CASE REPORT

A 92-year-old male patient presented with a 3-year history of an enlarging, painless mass on the right arm (Fig. 1). He did not have any history of trauma on the area. The patient reported no weight loss and had no other symptoms other than rapid enlarging of the tumor for the last 6 months. Physical examination showed an 8 x 5 cm flesh colored, firm, ill-defined mass on right arm (Fig. 1). No cervical or axillary lymphadenopathy was noted. The MRI showed a huge ill-defined soft tissue mass on the anterior aspect of lower right arm measuring 13 x 5.7 x 4.6 cm. T1-weighted images showed heterogeneous hypo and hyper-intense signal (fatty tissue) with increased enhancement in T2-weighted image. The initial clinical differential diagnoses included liposarcoma, lipoma, teratoma, lymphoma, and germ cell tumor (Fig. 2). An incisional biopsy was performed and the microscopic pictures are shown in (Fig. 3). After the diagnostic biopsy, a marginal excision was performed under general anesthesia and a partial thickness skin graft was used to cover the defect.
DIAGNOSIS: Well-Differentiated Liposarcoma (Atypical Lipomatous Tumor)

DISCUSSION

Grouped under the label of intermediate malignancy, well-differentiated liposarcoma (WDLS) was originally termed atypical lipoma and atypical lipomatous tumor to emphasize their almost invariably locally aggressive behaviour (almost never metastasize). The concept of atypical lipoma or atypical lipomatous tumors was first introduced by Evans and associates in 1979. This proposed terminology has generated considerable controversy. In 2002, the World Health Organization (WHO) unified the concept and viewed atypical lipomatous tumor and WDLS as synonyms. The WHO also distinguishes the 4 variants of liposarcoma (as proposed by Enzinger): well-differentiated (which includes adipocytic, sclerosing, inflammatory and spindle cell), myxoid, round cell, and pleomorphic variants, and a fifth variant, dedifferentiated.

WDLS is the most common type of liposarcoma (> 50% of cases of liposarcoma) and tends to occur in middle-aged or elderly patients and arises in deep subcutaneous tissues of the limbs or retroperitoneum with few reports of metastases. Histologically, it is characterized by the predominance of univacuolar adipocytes with variation in shape and size, and the presence of hyperchromatic cells in a broad fibrous septa, with or without lipoblasts.

Tumor location is thought to be the most important prognostic factor in patients with WDLS, principally because of resectability that correlates with rate of recurrence. Patients with WDLS arising in the extremities have a very significantly better prognosis compared to those arising in the retroperitoneum. Both CT scan and MRI are excellent methods of anatomical definition and can predict to a degree whether a tumor is benign or malignant. An entirely uniform tumor with a density of fat clearly is highly likely to be a benign lipoma, whereas the presence of heterogeneity and thicker septi will be more characteristic of WDLS.

The local treatment is the mainstay with maximal preservation of important structures. Like all other soft tissue tumors, WDLS tend to infiltrate along the lines of least resistance. As a consequence, it is essential to accurately diagnose all anatomical extensions of these tumors preoperatively with either CT or MRI. The accurate core biopsy and detailed anatomical imaging preoperatively to plan excision margins for these tumors and to enable appropriate one-stage rather than multistage surgery are very important. A complete marginal excision as a one-stage procedure with maximal preservation of important structures is the aim.

Radiotherapy and chemotherapy are generally not indicated for which shows poor response and low sensitivity to the therapy. Local recurrence is common for patients with WDLS and it becomes apparent within the first 6 months in most cases, but it may be delayed for 5 or 10 years following the initial excision. Recurrence is related to the incomplete excision and tumor tissue left behind at the time of surgery.

REFERENCES