

Ossifying Fibromyxoid Tumour of Soft Tissues: A Rare Case Report

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Abstract

A case of ossifying fibromyxoid tumour of the soft part, a rare neoplasm arising about the knee of a young man is reported. The tumour has the characteristic morphology, having a thin bony shell, vascular myxohyaline stroma with oval or slightly spindly epitheloid cells arranged in cord pattern. The important histomorphological features have been highlighted and illustrated. The histogenesis, biological behavioral variations and differential diagnosis has been briefly discussed.

Keywords: Ossifying fibromyxoid tumour; Rare soft tissue tumours; Histomorphologic features

Introduction

Ossifying fibromyxoid tumour (OFMT) of soft tissues is a rare neoplasm of uncertain histogenesis, first described in 1989 with 59 cases identified till then [1]. It most commonly presents as a slow growing subcutaneous mass in extremities, and less commonly in head and neck and trunk [2]. Males are affected slightly more than females and the age range is 14-83 years with a median of 50 years [1,2]. The tumour often has a vaguely lobular or nodular architecture with lobules composed of epitheloid, polygonal, round oval or slightly spindled cells arranged in nests, cords or trabeculae with myxohyaline stroma, usually with a shell of metaplastic bone at its periphery [3]. Some cases of OFMT have atypical cytomorphological features and more aggressive behaviour and it has been proposed to classify them as typical, atypical or malignant, based on morphological and behavioral patterns [4-6]. We are tempted to report this case to highlight the essential features of the rare neoplasm. The aim is to create more awareness about this less well recognised neoplasm among surgeons and pathologists who may mistake it for other lesions leading to under-reporting. Correct diagnosis is important as some of these tumours may have a more aggressive behaviour.

Case Report

A 28 year old male presented with a painless, slow growing, firm 3×2×2 cm subcutaneous mass just above the knee. The patient noticed the mass 6 months back and he was avoiding surgery until he noticed that it is growing and producing an easily visible swelling. It was slightly mobile transversely and unattached to the underlying bone. No radiological examination was done in the peripheral hospital where he reported. Other routine investigations were normal. Surgeon's suspicion was fibroma or neurofibroma.

At surgery, the mass appeared to be circumscribed, free from bone but partially attached to the quadriceps tendon. It was totally excised including the area of attachment.

On gross examination the mass was 3×2×1.5 cms, well circumscribed with slightly lobulated external surface. It had a firm and hard consistency with a thin, hard calcified shell of varying thickness covering most of the external surface. The cut surface was firm, white, slightly slimy and cartilaginous with grainy gritty areas. Four blocks were prepared including the external surface and calcified areas.

Microsection showed a distinctly nodular moderately cellular tumour with scattered small foci of microcalcifications. A thin shell of bone was obvious at the periphery with extension of calcification into

the septae dividing the lobule (Figure 1a). Foci of microcalcification were also noted within the tumour (Figure 1b).

The cells in the nodule were round, polygonal, ovoid or slightly spindled. The nuclei were bland vesicular with rather empty looking or scanty fine granular chromatin and occasional fine nucleoli. The cells were mostly in cords or trabeculae. In some areas they were scattered or in small groups with intervening tissue having a hyaline, vaguely cartilaginous appearing stroma (Figure 2a). There was no remarkable atypia, pleomorphism or increased mitosis. Myxoid areas were seen, but were sparse (Figure 2b). Prominent thin walled blood vessels were also noted in the stroma. The cells were diffusely positive for vimentin, S-100 and NSE but negative for desmin, EMA and SMA. These finding were classical as expected in typical OFMT. The patient has been followed up for 7 months and there is no evidence of recurrence.

