of increased uptake is seen in right upper paravertebral region. The latter represented an FDG avid enlarged lymph node.

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Fig. 7: Axial FDG-PET/CT fusion image showing elevated metabolic activity in bilateral proximal humeri.

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Fig. 13: Figure 13,14: Coronal T2-weighted image and gadolinium enhanced fat suppressed T1 weighted images respectively of the orbits in the same patient as figure 12 show similar the intraconal soft tissue to be encasing the optic nerves on both sides and showing avid homogeneous enhancement.

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**Fig. 27**: Figure 27,28,29: Coronal CT images show periaortic infiltration throughout the length of aorta causing a "coated aorta" appearance. The abnormal soft tissue encases supra-aortic branches and bilateral renal arteries. Irregular narrowing of abdominal aorta and renal arteries is noted.

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Fig. 30: Figure 30, 31: Axial contrast enhanced CT images show sheathing of aortic arch by abnormal periadventitial soft tissue with encasement of supra-aortic branch origins.

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Conclusion

ECD is commonly detected as a polyostotic bony abnormality on radiographs. However, it may show a multisystemic, extra-osseous presentation and present as a potential life-threatening disease, progressing to respiratory, renal or cardiac failure. Due to diversity and non-specificity of clinical features, it is likely to be misdiagnosed. The diagnosis relies exclusively upon radiological and pathological findings. Unless the radiologist is aware of the imaging features of this rare condition and entertains the diagnosis, useful time may be lost. Early diagnosis enables appropriate timely therapy to be performed and may enhance the patient's length and quality of life.

Personal information

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Legends:

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