



Fibromyxoma of bone : A case report and review of the literature

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In this study, a case of fibromyxoma of the proximal femur in a 59-year old woman is reported. The classification of this rare bone tumour is still a matter of debate and some investigators have suggested that these lesions represent a degenerative form of fibrous dysplasia. Some authors make a further distinction between fibromyxoma and myxoma of bone. In a review of 23 cases of fibromyxoma and five cases of myxoma, no differences in clinical, radiographic and biologic behaviour between fibromyxoma and myxoma were found. Apart from the age at diagnosis, the most important difference between fibromyxoma and myxoma was the degree of myxoid matrix. Therefore, we suggest that extragnathic myxoma is a regressive variant of extragnathic fibromyxoma and should be termed as the same entity. In contrast to monostotic fibrous dysplasia fibromyxoma / myxoma often causes pain and presents as a Lodwick IC lesion with a soft tissue mass. Therefore, fibromyxoma / myxoma should be distinguished from fibrous dysplasia because of its different clinical and radiographic features.

Keywords : fibromyxoma of bone.

INTRODUCTION

Fibromyxoma of bone is a rare benign fibrous tumour. Twenty-eight cases of extragnathic fibromyxoma and myxoma have been described since 1977 in the literature (1, 2, 5, 6, 10-14, 17, 18, 20, 22). It is usually found in the jawbones (3, 4, 6, 10, 11). Extragnathic fibromyxoma with abundant myxoid

tissue has been designated as myxoma, and in the presence of large amounts of differentiated fibrous tissue has been designated as fibromyxoma (3, 4, 11, 13, 17, 18, 20, 22).

In contrast to gnathic fibromyxoma the existence of fibromyxoma as a specific entity is still a matter of debate. In 1970, Arora and Hunter (3) regarded extragnathic fibromyxoma or extragnathic myxoma as resulting from myxoid degeneration of bone tumours like chondrosarcoma or fibrosarcoma. Furthermore, it can be difficult to distinguish between fibromyxoma and chondromyxoid fibroma (6, 9, 17). In the opinion of Unni (23) many cases of fibromyxoma reported in the literature are examples of myxoid fibrous dysplasia.

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Certainly, the rare occurrence of this fibrous tumour is a reason for difficulties in classifying fibromyxoma as a specific entity and to distinguish it from other tumours containing myxoid tissue. The aim of the current study was to determine possible clinical, radiological, histopathological and biological differences between fibromyxoma and myxoma of bone. Furthermore, clinical, radiological and histopathological differences between fibromyxoma / myxoma and other fibrous lesions of bone – especially fibrous dysplasia – are pointed out in a literature review.

CASE REPORT

A 59-year-old woman was admitted to our department with a 6-month history of progressive pain about her left trochanter. First, the pain occurred mainly while walking, but in the last few months it was also present at rest. The range of motion of the left hip was not limited. Clinical examination revealed tenderness over the greater trochanter. Medical history revealed no symptoms of polyostotic fibrous dysplasia.

Radiographs showed an osteolytic lesion of approximately 5.5 cm in diameter with a surrounding osteosclerotic rim involving the medullary cavity of the lateral neck of the femur and the greater trochanter (Lodwick (16) Grade IA) (fig 1). On computed tomography scans there were no signs of cortical destruction and / or periosteal reaction. On magnetic resonance imaging the lesion had a cystic appearance and Gadolinium application resulted in a moderate contrast enhancement (fig 2a-c). The fluorodeoxyglucose positron emission tomography revealed no elevated FDG-uptake (tumour to background ratio, 1.0). Taking all data together, we expected a benign lesion in nature. Therefore, we did a complete curettage of the lesion. The resulting defect was completely filled with autogenous corticocancellous bone and bovine spongiosa.

Histopathologically, the curettage showed a tumour composed of fibrous tissue with a variable myxoid stroma of low cellularity without a lobular architecture or a chondroid matrix. The tumour had an alcian blue-positive matrix. The fibrous tissue



Fig. 1. — Radiograph of the left hip showing a well marginated osteolytic lesion with a sclerotic rim involving the medullary cavity of the lateral neck of the femur and the trochanter major.

was reactive for vimentin and negative for S-100 protein. The proliferation index (Ki-67 index) was < 1%. The diagnosis was fibromyxoma (fig 3).

On follow-up examination 30 months after surgery, there was no evidence of recurrence (fig 4) or bone resorption and the patient was free of pain.

