Von Recklinghausen’s Disease with a Giant Lipoma

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Abstract

A 64-year-old man with a long history of von Recklinghausen’s disease presented with a three years history of a right shoulder mass. We report an extensive lipoma of the shoulder region and demonstrate the MRI findings.
Introduction

Although nonneuroectodermal tumors in association with von Recklinghausen’s disease have been reported previously, a huge lipoma in a patient with von Recklinghausen’s disease has not been described.

Case Report

A 64-year-old man with a long history of von Recklinghausen’s disease presented with a three years history of a right shoulder mass. He had severe neurofibromatosis of the entire skin, therefore, he had been regarding the mass as a part of neurofibromatosis. His past medical history had been pulmonary tuberculosis since 20 years old. On examination, the soft mass located over the right supraclavicular area and was sized of a baseball (Fig.1).

*Figure 1*: Many neurofibromas of the skin make their appearance from childhood, and are 1 to 10 centimeters in diameter. A growing mass of the right shoulder is discernible.
It located subcutaneously and showed no obvious adhesion to the adjacent structures. No spontaneous pain and tenderness was noted. The right shoulder motion was not restricted and neurological sign was unremarkable. Only right lateral bending of the neck was slightly restricted. Radiographs disclosed diffuse soft tissue tumor but no bony involvement. MR images revealed a large mass from the right supraclavicular area to subacromial joint (Fig. 2).

![Figure 2: A coronal T1-weighted MR image shows a high signal intensity of the large mass in the right supraclavicular area.](image)

The lesion showed a high signal intensity both on T1 and T2-weighted images, consistent with lipomatous tumors. The mass expands over the supraspinatus fascia without invesion into muscle. A rotator cuff tear or intracapsular lesions were not remarkable. At surgery, there was a large encapsulated tumor (15×8×8cm), which was removed easily and extended to the thoracic wall and the subacromial area. The brachial plexus and vessels were not involved into the tumor. Histologically this tumor was composed of mature fat cells showing only
slight variation in cellular size and shape, separated by thin fibrocollagenous tissues, without malignant findings (Fig. 3). These findings are consistent with lipoma. Post-operatively, the range of neck and shoulder motion gradually recovered, and his discomfort was relieved. A follow up MRI showed no recurrence of the tumor.

Figure 3: Histologically, this tumor showed a proliferation of fat cells showing only slight variation in cellular size and shape without any malignant findings.
Discussion

Von Recklinghausen’s disease is a disease of autosomal dominant inheritance, which has multiple café-au-lait spots and neurofibromas. One of the characteristics of disease is concomitant development of various neoplasms. If a tumor mass grows during the follow-up periods, a regrowth of preexisted benign neurogenic tumors or a malignant change of their tumors could be usually considered, but not lipomatous tumors such as lipoma or liposarcoma.

Malignant transformation in neurofibromatosis is estimated at 3%~17% [1, 5]. Malignant schwannoma is the most common malignancy, but also angiosarcoma, chondrosarcoma and rhabdomyosarcoma were reported [2]. In addition, patients with neurofibromatosis may also develop optic gliomas, astrocytomas, acoustic neuromas, neurilemmomas, and meningiomas, which are identified in 5 to 10% of the patients [3]. In our search of the literature, only one case of juxta-medullary spinal lipoma in a patient with von Recklinghausen’s disease was reported [4].

In our case, lipoma arised from subacromial space and extended to supraclavicular region. Deep lipomas are rare compared with superficial lipomas and their localization in the deep shoulder region is extremely rare.
References


