

A sarcoma of 23 years' duration: symptom duration is not a reliable parameter to exclude malignancy

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This report describes a 31-year-old woman with a 23-year history of a right buttock mass that was otherwise asymptomatic, but was proven to be a low-grade fibromyxoid sarcoma (a fully malignant soft tissue tumour with a potential for distant metastasis). This case illustrates that a long-standing tumour does not necessarily imply a benign pathology. A vigilant approach should be taken for any tumours that are of significant size (larger than 5 cm).

Case report

A 31-year-old woman consulted her gynaecologist for investigation of an ovarian cyst in April 2010. Whole body fluorodeoxyglucose (FDG) positron emission tomography scan incidentally found FDG uptake (standardised uptake value max, 1.6) in the right gluteal region. There was no evidence of any lung or bone metastasis. She was referred to us for further investigation.

Upon subsequent questioning, she recalled having noticed buttock asymmetry since the age of 8 years. The right buttock was proportionally larger than that on the left side, but there had been no definite increase in size since puberty. In all other respects she was asymptomatic and free of pain. Her activities of daily living were unaffected, and she enjoyed active sports, nor had she sought any medical advice for the lesion. Physical examination revealed a 12-cm non-tender intramuscular (gluteal muscle) mass. The sciatic nerve was intact.

Investigations yielded a normal blood picture, as well as normal routine renal and liver function test results and plasma calcium and phosphate levels. Magnetic resonance imaging (MRI) showed a well-circumscribed 6.3 x 7.2 x 9.3 cm lesion in the gluteus medius. It was homogeneously isointense to muscle on T1-weighted imaging, and on T2-weighted imaging it was heterogeneous with patchy contrast enhancement. Incisional biopsy yielded a spindle cell lesion. Mitosis and necrosis were not apparent. Overall, it was suggestive of a low-grade malignancy. Complete resection of the intramuscular soft tissue tumour was carried out, leaving a 1-cm margin in all directions except the deep surface where the tumour was abutting the periosteum of the iliac bone. Resection was therefore through the sub-periosteal plane. The wound was then closed primarily.

Pathology

The tumour appeared to be intramuscular and measured 6.5 x 5.3 x 12 cm (Fig 1). Microscopically, it was encapsulated by a thin layer of fibrous tissue, and composed of bland-looking spindle cells in an admixture of heavily collagenised, hypocellular, and myxoid zones and cellular nodules (Fig 2a). In areas, the nodules were arranged in short fascicles or whorled patterns (Fig 2b). Scattered small-sized curvilinear blood vessels and vessels with hyalinised walls were also noted. Collagen rosettes featuring a collagenous centre and palisading of spindle cells in various stages and sizes were very evident, but there was no mitosis or necrosis (Fig 2c). The thin fibrous capsule touched the deep margin (periosteum of the ilium) focally. The diagnosis was low-grade fibromyxoid sarcoma.

Fluorescence in-situ hybridisation evaluation following hybridisation-detected rearrangement of the *FUS* gene (16p11) with a standard pattern in 5% of the interface cells examined, and a complex rearrangement pattern with rearrangement plus duplication of the native *FUS* gene in 46% of the tumour cells. This was indicative of evolution of the original clone. The findings were consistent with the morphological diagnosis of low-grade fibromyxoid sarcoma.¹

Progress

The patient was given postoperative irradiation for better local disease control. No

Key words

Fibrosarcoma; Sarcoma; Soft tissue neoplasms

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FIG 1. Gross picture of the resected specimen

長達二十三年病史的肉瘤：病徵的長短不足以排除惡性腫瘤

一名31歲女士右臀部的腫塊已達23年，除此以外並無其他症狀。切除後證實為低度惡性纖維黏液樣肉瘤，即是一個有可能轉移的全惡性軟組織腫瘤。本病例顯示出現病徵時間的長短不足以排除惡性腫瘤的可能性。醫生對於患有很大腫瘤（大於5 cm）的病人應有高度警覺性。

recurrence was detected at the 9-month post-operative follow-up.

Discussion

It has been widely accepted among clinicians, including some orthopaedic surgeons, that a long-standing soft tissue tumour is a reliable indication of its benign nature. For example, Mannan and Briggs² stated "A lesion that has been present for 20 years and not changed in appearance is unlikely to be malignant". In our case, the lesion had been present for 23 years, and had not undergone any recent increase in size, yet, turned out to be a malignant sarcoma.

Size and depth of lesion are also widely used clinical parameters to assess whether a soft tissue tumour is benign or malignant.³ In patients older than 50 years, any soft tissue lesion greater than 5 cm in maximum diameter carries a 20% risk of malignancy, whilst such a deep-seated mass located proximally carries a 50% risk of malignancy. Accordingly, it has been advocated that an MRI scan⁴ and biopsy should be performed before surgical removal of any deep-seated (sub-fascial) lesion or mass larger than 5 cm in its maximum dimension, in order to avoid unplanned incomplete resection of a malignant tumour. In fact, Wong et al⁵ suggested that 83% patients with unplanned incomplete resections might have avoided subsequently more aggressive salvaging procedures, had the rule to adequately investigate 'sub-fascial' or '5-cm' lesions been followed.

