

---

# **Pseudotumoral Bilateral Involvement of the Breast in Erdheim-Chester Disease: CT Appearance**

[BREAST IMAGING: Case Report]

Ferrozzi, Francesco; Bova, Davide; Tognini, Giuseppe; Zuccoli, Giulio

From the Istituto di Scienze Radiologiche, Università degli Studi, Parma, Italy. The current address of Dr. D. Bova is Department of Radiology, Loyola University Medical Center, Maywood, IL 60153, U.S.A. Address correspondence and reprint requests to Dr. F. Ferrozzi at Istituto di Scienze Radiologiche, Università, Viale Gramsci 14, 43100 Parma, Italy.

## **Abstract**

**Abstract:** We report a case of pseudotumoral involvement of the breast in Erdheim-Chester disease. CT shows an enlargement of both breasts with inhomogeneous structure, microcalcifications, and foci of fatty density.

---

Erdheim-Chester disease is a rare pathologic entity of the adult, originally described by Chester in 1930 (1), characterized by a diffuse infiltration of different organs by foamy, lipid-laden histiocytes and giant cells (2,3). The typical pattern shows symmetrical lesions involving mostly the long tubular bones of the appendicular skeleton with typical osteosclerotic appearance at conventional radiography (4-6). To date, 37 cases of Erdheim-Chester disease have been described (4). Extraskeletal involvement is uncommon and usually documented in advanced systemic disease. The most frequently affected sites include the skin, conjunctiva, pituitary gland, brain, meninges, choroid plexus, orbits, gingiva, false vocal cord (7), thyroid gland, heart, lungs, mediastinum, pleura, liver, pancreas, spleen, small bowel, kidneys, adrenal glands, retroperitoneum, pelvis, and testes (5). To our knowledge, the first case reported of breast involvement was described by Tan et al. in 1995 (8). This was characterized by soft tissue infiltration of the left breast (8).

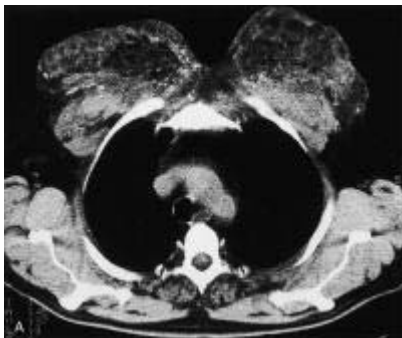
We report herein the CT findings of bilateral massive infiltration of the breast showing pseudotumoral appearance in a patient with plurivisceral manifestations of the disease.

## **CASE REPORT**

A 60-year-old man was referred for evaluation of giant bilateral gynecomastia. The gynecomastia had been present since early adulthood, but since the age of 50 had gradually increased in size with onset of cutaneous ulcerations bilaterally on the breasts. Past medical history revealed chronic pancreatitis with moderate malabsorption due to enzyme deficiency and lower extremity claudication (Leriche syndrome). The patient was affected by a manic-depressive psychosis and treated with antidepressant drugs. At 56 years old, he also reported the appearance of scrotal cutaneous induration and ulceration.

The pain caused by the skin lesions had been treated by nonsteroidal inflammatory medications. A few months prior to presentation, the patient exhibited induration of the right thigh involving the dermis and the subcutaneous tissues, originally attributed to sequelae of a posttraumatic hematoma but slowly worsening and expanding over a period of several weeks.

At the time of admission, the following imaging and diagnostic procedures were obtained: Bone scan revealed increased uptake of the radionuclide in both tibias without any other sites of increased activity; radiographs revealed mixed osteolytic and sclerotic lesions with epiphyseal sparing of both tibias; bilateral mammography gave nonsignificant results because of the very high density of the breast parenchyma; on CT, both breasts were enlarged, inhomogeneous in structure, with many foci of microcalcifications and fatty densities, and the soft tissues of the anterior thoracic wall were diffusely thickened and involved by the disease (Fig. 1). No other lesions were documented in the chest, abdomen, or pelvis.



