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Cytological Diagnosis of Deep Fibromatosis: A Case Series

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ABSTRACT

Fibromatosis forms a spectrum of clinico-pathologic entities, characterized by infiltrative proliferation of fibroblasts/ myofibroblasts that lack malignant cytologic features. The present study deals with the cytomorphologic appearance of fibromatosis and its cytologic differential diagnosis. FNAC was performed in 4 cases of fibromatosis with tumorous clinical presentation using a 22-gauge needle and 20ml syringe. Air dried smears stained with MGG were studied. FNAC revealed moderately cellular smears and showed scattered as well as aggregated spindle cells, some of them being embedded in hypocellular dense stromal fragments. The cells were oval to spindle shaped with tapering ends, bipolar nuclei, bland nuclear chromatin and generally inconspicuous nucleoli. Based on morphology and clinical presentation, a diagnosis of fibromatosis was rendered. Histopathological examination confirmed the cytodiagnosis of fibromatosis in every instance. Based on clinical history, physical findings, imaging data and cytologic observations, a correct diagnosis of fibromatosis can be rendered in most instances if one is conversant with the cytologic features.

KEYWORDS Fibromatosis, Fine needle aspiration, Cytology, Cytodiagnosis

INTRODUCTION

The term fibromatosis was originally proposed by Stout for mesenchymal tumors characterized by presence of fibroblasts, myofibroblasts and marked collagen [1]. Fibromatosis comprises a broad group of benign fibroblastic proliferations of similar microscopic appearance whose biologic behavior is intermediate between that of benign fibroblastic lesions and fibrosarcoma. Like fibrosarcoma, fibromatosis is characterized by infiltrative growth and a tendency toward recurrence, but these tumors never metastasize. [2]. We present 4 cases of fibromatosis, which were diagnosed on cytology, along with discussion of their differential diagnoses and review of literature.

MATERIALS AND METHODS:

Between the years 2012 and 2015, cases that were diagnosed as fibromatosis on cytology, with available histology, were reviewed for this study. Clinical and radiographic information was recorded. Cytomorphologic features were noted and cytologic diagnosis was confirmed on histology and an immunohistochemical panel was done in 2 out of 4 cases.

RESULTS:

The clinical features, duration of complaints, radiographic findings, cytomorphologic features and cytologic diagnosis of 4 cases of fibromatoses are summarized in Table 1 and Table 2.

Table 1. Clinical features of 4 cases of fibromatosis

S. NO.	Age (yrs)	Gender	Size (cm)	Site of tumor
1	30	M	6	Upper arm
2	34	F	10	Proximal thigh
3	40	F	12	Shoulder
4	42	F	16	Gluteal region

Table 2. Radiological and cytological features of 4 cases of fibromatosis

S. NO.	RADIOLOGICAL FINDINGS	CYTOLOGICAL FINDINGS	CYTOLOGICAL DIAGNOSIS
1	No in house studies prior to FNA	Moderate cellularity, with collagenized stroma, embedded spindle cells	Consistent with fibromatosis
2	Infiltrative soft tissue mass, not categorized	Moderate cellularity, with clusters of bland spindle cells	Favour fibromatosis
3	No in house studies prior to FNA	Moderate cellularity. Bland spindle cells embedded in collagenized stroma	Consistent with fibromatosis
4	Consistent with recurrent fibromatosis	Low cellularity, few spindle cells and fragments of collagenized stroma	Consistent with fibromatosis

In our study, out of 4 cases of fibromatosis, 3 were female and one male, with a mean age of 36.5 years (range 30-42 years). In 3 cases, reported duration of complaints varied from 12 months to 20 months (mean :14 months).1 patient had history of prior resection at the same site and now had a duration of complaint of 6 months. Various sites of tumor in our study were upper arm, proximal thigh, shoulder and gluteal region. On examination, the size of the tumors varied from 6 cm to 16 cm (mean 44). In all the 4 cases, swelling was firm in consistency and was fixed to underlying deep muscle.

