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General Surgery

LOW GRADE MYOFIBROSARCOMA: A RARE TUMOR AT RARE LOCATION

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ABSTRACT Low Grade Myofibrosarcoma (LGMS) represents an extremely rare and atypical tumor composed of myofibroblast with a high recurrence rate. Only few cases have been reported in the literature worldwide. In the present study we describe a rare case of recurrent LGMS in the anterior chest wall which is a rare location for this entity.

KEYWORDS : Myofibrobast, Chest wall, Myofibrosarcoma.

INTRODUCTION

Myofibrosarcomas are rare soft tissue sarcomas affecting the head and neck or extremities of adult patients. Myofibrosarcomas originate from myofibroblasts.[1] The natural course of myofibroblastic malignancies follows a slowly growing, infiltrating pattern of spread, but carries a risk of metastasis, even after many years. The latter attribute can be explained by the lack of a capsule.[2] Patients present with complain of painless swelling or an enlarging mass with or without a mass. The diagnosis of MS relies on pathological findings. Incisional or excisional biopsies are done for diagnosis. The treatment has not been clearly defined, an aggressive surgical resection with wide tumor-free margins, and occasionally, radiotherapy or chemotherapy.[3-4]

CASE PRESENTATION

A 30 year old male presented with a right sided recurrent anterior chest wall mass. Patient had similar complaints 1 year back for which excision and biopsy of the mass was done. The biopsy reports were not available. Now patient is presenting with rapidly enlarging mass in anterior chest wall in right infra-mammary region. General examination revealed no abnormality. On local examination, scar of previous excision was present on inspection at the summit of the swelling. On palpation swelling was non tender, measuring 12X10 cm, firm in consistency, fixed to anterior chest wall and skin over the swelling not fixed.(Fig. 1) A CT scan of the thorax showed features suggestive of Myofibrosarcoma.

Patient was planned for wide local excision under general anaesthesia. Mass measuring 12 cm in transverse direction and 10 cm in longitudinal direction was excised and defect closed primarily. Definitive histopathology was consistent with myofibrosarcoma. Two years after procedure there is no evidence of recurrence.

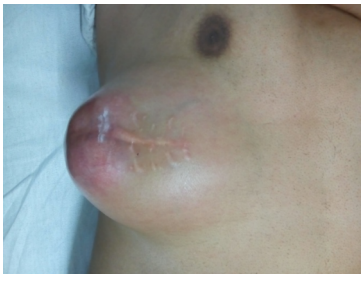

Figure 1: Clinical appearance of LGMS

DISCUSSION

Myofibrosarcoma (MS) is a rare infiltrative low or high-grade mesenchymal tumour that arises usually in a soft tissue. The tumor is composed of myofibroblasts. First described by Gabbiani et al.in 1971, they are mesenchymal spindle- shaped cells that share ultrastructural features with both fibroblasts and smooth muscle cells.[5,6] It has been known to arise mainly at the head and neck regions, although it could be rarely found at the extremities and trunk, and thus, only a small number of cases have been reported in the literature worldwide.[7,8] LGMS usually occurs in adult patients with a slight male predominance. Children are rarely affected.[3] Etiology of myofibrosarcoma remains unknown. It was initially thought to be of purely inflammatory origin but was subsequently found to have the potential for recurrence, infiltrative local growth, and even malignant transformation.[9] Clinically LGMS usually behaves as a slow growing low grade malignant sarcoma and exhibits a pattern of aggressive local spread with common local recurrences and eventual metastatic dissemination, only after a prolong period of time.[3,6,10] The diagnosis of MS relies on pathological findings. Histopathologically most cases are characterized by a diffusely infilterative growth pattern composed of spindle or stellate cells arranged in fascicles. More rarely a prominent collagenous matrix with focal hyalinization and numerous thin-walled capillaries or inflammatory cells have also been reported. Because this tumor shares histological features, such myofibroblastic differentiation, with other malignant neoplasms (i.e. fibrosarcoma), conventional microscopy is generally insufficient for a definitive diagnosis so that immunohistochemical analysis, or electron microscopy may also be required.[3,4] LGMS has a higher recurrence rate. Although LGMS is a low-grade malignancy, 43.9% (18/41) of cases in the oral and maxillofacial region develop local recurrence, although metastases occur rarely.[3] Summarizing the published reports, in cases treated with local excision alone, 75% (8/12) relapsed, while only one of 14 cases with wide excision, with or without post-operative radiotherapy, relapsed. Thus, the treatment of LGMS requires wide excision, with tumour-free margins and post-operative radiotherapy or chemotherapy if needed.

CONCLUSION

LGMS, an aggressive mesenchymal tumor has a predilection for head and neck region. In the present case MS was arising from chest wall, which is a rare site for this tumor. Wide local excision is treatment of choice as tumor has high recurrence rate if tumor free margin is not achieved.

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