Case 2487

Angiofollicular lymphoid hyperplasia (Castleman's disease)
Published on 03.03.2005

DOI: 10.1594/EURORAD/CASE.2487
ISSN: 1563-4086
Section: Breast imaging
Imaging Technique: Mammography
Imaging Technique: Ultrasound
Case Type: Clinical Cases
Authors: Angelatou O, Moutaftsis E, Panou A, Pantziara M, Karatzas T
Patient: 44 years, female

Clinical History:
A 44-year-old woman came to our mammography department for her annual check-up.

Imaging Findings:
Our case concerns a 44-year-old, asymptomatic woman who came to our mammography department for her annual check-up. The mammography results showed multiple enlarged and homogenously radiopaque lymph nodes in the left axilla. A supplementary ultrasound investigation showed multiple, ovoid hypoechoic lesions with a maximum diameter of 1.8 cm. The differential diagnosis included many malignant and benign entities. The definitive diagnosis was obtained after a surgical excision and pathologic examination of the enlarged lymph nodes.

Discussion:
Angiofollicular lymphoid hyperplasia is a rare and usually benign disease of an unknown origin with the characteristic form of lymph node hyperplasia. It is also known as giant lymphnode hyperplasia, or angiomatous lymphoid amartoma. It was first described by Castleman and his collaborators as a disease localized in the mediastinum. However, a less frequent generalized or multicentric form of Castleman's disease has also been reported to occur. This disease can affect any group of lymph nodes, with the mediastinum being the most frequent site of occurrence. The axillary lymph nodes are an unusual location to get afflicted with the disease since only 2% of the known cases involve the axilla. CLASSIFICATION: As mentioned above, angiofollicular lymphoid hyperplasia has two forms: 1) localized and 2) generalized or multicentric. The first form is categorized (according to Keller and collaborators) as (a) hyaline vascular type (76%–91%) and (b) plasma cell type(10%–24%). The probable causes for the former type are chronic antigenic stimulation and developmental abnormality of the lymphoid tissue. Mediastinal and cervical nodes may usually be involved. The pathogenic agent in the latter case could be chronic viral antigenic stimulation. The generalized or multicentric type could further be divided into lymphoid hyperplasia with or without neuropathy. CLINICAL PRESENTATION AND LABORATORY FINDINGS: The majority (97%) of patients with the localized disease of the hyaline type are asymptomatic and in the event that symptoms do occur, these are due to compression of the surrounding structures by the developing lesion and depend on the location of the disease. On the contrary, patients who are suffering from the plasma cell type of the disease are more likely to present with symptoms such as fever, fatigue, weight loss and retardation of development in children. Laboratory findings include hypochromic microcytic anemia, increased erythrocyte sedimentation rate and polyclonal hypergamaglobulinemia. The increased amounts of IL-6, which are produced by hyperplastic lymph nodes, are considered to be the cause of both clinical and laboratory findings. Finally, the generalized form of the disease may manifest with severe systemic symptomatology which includes multicentric peripheral lymphadenopathy, hepatosplenomegaly, enlargement of the
salivary glands, skin lesions (hypertrichosis, hirsutism, sclerodermatous thickening, hyperpigmentation and hemangiomas), symmetrical peripheral neuropathy and papilledema or pseudotumor cerebri. Herget et al have postulated its association with the POEMS syndrome (polyneuropathy, organomegaly, endocrine abnormalities, M-protein, skin changes). HISTOLOGY: The hyaline vascular type of Castleman’s disease consists of altered germinal centers, which include small hyperplastic arteries and sometimes capillaries. Follicles present a lack of germinal centers, normal cell components and are atrophied. Lymphocytes are arrayed in an “onion skin” configuration. Furthermore, the interfollicular region is expanded with numerous post-capillary venules. Plasma cells are relatively absent. The plasma cell type histologically resembles a florid, follicular lymphoid hyperplasia with layers of plasma cells in the interfollicular region. The prominent vascular changes seen in the hyaline vascular type are absent in this histological type. Generalized or multicentric Castleman’s disease usually not only exhibits some of the vascular abnormalities, characteristic of the hyaline vascular type of this disease but also contains a marked plasmocytosis as in the plasma cell type. IMAGING MAMMOGRAPHY: Enlarged, homogeneously radiopaque axillary lymph nodes. ULTRASONOGRAPHY: Ovoid hypoechoic lesions with no hyperechoic center. COLOR DOPPLER SONOGRAPPHY: Hypoechoic tubular structures with a measurable blood flow and many feeding vessels. CT: A well-defined mass with or without central calcification exhibiting a marked contrast enhancement in the hyaline vascular type and a slight enhancement in the plasma cell type. MRI: Homogeneous mass with a low signal intensity on T1-weighted and high signal intensity on T2-weighted images. DIFFERENTIAL DIAGNOSIS: The differential diagnosis of enlarged radiopaque lymph nodes includes metastatic breast cancer, metastases from non-breast primary cancer, lymphoma, leukemia and inflammatory diseases such as collagen vascular disease, sarcoidosis, HIV-related lymphadenopathy or reactive lymphadenopathy associated with breast abscess. TREATMENT - PROGNOSIS: Surgical excision is the treatment of choice especially in the localized type. Nevertheless, because of the hypervascularity of the lesion, especially in the hyaline vascularity type, preoperative embolism for debulking may be useful. The generalized or multicentric form of the disease is treated with corticosteroids, chemotherapy and radiation therapy, although surgical intervention may be indicated for peripheral neuropathy. The prognosis of the localized form is generally good, although a small proportion may relapse after the surgical excision in spite of the complete removal of the lesion. Some lesions, however, especially those of the plasma cell type, may exhibit a malignant degenerative potential. The generalized form of Castleman’s disease has a 50% mortality rate with an average survival time of approximately 27 months. Death is usually caused by complications such as infections or septicemia, as well as malignancies such as Kaposi’s sarcoma, malignant lymphoma or epithelial neoplasm. Differential Diagnosis List: Angiofollicular lymphoid hyperplasia (Castleman's disease).

Final Diagnosis: Angiofollicular lymphoid hyperplasia (Castleman's disease).

References:

Meador TL, McLarney JK. Related Articles, Links
Figure 1

Description: Fatty involution of the breast without any focal lesion. Enlarged, radiopaque axillary lymph nodes. Origin:
Figure 2

Description: Enlarged lymph nodes are sonographically visualized as ovoid, hypoechoic lesions without a hyperechoic center. Origin: