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Case report

Recurrent Retroauricular Myxofibrosarcoma in Libya: A Case Report

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Abstract
*Myxofibrosarcoma (MFS) is a rare malignant soft tissue sarcoma, typically affecting elderly patients and commonly arising from the extremities. Its occurrence in older individuals with atypical locations, such as retroauricular, is uncommon. A 50-year-old female presented with a history of recurrent left huge retroauricular mass, a painless and hard mass, not mobile, not tender, attached to the skin, approximately 4*3 cm. Initial imaging suggested a benign lesion; however, histopathological results of the excised mass revealed a spindle cell neoplasm with a lot of myxoid changes that indicate High-grade Myxofibrosarcoma. Immunohistochemistry confirmed a diagnosis of high-grade MFS. The patient underwent surgical excision of the retroauricular mass with clear margins and remains under regular follow-up with no signs of recurrence to date. In this case, the diagnostic tools of MFS are mainly used in elderly patients with atypical anatomical locations. The imaging is a nonspecific tool to confirm diagnosis, and the histopathological examination is an accurate diagnostic tool for MFS. The mainstay of MFS treatment is surgical excision, and in this case, the patient received Adjuvant Radiotherapy and systemic chemotherapy therapy (doxorubicin and ifosfamide) due to high-grade or multiple recurrences. Adjuvant therapy and Regular follow- up; are essential to prevent local recurrence. This case contributes to the limited data on MFS in sub-Saharan Africa, to increase awareness and reporting, and to better understand epidemiology and management strategies in this region.*
Keywords. *MFS, Malignancy, Multidisciplinary Team.*

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Introduction

Myxofibrosarcoma (MFS) is a rare malignant soft fibrous tissue tumor, composed of myxoid matrix and pleomorphic spindle cells, and historically composed of myxoid malignant fibrous histiocytoma. MFS mainly affects the elderly patient, with a high incidence among the 50th and 70th years of life, with little predominance in males. The main origin of MFS is in its extremities, especially in the lower limbs, and less in the trunk, head, and neck regions. The tumor usually presented as a painless and hard mass, not mobile, not tender, attached to the skin, slowly-progressive [1] The first case of MFS was reported in 1977 and the MFS is representing nearly 5% of soft tissue tumor in the extremities and trunk [3] the differential diagnosis of MFS as a histologically includes myxoid fat-free spindle cell lipomas, spindle cell melanoma, myxoid spindle cell squamous cell carcinoma, carcinoma myxoid dermatofibrosarcoma protuberans, pleomorphic dermal sarcoma, and atypical fibroxanthoma. The immunohistochemical stains it's a confirm a diagnosis of MFS, as a critical evaluation in sarcoma to ensure appropriate pathological diagnosis and management [4]. We report the case of a high-grade myxofibrosarcoma surgically resected, but with previous recurrence and no metastasis to cervical lymph nodes; to increase awareness and reporting, and better understand epidemiology and management strategies in this region.

Case Presentation

A 50-year-old female, K/C of HCV, presented to our clinic with a history of recurrent huge swelling on the left retroauricular region of the head. are used to notice a small and gradual increase in size. The swelling was painless, attached to the skin, and was not associated with any skin changes, discharge, ulceration, or pruritus. She did not complain of any fevers, weight loss, or night sweats. On examination, she was clinically stable with vitals normal. On local examination, there was a swelling on the retro auricular region of the left side with the firm consistency on palpation and measuring approximately 4cm × 3 cm, tender on deep palpation, not mobile, not hotness on the skin over the swelling (Figure 1) her laboratory finding a hemoglobin of 12 g/dl, with normal leukocyte (7.85 × 10 .9 creatinine of 68.8 µ /ul) and platelet counts (256 × 10 3 /ul), mol/l, AST of 21.8 u/l, ALT of 14.6 u/l and an ESR of 15 mm/h. A Neck ultrasound of the mass reported a hypoechoic, solid, heterogeneous ovoid superficial lesion seen on the left retroauricular region measuring approximately 3.5×4 and 0.18 cm deep from the skin. MRI Neck showing well-defined capsulated mass lesion (4*3.5) seen between left occipital and temporal attachment, posterior to SCM extending retro

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auricular scalloping adjacent bone, with noted heterogenous enhancement, and also another left lesion, left retropharyngeal just medial to the carotid, affecting the carotid sheath with the same radiological character and scattered lymph nodes all in a reactive pattern (Figure 2).