Out of 4 cases, in 2 cases radiographic findings were not available prior to fine needle aspiration cytology. Of the remaining 2 cases, in one case the radiographic impression prior to FNAC was consistent with recurrent fibromatosis and another with infiltrative soft tissue mass, not further categorized. Fine needle aspiration cytology from the cases revealed low to moderately cellular smears and showed small cohesive cell clusters, dispersed cells and collagenised stromal fragments. The cells were oval to spindle shaped, showing mild anisokaryosis, having fusiform nuclei, finely granular chromatin and small nucleoli. Well demarcated, pale cytoplasm was seen in preserved cells and bipolar cytoplasmic processes are seen. The cells were often embedded in or were in close proximity to stroma. Mitosis were not seen in any of the cases. (Figure:1 and 2). Based on morphology, cytologic diagnosis of fibromatosis was given.

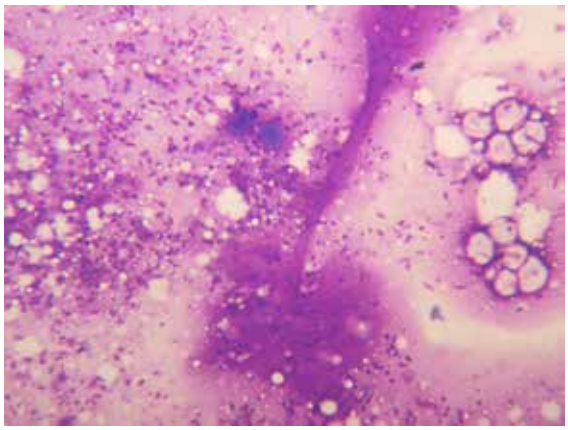

Figure 1. Low power view of fine needle aspiration material from deep fibromatosis shows dispersed cells and fragments of collagenised stroma (May Grunwald Giemsa stain, original magnification x 100).

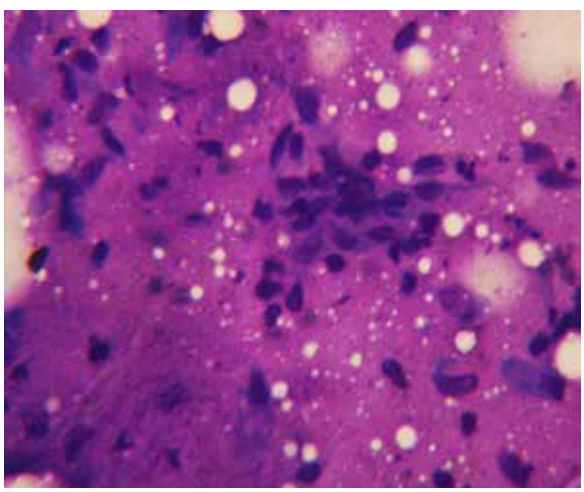

Figure 2. High power view shows loosely attached fibroblast-like cells embedded in stroma (May Grunwald Giemsa stain, original magnification x 100).

Histopathologic examination in all the 4 cases confirmed the diagnosis of fibromatosis. (Figure:3). In 2 cases, out of 4, immunohistochemical panel was done, which showed nuclear positivity for β-catenin (Figure 4). Desmin, CD-34, and c-Kit were negative in these 2 cases.

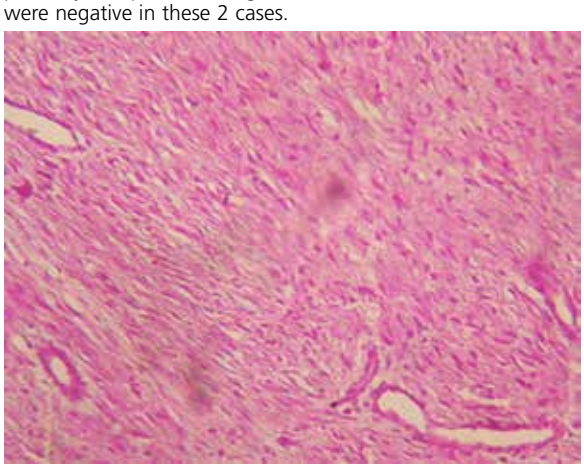

Figure 3. Low-power view of fibromatosis with bland appearing spindle shaped cells arranged in short intersecting fascicles separated by variable amount of collagen (hematoxylin-eosin, original magnification, x 100).

