

Proliferating myositis and proliferating fasciitis: benign lesions often misdiagnosed as sarcomas

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(Index words: Clinical features, immunohistochemistry, histology)

Introduction

Proliferative myositis and proliferative fasciitis are benign lesions that are often misdiagnosed as sarcomas [1, 2], leading to unnecessary mutilating surgery and chemotherapy. We report two such initially misdiagnosed cases.

Case history

Case 1

A 77- year old man presented with painless, mobile swelling of the right anterior chest wall of 3 months' duration. Ultrasound scan revealed an elliptical lesion measuring 4 x 3 cm within the pectoralis major muscle. It had a hyperechoic centre and hypoechoic periphery. A diagnosis of a spindle cell sarcoma was made on an incisional biopsy. This diagnosis was reviewed and confirmed by a second pathologist and a radical mastectomy was performed. The patient remained well 26 months after surgery.

Case 2

A 25-year old woman presented with a well demarcated rapidly enlarging tender nodule measuring 2 cm in diameter in the right supraclavicular area of 2 weeks' duration. The lesion was excised. It was diagnosed as a rhabdomyosarcoma. The patient is without recurrence 24 months after surgery.

Both cases were subsequently referred to the third author for review. In Case 1, the lesion comprised a poorly demarcated intramuscular spindle cell proliferation. There were large ganglion-like cells with basophilic cytoplasm, vesicular nuclei and prominent nucleoli in a background of plump spindle shaped cells (Figure 1). There were scattered mitotic figures. The stroma was myxoid and showed red cell extravasation. There was extension of lesional cells in between atrophic muscle fibres. The histological features were those of proliferative myositis.

The lesion in Case 2 was composed of cells similar to those in Case1, but without involvement of skeletal muscle. There were scattered mitotic figures. The

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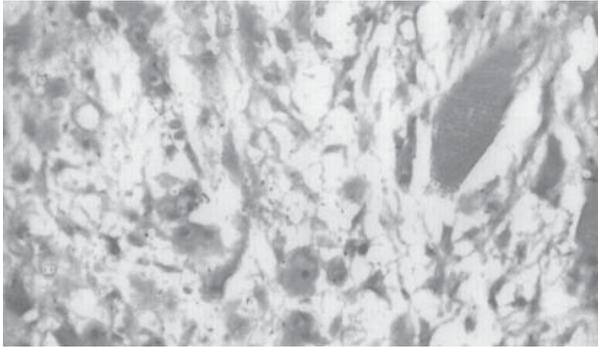


Figure 1. Proliferating myositis showing large ganglion-like cells in a spindle cell background (H & E stain x 400).

ganglion-like cells stained positively with smooth muscle actin and negatively with desmin, myoglobin and myoD1, which are specialised immunohistochemical markers for skeletal muscle differentiation. The histological features were those of proliferative fasciitis.

Discussion

Proliferative myositis and fasciitis are benign, self-limiting mass forming processes involving intramuscular and subcutaneous sites respectively [1, 3]. They predominantly affect middle-aged or older adults. Proliferative myositis shows a predilection for the shoulder girdle, flat muscles of the trunk, upper arm and thigh. Proliferative fasciitis shows a predilection for the upper and lower limbs. A characteristic clinical feature common to both lesions, which should alert clinicians and pathologists to the diagnosis, is the history of rapid growth within a short period. Unlike sarcomas, these lesions are almost always less than 5 cm in maximum dimension and are most often less than 3 cm [4]. Few sarcomas will grow as rapidly as these lesions and the ones that do so will have areas of necrosis, unlike proliferative fasciitis and proliferative myositis.

Proliferative fasciitis and myositis mimic sarcoma due to the microscopic appearance of bizarre cells, increased mitotic activity and infiltrative growth pattern. The large basophilic ganglion-like cells stain positively with smooth muscle actin and muscle specific actin. This should not be interpreted as representing skeletal muscle differentiation because there is negative staining with myoglobin and desmin. A characteristic and distinctive architectural feature in proliferative myositis which helps to distinguish it from a sarcoma is extension of the lesion along fibrous

septa and between individual atrophic muscle fibres giving rise to a “checkerboard appearance”. Both lesions are devoid of atypical mitoses and necrosis. In fine needle aspirations, these lesions can be distinguished from malignant cells by their thin, smooth nuclear membranes and fine chromatin pattern [5].

The cause and mode of development of proliferative myositis and fasciitis remain unexplained. Although traditionally regarded as reactive lesions, there is accumulating evidence mainly from cytogenetic analysis that these are clonal proliferations [6]. In both conditions the prognosis is excellent and the lesions can be adequately treated with local marginal excision. Recurrence is extremely infrequent [2, 6, 7]. Any microscopic residual lesional tissue undergoes spontaneous attrition by scarring [6]. Awareness of these entities and correct diagnosis will help to avoid unnecessary mutilating surgery and chemotherapy.

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