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Myxoinflammatory fibroblastic sarcoma: single institution experience and pooled analysis of 138 published cases

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Myxoinflammatory fibroblastic sarcoma (MIFS) is a rare soft tissue sarcoma first recognized and named at the end of nineties. Since then, only few case reports and small series have been published. It is generally considered a low-grade sarcoma that typically arises in the extremities but large cohorts with long-term follow-up are lacking. The aim of this study is to review our experience and perform a systematic review of published cases focusing on the risk of recurrence.

Database of the Rizzoli Institute was retrospectively queried to identify all patients with a pathological diagnosis of MIFS observed from 1997 to 2012. Similarly, all literature of those years was searched to capture all MIFS reported cases.

Five patients underwent surgery for MIFS in our Institute and 133 cases were found in literature. Not all clinical and pathological data were available for every patient. There were 76 men (55%), median age was 45 years (IQR: 34-56). Median size was 3 cm (IQR: 2-5); the most common sites of presentation were hand (47%) and foot (21%). Pain was present at diagnosis in 14/82 patients (17%) with median symptoms duration before surgery of 7 months (IQR: 3-12). Initial surgery was performed for a suspected benign tumor in 88 patients (74%). Marginal or intralesional resections were reported in 45/71 cases (63%), and re-excision during same hospitalization was performed in 32/45 cases (71%). At a median follow-up of 26 months, a recurrence was observed in 26/118 patients. Median time to recurrence was 15 months (IQR: 7-26). Relapse-free survival (RFS) at 1, 3 and 5 years was 93%, 72% and 67%, respectively. Only symptons duration less than 7 months was found to be significantly associated with a worse RFS at univariate analysis (p=0.046). Metastatic disease was observed in 3 patients (one patient with lymph node metastasis, one patient with metachronous lung metastasis and one patient with synchronous lung metastasis observed at our Institute).
MIFS is a rare sarcoma. Clinical findings confirm the “low-grade” nature of MIFS, however, some patients could be affected by aggressive tumour with distant metastases. Extensive preoperative evaluation, wide surgical excision and follow-up are mandatory.

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