DISCUSSION

The pathologic classification of fibrous bone lesions remains difficult (17). The existence of extragnathic fibromyxoma as a specific entity is still a matter of debate (3, 11). Particularly, the differentiation of fibromyxoma from fibrous dysplasia

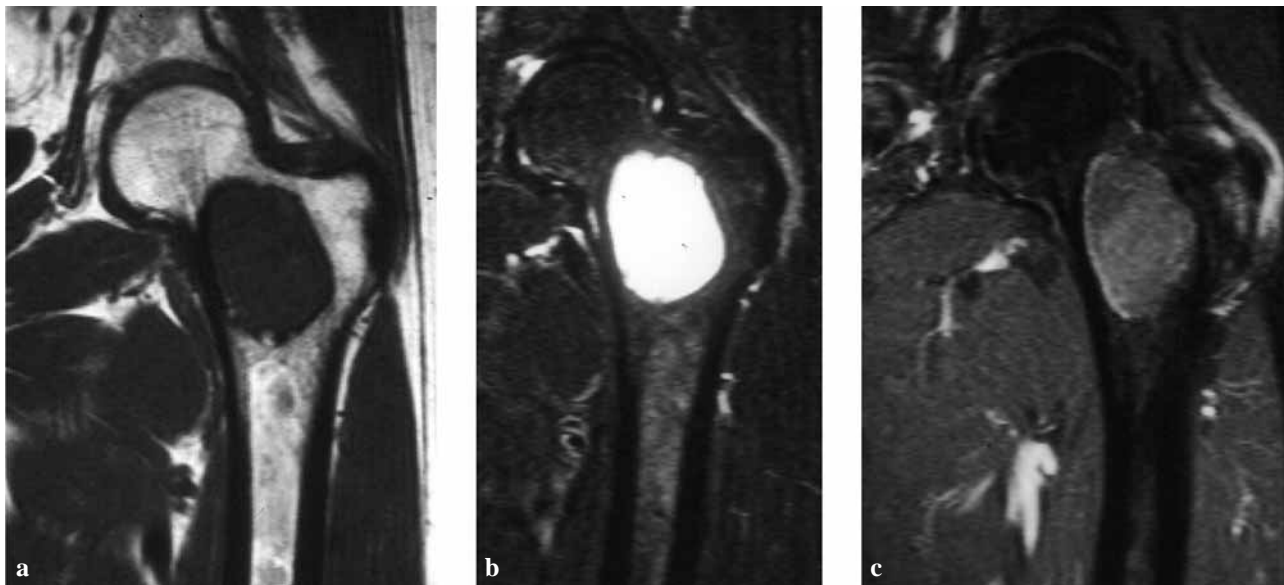


Fig. 2. — MRI of the lesion in the coronal plane. The T1-weighted image (a) shows a hypointense lesion. The T2-weighted image (b) shows a hyperintense lesion. The diagnosis of a bone cyst cannot be excluded. The moderate contrast enhancement after Gadolinium application (fat-saturated) (c) indicates that a solid lesion is present.

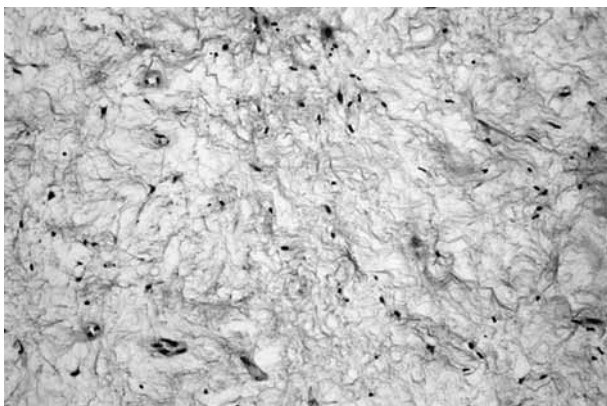


Fig. 3. — Morphology of fibromyxoma ($\times 290$, H & E staining) shows a stroma with many spindle-shaped cells in between a myxoid ground substance.

remains controversial (23). Some authors make a further distinction between fibromyxoma and myxoma of bone (17, 18). By reviewing the literature, possible differences or similarities between these entities should be worked out.

In the current study a case of an extragnathic fibromyxoma is reported. Since 1978, 23 cases of extragnathic fibromyxoma have been reported in

the literature (1, 2, 5, 6, 10, 12-14, 17). In these studies, the tumour affected 13 female and ten male patients observed between 2 to 68 years (median age at diagnosis : 33 years). Predilection sites were the metaphyses of the long bones. The most common reported sites were the proximal femur (30.4%) and the proximal tibia (30.4%). A location in flat bones was less common (12, 14). Pain was present in 82.6% of cases and was the most common symptom in fibromyxoma. In contrast, pain in monostotic fibrous dysplasia is rare (8).

Also in these studies the radiological feature of fibromyxoma was always a lytic lesion (1, 5, 12, 17). Often, fibromyxoma appeared like in the current case as a central, well defined osteolytic lesion, which was not locally aggressive and had a surrounding sclerosis without signs of malignancy (Lodwick (16) Grade IA) (4, 10, 12, 17). Sometimes the tumour presented radiographically as a malignant lesion with ill-defined margins and interruption of the cortex (Lodwick IC), with extension into the soft tissue (1, 2, 5, 14). Because of the variable radiologic features of fibromyxoma, there are various differential diagnoses of benign and malignant lesions such as fibrous dysplasia, chondromyxoid



Fig. 4. — Radiograph of the left hip showing a good osseointegration of the autologous cancellous bone 30 months post-operatively.

fibroma, bone ganglion cyst, solitary bone cyst, aneurysmal bone cyst, giant cell tumour, low-grade chondrosarcoma or plasmocytoma (2, 10, 14, 17).