Discussion

The histogenesis of OFMT is uncertain. Most workers with

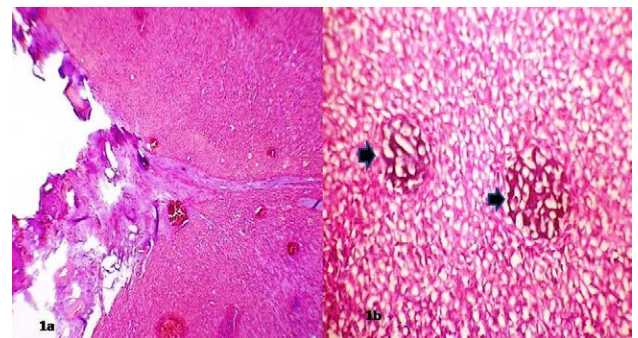


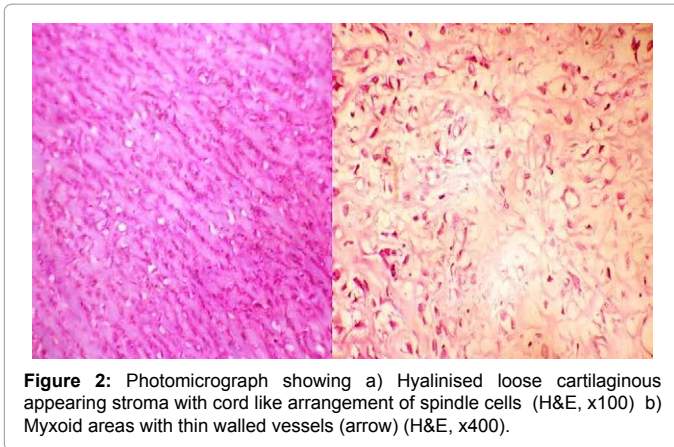
Figure 1: Photomicrograph showing a) Thin bony shell with extension of calcification into the septae (H & E, x100) b) Moderately cellular tumour with scattered foci of microcalcification (H&E, x400).

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Received July 10, 2014; Accepted August 27, 2014; Published September 02, 2014

Citation: Panicker NK, Buch AC, Garg K, Satav VH (2014) Ossifying Fibromyxoid Tumour of Soft Tissues: A Rare Case Report. J Cancer Sci Ther 6: 322-323. doi:10.4172/1948-5956.1000288

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immunohistochemical and ultrastructural evidence suggest schwannian or chondroid differentiation [1,7,8]. Some have also considered myoepithelial differentiation [5,9]. Neuroepithelial differentiation has also been postulated [10]. Immunocytochemically, the tumour cells have a scrambled phenotype, the most consistent positive reactivity has been with vimentin, S-500, NSE and CD57. Here also there is much variability from tumour to tumour [1,11]. Hence there is no consistent immunological marker for this tumour. Our case also showed positive staining for S-100 and vimentin but was negative for desmin. Most workers agree that cytomorphologically and behaviorally intermediate and malignant forms of tumour exist others hold that the small numbers of intermediate and malignant examples identified are probably other tumour types [11]. Our case had innocuous histology with other classical features of OFMT.

Most cases of OFMT are cured by adequate surgical excision. Recurrence has been reported in up to 22% cases and metastasis in 6% [4,10]. Though those tumours with high cellularity, high nuclear grade or increased mitotic activity (more than 2/50 HPF) are expected to behave aggressively, occasional typical benign appearing OFMT may reoccur or produce metastasis [12].

Some tumours have predominant myxoid areas [2,12]. Most others have predominant hyalinised collagenous areas, occasionally with foci of calcification and cartilaginous areas as seen in our case [13,14]. Our case had only small areas of loose myxoid tissue, but the areas of external shell of bone, hyaline cartilaginous areas with cord like pattern and microcalcification were striking.

The tumour need to be distinguished from epitheloid schwannoma, condroid syringoma, myxoid chondrosarcoma and epitheloid leiomyoma. It has many morphological and immunohistochemical similarities to nerve sheath tumours. But the calcifying shell, nodularity and cytological features are helpful distinctions. Chondroid syringoma has distinct epithelial elements and markers. Myxoid chondrosarcoma

is also cytologically different and display intracytoplasmic glycogen not present in OFMT. Only occasional OFMT show muscle antigen unlike smooth muscle tumours which are invariably S-100 negative.

We conclude by stating that OFMT is rare tumour of the soft tissue with sufficiently distinctive clinical and histomorphological features aiding its identification. We are of the impression that the neoplasm is probably not all that rare as a fair number may not be recognized as such and tend to be dumped with conditions having overlapping morphology.

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