



An Unusual Axillary Mass: Castleman's Disease

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Abstract

Castleman's disease is the giant lymph node hyperplasia first identified in 1956 by Benjamin Castleman et al. It is a very uncommon lymphoproliferative disorder without a fully known etiology. It is also known as angiofollicular lymph node hyperplasia, giant lymph node hyperplasia, lymphoid hamartoma, giant benign lymphoma and follicular lymphoreticuloma. Clinically it has localized and multicentric types while histopathologically it has hyaline, vascular, plasma-cell and mixed types. Thymoma, sarcoidosis, tuberculosis, lymphoma and malign lesions are the medical conditions which are to be primarily considered in the differential diagnosis. The present study presents a case with Castleman's Disease localized in axilla.

Keywords: Castleman; Lymphoproliferative disorder; Axilla

Case Presentation

A 37-year-old female patient with no systemic disorders presented to our clinic with the complaint of swelling under the left armpit. Furthermore, the patient had the complaints of chronic weakness and tiredness. She had a history of tonsillectomy operation in childhood and 10 pack-year of smoking. The axillary mammary ultrasonography revealed a 9x5 mm hypoechoic lesion with blurred borders in the left breast at 1 or 2 'o'clock position 1 cm away from areola and an approximately 75x26 mm fusiform hypoechoic solid lesion with neat borders and a dense core, and vascularization as revealed by CDUS in the left axilla inferior. This profile was interpreted as a pathological giant lymphadenopathy (Figure 1). The PA chest radiography revealed no pathology (Figure 2). In the physical examination, palpable lesions were observed in the breast and axilla. It was determined that the lesions in the breast and axilla were to be excised for the purposes of diagnosis and treatment. Preoperatively, the lesion in the axilla was marked stereotactically using a wire. The lesion was adjacent to the axillary artery and vein. The approximately 7 cm lymph node, which was marked using the wire, in the axillary area, was nearly totally excised. Upon the absence of any postoperative complications, the patient was discharged from the hospital on the postoperative day 3. The pathological workup concluded that the mass in the axilla was Castleman's Disease-Hyaline vascular type. Then, the patient was referred to the departments of Radiation Oncology and Medical Oncology. However, chemo-radiotherapy was not scheduled for the patient. She was discharged after scheduling PET-CT for her. Considering the possibility of the disease to show a multicentric character, the patient was initially asked for to pay a control visit 1 month later for a close follow-up.

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Discussion

As in our case, Castleman's disease may develop as localized or disseminated and may be classified in different ways as well. As for localization, it may display unicentric or multicentric involvement. Multicentric type is particularly associated with Human Herpes-8 secreting viral interleukin-6, an interleukin-6 homologue, and it is seen more commonly in the patients with immune suppression. There was no finding of immune suppression in our patient. The types with multicentric localization have the risk of become malignant [1-4]. Thus, the patient was followed up closely. These masses mostly come up as asymptomatic mediastinal masses which are followed by neck, pelvis, retroperitoneum and axilla [5]. When in multicentric form, the mean survival is 2 years with an approximately 50% mortality rate, and surgery is not a highly preferred treatment approach. In this form, there is a high risk for multiple myeloma, B-cell neoplasms and Kaposi's sarcoma [6]. For the localized form, the mean age is 23 years and the risk of malignancy is very low. The 5-year survival rate is 100%. It is permanently cured with surgery and the systemic symptoms are seen to disappear [7]. The hyaline vascular type detected in our patient has a benign clinical profile and it is more commonly seen in localized form [8]. Plasma cellular type, however, localization is

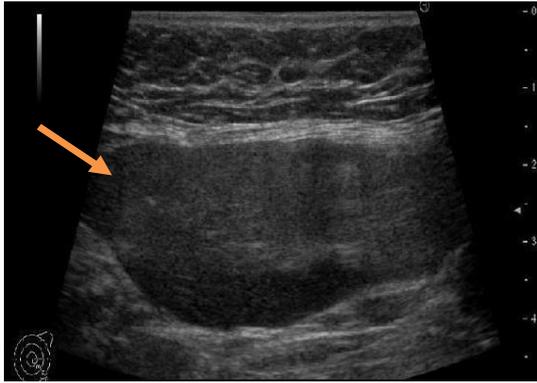


Figure 1: The arrow indicates ultrasound view of 75x26 mm fusiform hypoechoic solid mass.

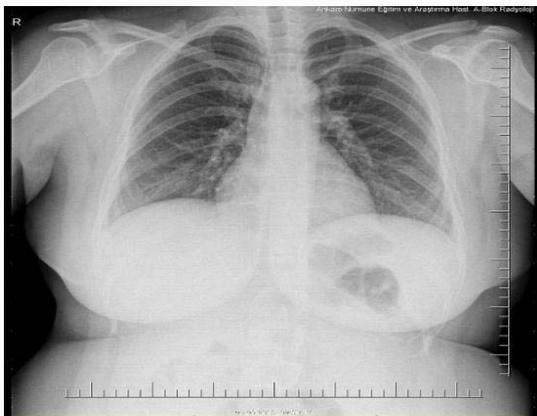


Figure 2: Normal PA chest radiograph.

usually the mediastinum (52%), though it may also be the neck, axilla, retroperitoneal space, mesentery and pelvis [9]. It has an aggressive clinical profile and it is more commonly seen in multicentric form. Eventually, the approved approach for Castleman's disease, both diagnostically and therapeutically, is surgical excision. Total excision of the mass is sufficient for a curative treatment in local forms, and anemia, asthenia, fever and weight loss are normalized. The first option for the treatment of hyaline vascular type is the complete resection. The response to surgery is quite well and the 5-year survival rate is 100%. Though being rare, there is a risk of malignancy in

the unifocal type of the hyaline vascular variant. For the cases with failed total resection or ineligible for resection, solely radiotherapy or radiotherapy accompanied by steroid treatment is recommended, and there are studies reporting favorable results with these treatments. Radiotherapy has a limited effect on the size of the mass and reversal of the hematological abnormalities [10]. Castleman's disease should be taken into consideration for the giant lymphadenopathies particularly in mediastinum, pelvis and axilla, and surgical approach should not be avoided as a nearly total recovery may be achieved postoperatively.

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