Low-grade fibromyxoid sarcoma was first reported as a distinct pathological entity by Evans in 1987.⁶ It is a deceptively innocuous fibroblastic spindle cell tumour that manifests aggressive biological behaviour. Microscopically, it is characterised by contrasting fibrous and myxoid areas of varying size with swirling whorled growth patterns and arcades of curvilinear blood vessels. Poorly formed collagen rosettes featuring hyalinised collagen surrounded by a cuff of epithelioid fibroblasts are sometimes seen.

Tang et al⁷ comprehensively reviewed all reported cases of low-grade fibromyxoid sarcoma in the literature (up to the year 2008). Of the 273 cases,

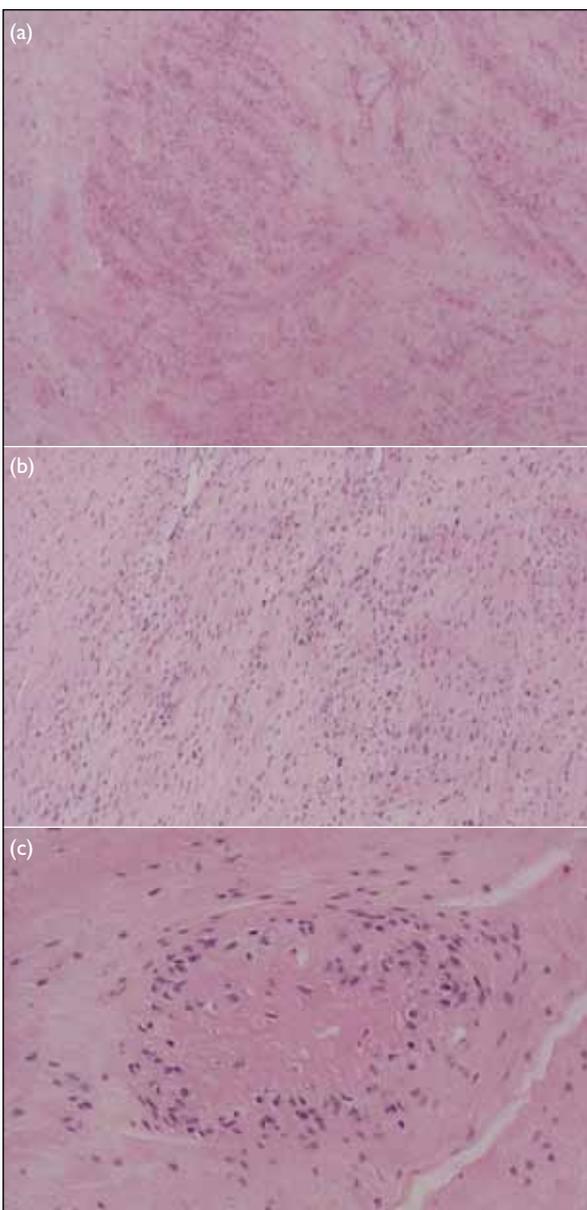


FIG 2. (a) Curvilinear blood vessels in a hypocellular fibromyxoid background (H&E, x 20). (b) Spindly tumour cells arranged in focally swirling pattern (H&E, x 40). (c) Giant cell rosettes or collagen rosettes with tumour cells surrounding a nodule of hyaline collagen (H&E, x 100)

the duration of clinical symptoms was reported in only 52 cases. Thirteen (25%) of these patients had had the lesions for more than 5 years; six (nearly 12%) had had them for more than 20 years. While this sarcoma is fully malignant, patients with this condition may present for medical consultation after a considerable period.

Data from older literature show that low-grade fibromyxoid sarcoma carries a local recurrence rate of about 68%, a 41% risk of metastases, and an 18% risk of causing death, which suggest that it can become very aggressive.⁸ However, the poor outcomes encountered in this early report may have been due to inappropriate management, as most of the lesions were treated as benign. Folpe et al⁹ reported a local recurrence rate of 9%, a metastatic rate of 6%, and a death rate of 2%. Tang et al's review⁷ reported that in 273 cases, the local recurrence rate was 29% and the metastasis rate was 18%. Due to the high local recurrence and metastasis rates, as well as its delayed presentation, long-term follow-up is essential.

The preferred treatment⁷ is surgical resection with adequate margin including radical or wide en-bloc resection; adjuvant radiotherapy was

reported as curative but efficacy in the long-term control has not been proven.

Conclusion

While a long duration of symptom from a soft tissue mass is generally taken as a reliable indicator to differentiate benign from malignant lesions, our patient with a low-grade fibromyxoid sarcoma serves to illustrate an important exception to this widely held myth. Recognition of such clinical features is crucial to avoid underdiagnosing malignant soft tissue tumours and offering inappropriate limited resections. Therefore, any tumours of a size greater than 5 cm or those that are deep-seated should have appropriate imaging (an MRI scan) and a preoperative biopsy, irrespective of the duration of the symptoms. In cases of doubt, referral to a specialised tumour centre for further assessment may minimise risk of unplanned incomplete resection.

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