In contrast to fibromyxoma with a variable radiographic feature, monostotic fibrous dysplasia mostly presents radiologically as a Lodwick IA (16) lesion without a soft tissue mass (8). In fibromyxoma and myxoma, a usually slow-growing extraosseous mass has been reported in 30.4% of patients (1, 5, 6, 12-14). Furthermore, in the current case, in MRI scans the lesion was suggestive of a cystic lesion with homogenous high-signal intensity on T2-weighted images without signs of a solid tumour component. Therefore, fibrous dysplasia was unlikely. In the literature, cystic degeneration

of fibrous dysplasia is reportedly rare, but in these cases pathology findings showed that more than 90% of the lesion was cystic (19). Compared with this, in the current case no serous fluid was found.

According to the literature, macroscopically fibromyxoma is a grey to white, firm to gelatinous lesion with varying amounts of calcification (1, 2, 5, 13). In general, as in the current case, fibromyxoma consisted histologically of a mixed myxoid and fibrous stroma. In the stroma were many stellate or spindle-shaped cells (fig 3). Histologically, the curetted material was examined completely. Other tumours with myxoid tissue, especially chondromyxoid fibroma and also other tumours such as chondroma, chondrosarcoma, fibroma, fibrosarcoma, bone ganglion, or fibrous dysplasia could be excluded (2, 4, 6, 18).

Some authors have suggested that extragnathic fibromyxoma bears certain similarities to chondromyxoid fibroma. In general, patients with chondromyxoid fibroma are younger (second and third decades) than patients with fibromyxoma (median age at diagnosis : 40 years) (7, 17, 23). Chondromyxoid fibroma is located in the tibia in approximately 30% of patients (7, 23). In contrast to fibromyxoma, a localisation in the proximal femur is uncommon (7). Unni (23) reported a definite male predilection in chondromyxoid fibroma, whereas in fibromyxoma no gender predominance was found. Histologically, fibromyxoma contained no chondroid structures with slight lobulation (2, 18).

We did not diagnose myxoma in the current patient. Myxoma of extragnathic bones was first described by Stout (21) in 1948. So far, only five cases of extragnathic myxoma have been described (18, 20, 22). In a large series of 11,000 bone lesions, the Mayo Clinic reported three myxomas of extragnathic bones, which were all located in the femur (18). Two other myxomas were found in the distal humerus and in the proximal tibia, respectively (20, 22).

Myxoma bears many similarities to fibromyxoma. The clinical and radiographic presentations of myxoma do not differ from fibromyxoma. Myxoma and fibromyxoma are benign lesions without the potential to metastasise (18, 20, 22). The most important difference between both tumours

is the degree of myxoid matrix and the age distribution in patients affected. Patients with myxomas are over the age of 60 years, whereas patients with fibromyxomas have an even age distribution. Therefore, we suggest that fibromyxoma and myxoma are the same entity with similar clinical and radiographic appearances and biologic behaviour. We think that the amount of myxoid matrix is not sufficiently specified to distinguish both entities. The biological behaviour is not dependent on the amount of fibrous or myxoid tissue (11, 21). Both entities exhibit a slow growth rate and do not metastasise (2, 4, 13, 21). Possibly in older patients more myxoid matrix is found, which could be a sign of tumour regression. In contrast to other authors we suggest that the differentiation of fibromyxoma and myxoma is not useful (13, 17, 18). We propose that both terms should be used synonymously to avoid confusion in the terminology.

In agreement with other authors, we suggest that complete curettage or excision is a sufficient treatment for both fibromyxoma and myxoma (1, 2, 5, 17, 18, 20, 22). Of course, if extensive curettage is necessary as in the current case, a bone graft must be used for reconstruction (1, 5, 10, 17). In fibromyxoma local recurrences have been reported in three of 23 patients (13%) (17). In fibrous dysplasia, recurrences after curettage and reconstruction with autogenous and / or allogeneous bone are more common. In fibrous dysplasia, there are reports describing the resorption of autogenous bone in 36% to 55% of cases. With the use of allogeneic material, bone resorption can be reduced to 18% (8, 15). In the current case, autogenous corticocancellous bone and bovine spongiosa was used to fill the defect without signs of resorption 30 months after surgery.

The current study showed that there are some clues indicating that the differentiation of fibromyxoma and myxoma is not useful because of similar clinical, radiological and biological features. However, the differentiation of fibromyxoma / myxoma from fibrous dysplasia is important, because of differences in these features. It is important to be aware of these tumours as differential diagnoses. The study of further cases is the only possibility to gain further information about these tumours.

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