**FIG. 1.** Enhanced CT images at the level of the aortic arch (**A**) and hepatic dome (**B**) demonstrate an enlargement of both breasts, inhomogeneous in structure, with microcalcifications and foci of fatty density. The soft tissues of the anterior thoracic wall are diffusely thickened and involved by the disease.

[\[Help with image viewing\]](#)

[\[Email Jumpstart To Image\]](#)

---

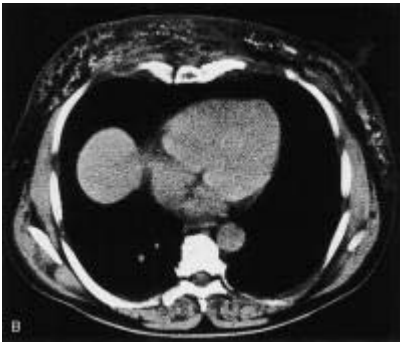


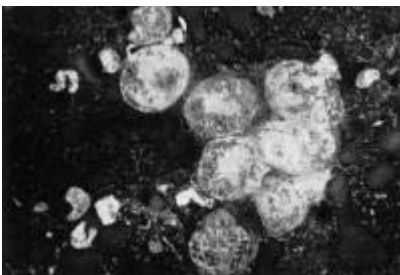
Figure 1. Continued

[\[Help with image viewing\]](#)  
[\[Email Jumpstart To Image\]](#)

---

Fine needle biopsy of the breast (Fig. 2) and of the right thigh showed numerous foamy lipid-filled cells with voluminous, often irregular nuclei, sometimes macronucleated, many giant multinucleated cells of histiocytic aspect, and scarce plasma cells. The immunohistochemical evaluation was negative for S-100 protein. The data collected suggested the diagnosis of Erdheim-Chester disease. Antiblastic therapy was proposed to the patient. However, he refused any intervention and follow-up.

---



**FIG. 2.** Pathologic specimen shows numerous foamy lipid-filled cells with voluminous, often irregular nuclei, sometimes macronucleated, and many giant multinucleated cells of histiocytic aspect.

[\[Help with image viewing\]](#)  
[\[Email Jumpstart To Image\]](#)

---

## DISCUSSION

Erdheim-Chester disease is a histiocytic disorder of unknown origin that may produce considerable abnormalities in many of the body's tissues and organs (9).

Pathologic studies have demonstrated infiltration of soft tissues and parenchyma characterized by fibrous tissue and hyalinization, interspersed with macrophages and other mononuclear cells, including histiocytes, lymphocytes, and plasma cells (3). The preponderant lipid-laden histiocytes or foam cells are different from the Langerhans cells that define "Langerhans cell histiocytosis" because the former have no intracytoplasmic granules and do not immunostain for S-100 protein (3,10,11).

The most typical site of involvement is the bone, where it is characterized by irregular thickening of trabeculae in both the cortical and the cancellous bone.

There is often synchronous bone marrow fibrosis, with discrete yellow foci representing lipid granulomata (9). The radiographic appearance shows diffuse bony sclerosis that predominantly involves the metaphysis and the diaphysis of the long tubular bones of the appendicular skeleton, with some degree of symmetry. The epiphyseal segment may be affected as well, although the degree of involvement is usually less than that in the neighboring metaphysis (9,12). Endosteal and periosteal bone production may be marked, leading to obliteration of the corticomedullary interface (9). Thirty-seven percent of the patients also demonstrate a lytic component in addition to the classic sclerotic changes (8). Flat bones of the axial skeleton are less frequently involved (5).

Organ involvement generally begins in the perivascular region, with extension to the surrounding soft tissues and with relative sparing of the parenchyma (13). Retroperitoneal and orbital tissues, pericardium, myocardium, lungs, kidneys, liver, and pancreas represent the most frequent sites of involvement (6,8).

Clinical manifestations are highly variable and often nonspecific. In some patients, symptoms and signs are completely absent, delineating a rather benign process, with abnormalities limited to the skeleton. In other patients, prominent clinical abnormalities accompany the histiocytic accumulation in osseous and, more importantly, extraosseous sites, in the aggressive systemic character of the disease (9).