Figure 1. Local Retroauricular mass

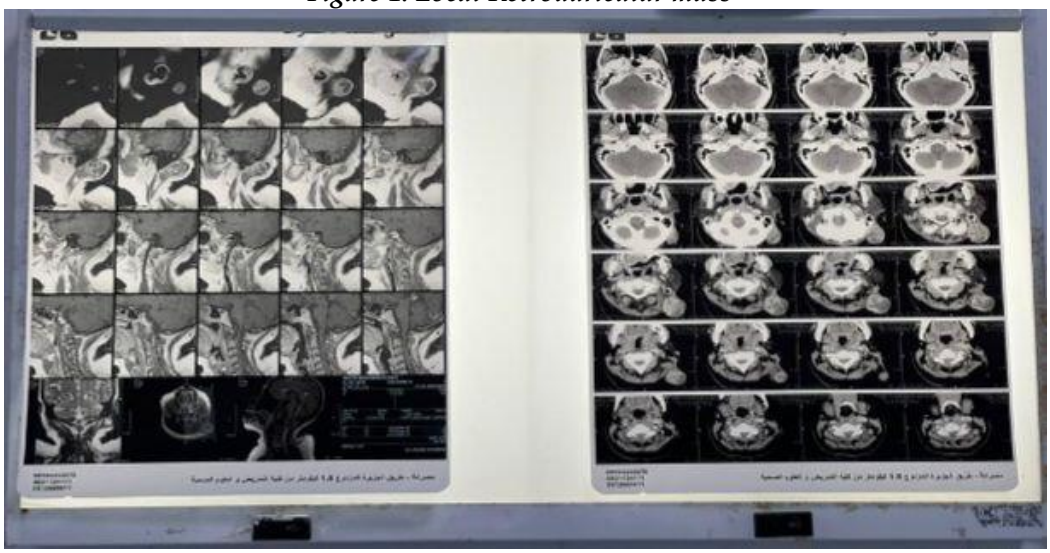

Figure 2. Neck MRI Imaging that revealed a retroauricular swelling

The CT Chest and Abdomen was done to exclude Metastasis and is normal. She was counseled and consented to a wide local excision. Intraoperatively, the whole mass was excised under General anesthesia with a 2 cm normal skin margin (Figure 3) and no evidence of lymph vascular invasion. The lesion was firm on palpation and was submitted for histopathology analysis. The histopathology reported on gross examination is Recurrent High-grade Myxofibrosarcoma with all surgical margins free of tumor invasion, closed margin (Deep margin 3mm), and others more than 1cm, and microscopically, showed the lesion surrounded by a collagenous capsule with moderately cellular as a spindle shaped cells in sheets in a collagenized and myxoid stroma. The lesion was nodular with Mild nuclear atypia. And seen mitosis with necrosis. The margins were free of the lesion.

A diagnosis of a spindle cell neoplasm with myxoid changes was made, and immunohistochemistry was suggested. Immunohistochemistry reported a negative stain in the neoplastic that excluding the structure of carcinoma (Pan CK, SMA, Desmin, CDK4, BRAF V600), with spindle cell neoplasm and myxoid changes were made in the excisional mass. The patient was seen on two follow-up visits after one and three weeks, respectively, for dressing of the wound, and was referred to the national oncology centre for further follow-up. The patient received Adjuvant Radiotherapy and systemic chemotherapy therapy (doxorubicin and ifosfamide) due to high-grade or multiple recurrences.

