



Imaging of High Grade Myxoid Fibrosarcoma- A Rare Case Report

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Abstract

Myxofibrosarcoma is a myxoid variant of malignant fibrous histiocytoma that most commonly involves the extremities of elderly people. However, a primary myxofibrosarcoma with bone invasion in young adults is extremely rare. Herein, we report the case of a 52-year-old Women presenting to our hospital, with complaints of oral ulcers, constipation since 2 weeks and pain in anal area since 1 week. Patient also complained of pain in back of right thigh and right hand/shoulder/TMJ since 1 week and unable to walk and sit due to pain. She was then referred to department of Radiodiagnosis for an MRI lower extremities, based on the findings observed a PET-CT was advised and patient underwent subsequent core biopsy of lung mass and immunohistochemistry.

INTRODUCTION

Myxofibrosarcoma is a myxoid variant of malignant fibrous histiocytoma (MFH), characterized by a nodular appearance, prominent myxoid matrix, elongated curvilinear capillaries, and location mostly in dermal and subcutaneous tissues in the extremities of elderly people^{1,2}.

Several clinical studies of myxofibrosarcomas showed that approximately 80% of the tumors occurred in the extremities and about 12% in the trunk, with a high incidence in the dermal or subcutaneous tissues³. Other sites for myxofibrosarcomas included head and neck

region, retroperitoneum, pelvis and heart⁴. However, a primary myxofibrosarcoma with bone invasion was rarely reported⁵. In addition, the peak age incidence of myxofibrosarcomas was in the fifth to seventh decades with a slight male predominance⁶.

CASE PRESENTATION

We report this rare case report of a 51 year old woman presenting to our hospital, with complaints of oral ulcers, constipation since 2 weeks and pain in anal area since 1 week. Patient also complained of pain in back of right thigh and right

hand/shoulder/TMJ since 1 week and unable to walk and sit due to pain. She was then referred to department of Radiodiagnosis for an MRI lower extremities, based on the findings observed a PET-CT was advised and patient underwent subsequent core biopsy of lung mass and immunohistochemistry.

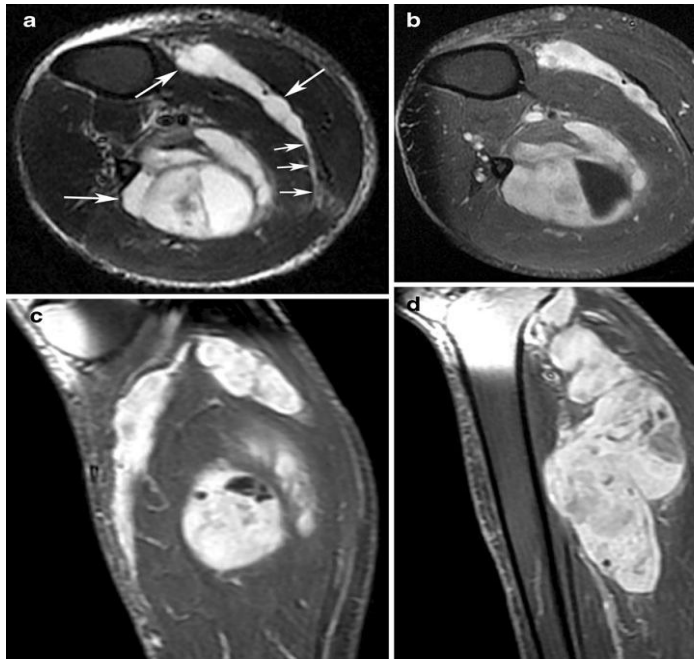


Fig. 1 51 year old woman with high-grade myxofibrosarcoma (MFS). a Axial short-tau inversion recovery image through the proximal calf demonstrates multinodular, very high signal masses (large arrows) with a deep location. Note the prominent, long, high-signal extension (small arrows) in the posterior calf. Such extension along fascial planes, between muscles in this case, is a common imaging manifestation of MFS. High signal in the subcutaneous fat represents edema. b, c Axial and sagittal T1-weighted images with fat suppression obtained after gadolinium contrast administration demonstrate marked enhancement of the masses, including the tail. Tumor contacts the fibular periosteum in b, but no gross osseous invasion was found at pathological examination. d Sagittal T1-weighted post-contrast image with fat suppression shows the multinodular morphology of the largest component of the mass.