Histologic examination is crucial for diagnosis, particularly for differentiating it from other diffuse skeletal diseases that may show similar pattern of bone involvement, such as mastocytosis, myeloid metaplasia, metastatic disease, lymphoma, fluoride intoxication, and toxic osteoarthropathy (6,12). To our knowledge, Tan et al. (8) reported the first case of breast involvement in Erdheim-Chester disease; it was characterized by monolateral soft tissue infiltration of the breast. Our case is a pseudotumoral bilateral involvement of the breast.

Such a voluminous gynecomastia might suggest neoplastic lesions like histiocytoid carcinoma or sarcoma, but the synchronous presence of the classic skeletal lesions and the involvement of the soft tissues of the thigh allowed a correct diagnosis.

### **Acknowledgment:**

The authors thank Dr. Marc Boillat de Corgemont Sartorio for reviewing the manuscript.

### **REFERENCES**

1. Chester W. Über Lipoidgranulomatose. *Virchows Arch [A]* 1930; 279:561–602. [\[Context Link\]](#)
2. Jaffe HL. Lipid (cholesterol) granulomatosis. In: *Metabolic, degenerative and inflammatory diseases of the bone and joints*. Philadelphia: Lea & Febiger, 1972: 535–9. [\[Context Link\]](#)
3. Athanasou NA, Barbatis C. Erdheim-Chester disease with epiphyseal and systemic disease. *J Clin Pathol* 1993; 46:481–2. [Bibliographic Links](#) [\[Context Link\]](#)

4. Kenn W, Stäbler A, Zachoval R, et al. Erdheim-Chester disease: a case report and literature overview. *Eur Radiol* 1999; 9:153–8. [\[Context Link\]](#)
5. Miller RL, Sheeler LR, Bauer TW, et al. Erdheim-Chester disease. Case report and review of the literature. *Am J Med* 1986; 80:1230–6. [\[Context Link\]](#)
6. Veyssier-Belot C, Cacoub P, Caparros-Lefebvre D, et al. Erdheim-Chester disease. *Medicine* 1996; 75:157–69. [\[Context Link\]](#)
7. Sandrock D, Merino MJ, Scheffknecht BHB, et al. Scintigraphic findings and follow-up in Erdheim-Chester disease. *Eur J Nucl Med* 1990; 16:55–60. [Bibliographic Links](#) [\[Context Link\]](#)
8. Tan APA, Tan LKA, Choo IHF. Erdheim-Chester disease involving breast and muscle: imaging findings. *AJR* 1995; 164:1115–7. [Bibliographic Links](#) [\[Context Link\]](#)
9. Resnick D, Greenway G, Genant H, et al. Erdheim-Chester disease. *Radiology* 1982; 142:289–95. [\[Context Link\]](#)
10. Favara BE, Jaffe R. Pathology of Langerhans cell histiocytosis. *Hematol Oncol Clin North Am* 1987; 1:75–95. [\[Context Link\]](#)
11. Lieberman PH, Jones CR, Steinman RM, et al. Langerhans cells (eosinophilic) granulomatosis: a clinicopathologic study encompassing 50 years. *Am J Surg Pathol* 1996; 20:519–52. [Ovid Full Text Bibliographic Links](#) [\[Context Link\]](#)
12. Brower AC, Worsham GF, Dudley AH. Erdheim-Chester disease: a distinct lipoidosis or part of the spectrum of histiocytosis? *Radiology* 1984; 151:35–8. [Bibliographic Links](#) [\[Context Link\]](#)
13. Fink MG, Levinson DJ, Brown NL, et al. Erdheim-Chester disease. Case report with autopsy findings. *Arch Pathol Lab Med* 1991; 115:619–23. [Bibliographic Links](#) [\[Context Link\]](#)

Index Terms: Breasts, neoplasms; Bones; Histiocytosis; Computed tomography

---

*Accession Number: 00004728-200003000-00018*

---

Copyright (c) 2000-2007 [Ovid Technologies, Inc.](#)  
Version: rel10.5.1, SourceID 1.13281.2.21