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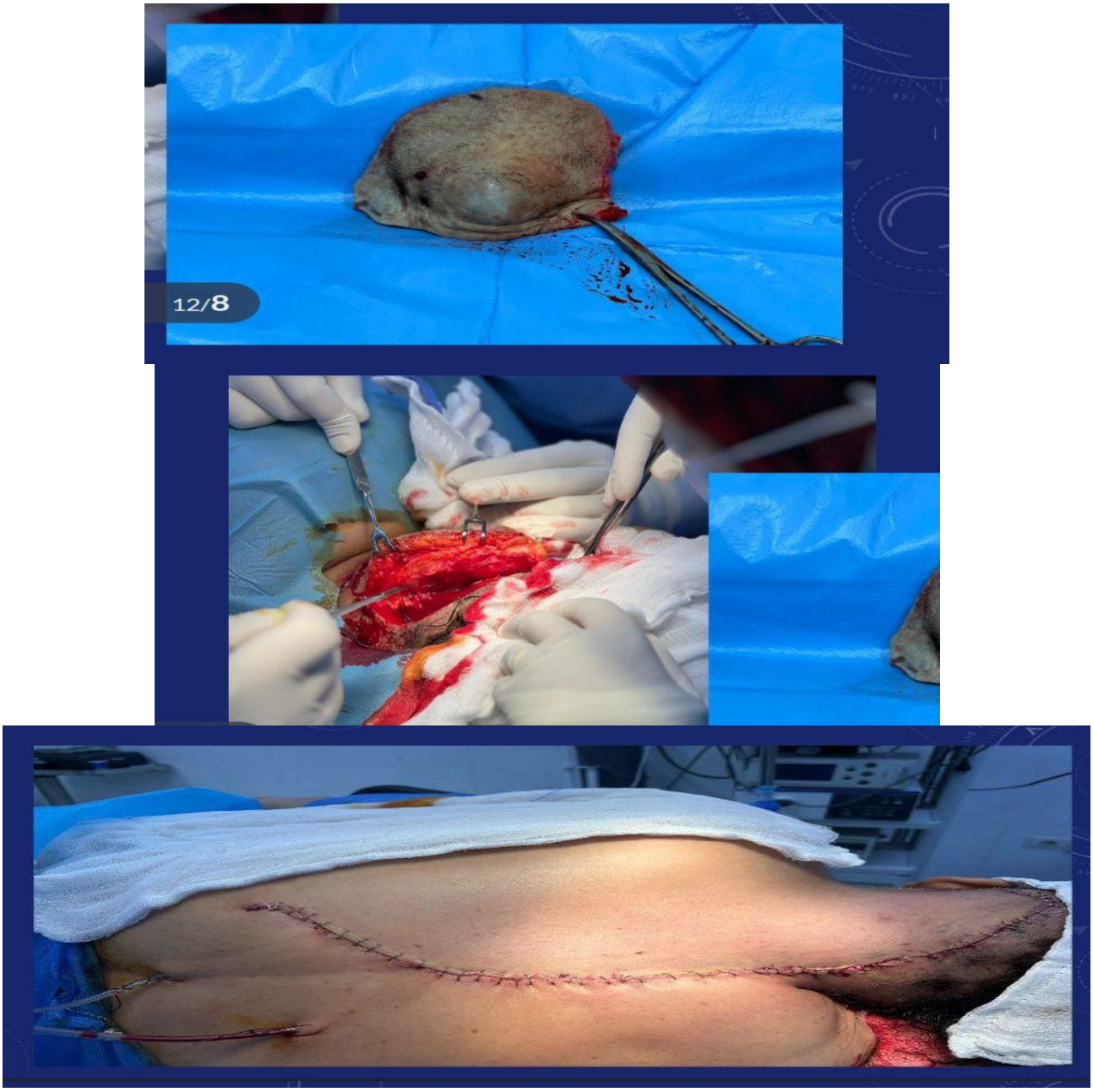

Figure 3. Operative images

Discussion

Myxofibrosarcoma is a rare malignant soft tissue sarcoma tumor; about 5% representing from all tissue sarcoma tumors. It's mainly affected individuals aged from 50 to70 years with a slight predominance in males. common origin from extremities, mainly in the lower limbs [1] and rarely in the trunk, head, and neck. Mostly about two-thirds of the cases developed in the dermal and subcutaneous tissues, and one-third in the fascia and skeletal muscle [2]. The histological classification of Myxofibrosarcoma is low, intermediate, and high grade and characterized by hypocellularity with hyperchromatic and pleomorphic nuclei associated with highly myxoid structures. The high-grade tumors are dense cellular, pleomorphic, with numerous mitotic, hemorrhage, and necrosis, compared with intermediate-grade tumors characterized by more cellular and a lot of nuclear atypia [2].

Skin biopsy is an important diagnostic tool; when taken superficially, it may show misleading features, such as a benign characteristic, but when taken deeper sample show histomorphology features of malignant cells [2]. Therefore, superficial biopsies may classify the tumor as a high-grade tumor or as a benign neoplasm. This characteristic is very important to represent the lesion and is adequate for histopathological examination [2]. Additionally, the immune infiltration of NK cells indicates improvement in the disease-specific survival rate [3]. Higher levels of PD-L1, PD-1, and lymphocytes infiltration are observed in MFS compared to other soft tissue sarcomas, with infiltration of regulatory T

cells that are associated with an increased risk of local recurrence; however, with or without margin positivity [3]. Finally; Head and neck MFS is a very rare site of the tumor, and in these anatomical regions, it can cause misdiagnosis and inappropriate surgery. For this reason, management of these patients in Surgery and radiochemotherapy is strongly recommended to prevent the recurrences [4].

Conclusion

This case presented the diagnosis of myxofibrosarcoma, especially in elderly patients and uncommon locations like the Neck. The patient's presentation with a painless, gradually enlarging mass for two years, along with imaging studies, was highly suspicious of myxofibrosarcoma due to a history of recurrence for two times before. The Histopathological examination is a definitive diagnosis. Surgical excision with clear margins remains the gold standard treatment of Myxofibrosarcoma. Regular follow-up is important due to the high propensity for local recurrence. The Multidisciplinary assessment is important in all MFS, which will be reflected in the best Oncological and functional outcome. This case contributes to the limited data on MFS in sub-Saharan Africa, to increase awareness and reporting, and to better understand epidemiology and management strategies in this region.

Ethical Approval

This case report was conducted in accordance with the ethical principles outlined in the Declaration of Helsinki. The patient provided informed consent for the use of her clinical data and images for academic and research purposes. Institutional approval was obtained from the hospital ethics committee prior to the preparation of this report. All identifying information has been anonymized to protect patient confidentiality.

Conflict of interest. Nil

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