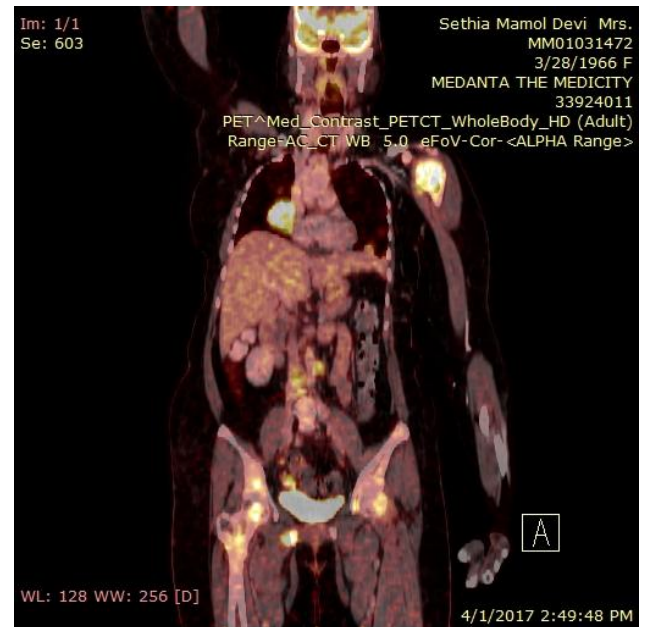


Fig 2: PET-CT shows metabolically active skeletal lesions, bilateral pulmonary and pleural based lung lesions, pelvic deposit and the multiple lymph nodes(retroperitoneal , pelvic and right prevascular) are suggestive of metastases.

DISCUSSION

Myxofibrosarcoma, also known as a myxoid variant of MFH, is one of the most common sarcomas in the extremities of elderly people, characterized by a high frequency of local recurrence. Distinctive histological features included the following: a commonly nodular growth pattern; a myxoid matrix containing elongated, curvilinear capillaries; and fusiform, round or stellate tumor cells with indistinct cell margins, slightly eosinophilic cytoplasm, and hyperchromatic atypical nuclei³.

The most common sites for this tumor are the extremities, predominantly subcutaneous. Primary myxofibrosarcomas with bone invasion were rarely occurred, with no more than 15 previous cases worldwide being reported in the literature⁵⁻¹¹. Five of these patients developed myxofibrosarcoma in a long bone of the extremities, with an average age of 53.2 years (range, 31 to 84).

The etiology of myxofibrosarcoma remains unknown. It was previously reported that 2 patients developed a myxofibrosarcoma after radiotherapy^{12,13}. In the current case, the patient

developed a low-grade myxofibrosarcoma in the left thigh after a multiple fracture and metal implantation suffered about 3 years ago. The patient was a farmer with no known history of potential environmental hazardous substances exposure, and he was an occasional smoker and did not drink alcohol. Except for receiving a metal implantation for osteosynthesis, the patient had no relevant medical or family history. In addition, the patient has no clinical or radiological evidence of a neoplasm elsewhere in the body, excluding the possibility of a secondary lesion.

CONCLUSION

Fibrous soft-tissue tumors represent a heterogeneous group of lesions that can involve both superficial and deep musculoskeletal structures.

The fibrosarcomas commonly demonstrate a large size, tendency toward aggressive behaviour and high recurrence rates after surgery. Although CT and MRI features of fibrous soft-tissue tumors are not pathognomonic, it is important for radiologists to recognize the imaging characteristics and common sites of occurrence of these lesions to help determine an appropriate differential diagnosis and guide the management of these lesions.

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