A case of recurrent uterine inflammatory myofibroblastic tumour and a review of the literature

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ABSTRACT

Introduction: Uterine inflammatory myofibroblastic tumour (UIMT) is a rare mesenchymal tumour with low malignant potential, which has been known by various names in the past. According to the 2020 World Health Organisation (WHO) definition, it consists of spindle-shaped myofibroblasts and inflammatory cells; its clinical manifestations and imaging features lack specificity, resulting in a high rate of preoperative misdiagnosis. Treatment is primarily surgical, but some cases are invasive and prone to recurrence post-operatively. This paper reports a case of recurrent UIMT and reviews the relevant literature to provide guidance for clinical diagnosis and management. **Case Presentation:** A 53-year-old female patient presented with one month of weight loss and a pelvic mass discovered approximately one and a half months prior, accompanied by frequent urination and severe anaemia. Tumour markers (CA-125, HE4, NSE) were elevated; PET-CT and PET-MRI revealed a 12–13.7 cm mass in the abdomen and pelvis, suggesting possible metastasis. The patient had been misdiagnosed with dedifferentiated liposarcoma at another hospital; however, a consultation at our hospital suggested a spindle cell tumour. The patient underwent extensive surgery combined with six cycles of chemotherapy, and postoperative pathology confirmed the diagnosis of UIMT (ALK-positive). Tumour recurrence was detected in August 2023, and the patient underwent a second surgical intervention; pathology again confirmed UIMT. Follow-up until more than a year revealed no significant abnormalities. While the aetiology of UIMT remains unknown, but it is closely associated with ALK gene abnormalities; ALK D5F3 immunohistochemical testing shows high concordance with FISH results. This condition is easily confused with uterine fibroids and uterine sarcomas; histopathological examination is key to definitive diagnosis. Surgery is the treatment of choice, with targeted therapy (such as alectinib) serving as an adjunct. Prognosis is related to tumour size and the extent of surgical resection; recurrence does not necessarily indicate a poor prognosis. **Conclusion:** UIMT is clinically rare and prone to misdiagnosis; diagnosis relies on histopathological examination. Surgery combined with close postoperative follow-up constitutes the primary management strategy, with targeted therapy serving as an adjunctive option. Current research consists largely of small-sample case reports; large-scale, multicentre studies are required to refine clinical management guidelines.