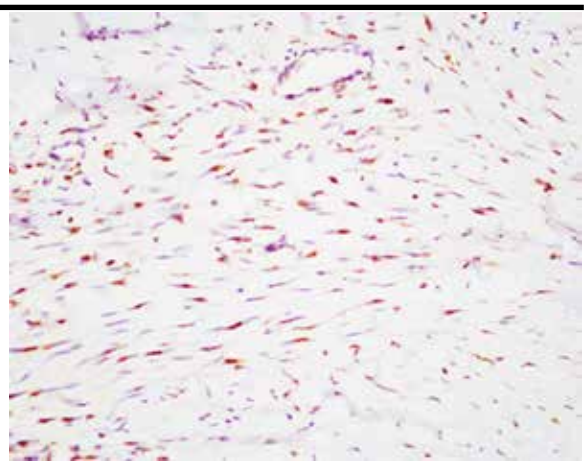

Figure 4. Nuclear beta-catenin immunoreactivity in fibromatosis (Original magnification x 100).

DISCUSSION

Fibromatosis are the group of locally aggressive neoplasms. They share the capacity for infiltrative, destructive, and commonly recurrent growth but have no capacity to metastasize. They are commonly sub-classified into superficial (e.g. palmar and plantar) and deep (desmoid) groups [2,3]. Deep (musculoaponeurotic) fibromatosis, involve deep structures, particularly musculature of trunk and extremities [2].

All fascial and musculoaponeurotic fibromatosis are capable of recurrence after excision, the recurrence rate of individual entities varies substantially [2]. There are few case series as well as reports of FNAC in fibromatosis which had recurrence of the same lesion [4, 5, 6]. In our study 25% of cases had a history of recurrence after surgery at the site of FNAC.

Aspirates from fibromatosis have variable cellularity [7, 8]. Cellularity is usually low (6 of 8 reviewed tumors) but does show a range, and some patients have tumors with moderate cellularity (2 of 8 reviewed tumors) [5]. Aspirates from aggressive fibromatosis are more cellular although mitosis are rare to absent [7]. Due to often abundant collagenous stroma, desmoid fibromatosis are very firm to palpate and a rubbery resistance is felt when needling; often vigorous aspirations are needed to collect sufficient material for examination [9]. Based on our findings,3 out of 4 cases had moderate cellularity and small amount of collagenous stroma, while, in one case, cellularity was low and there was abundance of collagenous stroma.

Differential diagnoses of fibromatosis include scar or keloid, mesenchymal repair, nodular fasciitis, schwannoma, leiomyoma, solitary fibrous tumor (SFT), gastrointestinal stromal tumor (GIST), fibrosarcoma, synovial sarcoma and malignant melanoma [10, 11]. Low grade malignant peripheral nerve sheath tumor (MPNST) as well as low-grade fibromyxoid sarcoma may also be cytologically misdiagnosed as desmoid tumor [9].

Keloid are less cellular than fibromatoses. In mesenchymal repair, the number of stromal cells is less than observed in fibromatoses, the cells display more stellate configurations and a greater degree of inflammation may be noted [2]. Uniform spindle cell population, fragments of collagenised stroma, absence of myxoid background and ganglion like cells exclude nodular fasciitis; while tumor fragments with elongated or comma shaped nuclei in fibrillar background are hallmarks of neurilemmoma [9].

In leiomyoma the most important clue to the diagnosis is the presence of blunt ended,'cigar shaped', elongated or ovoid nuclei of varying size, nuclei sometimes contain vacuoles. In solitary fibrous tumor the yield is usually haemorrhagic and in tissue fragments, branching network of capillaries is usually seen [9].

Fibrosarcoma is much more cellular than fibromatosis, and cells are arranged in a more consistent sweeping fascicular growth pattern [2]. In synovial sarcoma, branching capillaries with attached tumor cells are seen which have spindly to ovoid to rounded nuclei. Mitosis is commonly seen and so are mast cells.

Low-grade fibromyxoid sarcoma have alternating collagenous and myxoid background and presence of curvilinear vessel distinguishes it from fibromatoses. In MPNST the tumor cells are spindly with fusiform nuclei which are comma-shaped, wavy or buckled, in a fibrillary background.

Invariably most fibromatosis have β-catenin nuclear staining. β-catenin reactivity is also seen in SFT, endometrial stromal sarcoma and synovial sarcoma. SFT is also known to be positive for CD34 and CD99. Smooth muscle neoplasm is positive for desmin and majority of GIST show positivity for c-Kit. [7]. Therefore, a recommended panel based on similar appearing spindled tumors should include at least b-catenin, c-Kit, CD34 and desmin [5].

To conclude, FNAC is reasonably useful in diagnosing fibromatosis in association with clinical and radiological findings. Although histologic confirmation is preferred and b-catenin staining is extremely useful.

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