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Ossifying fibromyxoid tumor of the trunk mimicking hydatid cyst: A case report

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ABSTRACT

INTRODUCTION: Ossifying fibromyxoid tumor (OFMT) is a rare lesion that generally occurs in the soft tissues of proximal limbs, head or neck and presents as a slowly growing mass. Abdominal or trunk locations are extremely rare.

PRESENTATION OF CASE: We report a case of 50-year-old man who presented with a painless, slow growing epigastric mass for 5 years. Radiologic assessment revealed a well circumscribed median subcutaneous parietal mass lesion present in front of the xiphoid process suspicious of a calcified hydatid cyst. Diagnosis of OFMT was made on histopathological examination of the resected specimen.

DISCUSSION: OFMT most often presents as a single swelling arising from the subcutaneous soft tissues or skeletal muscles of the extremities. Multifocal presentation is exceedingly rare. Radiologically, a peripheral shell of bone is seen in more than 50% cases. On MRI, myxofibrous stroma appears isointense to muscle on T1 and of intermediate to high signal intensity on T2. Surgical excision is the mainstay of treatment. Histologically, the tumor has a thick fibrous capsule with a complete or partial underlying layer of metaplastic woven or lamellar bone. Tumor is composed of uniform round, ovoid, or spindle-shaped cells arranged in nests and cords embedded in a variably myxoid and collagenous Alcian blue-positive stroma. On immunochemistry, the tumor cells are positive for S100 protein and desmin in 90% and 50% cases respectively.

CONCLUSION: OFMT is a rare soft tissue tumor with malignant potential often misdiagnosed as a benign lesion. Complete surgical excision should be performed to prevent local recurrence.

1. Introduction

Ossifying fibromyxoid tumor (OFMT) is a rare soft tissue neoplasm first described by Enzinger et al. in 1989 [1]. This tumor of intermediate malignancy potential and uncertain line of differentiation displays a ubiquitous distribution but occurs frequently at extremities [2]. Clinically, OFMT presents as a slowly growing well-circumscribed mass arising from the subcutaneous tissues or muscles. Used to be considered of unknown etiology, it is currently well established that is a translocation-associated neoplasm without recapitulation of a normal line of differentiation [3]. Although OFMT is rare, it is likely an underdiagnosed entity because of the histological and immunohistochemical overlapping features with several soft tissue neoplasms, the lack of bone in some cases, and the absence of recognition by non–soft tissue pathologists moreover with unusual sites or deep locations [2]. We report a case of 50-year-old man with a painless, slow growing epigastric mass for 5 years which was confirmed to be OFMT postoperatively on histological examination of the resected specimen. This case has been reported in line with the SCARE criteria [4].

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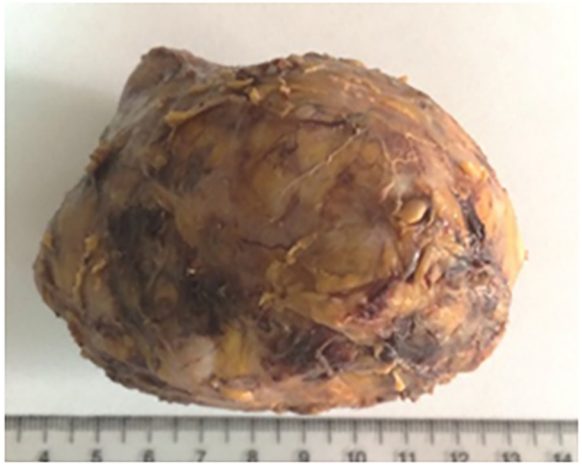

Fig. 1. Gross appearance of well-circumscribed encapsulated mass.

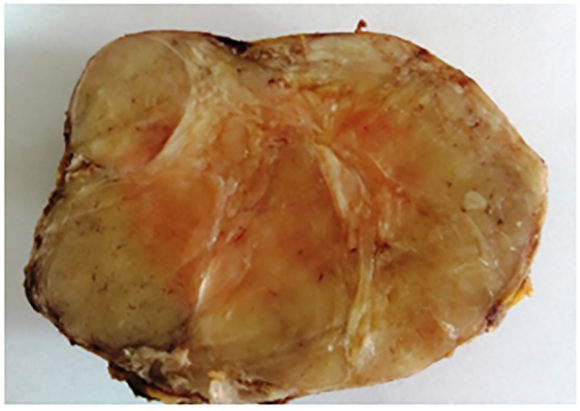

Fig. 2. Cut section showing a coarsely lobular tumour displaying a glistening yellow color with myxoid areas. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

2. Case description

A 50-year-old man, without past medical history presented with an epigastric mass. The mass was painless and had gradually increased to the present size of 4 × 3 cm in the last 5 years. There was no history of fever, night sweats, decreased appetite, or weight loss. On local examination, the mass was located over the xiphoid process in the subcutaneous plane, non-tender, firm in consistency, not fixed to the underlying bone with the normal overlying skin. Abdominal examination was unremarkable and no other subcutaneous masses or lymphadenopathy was noted. Routine blood investigations including hematological and biochemical tests were normal. Tumor markers were within normal range (Carcinoembryonic antigen: 0.9 ng/ml, Carbohydrate antigen 19-9: 2 U/ml). Abdominal ultrasonography (USG) revealed a well circumscribed subcutaneous lesion present in front of the xiphoid process, oval in shape measuring 38 × 34 mm with tissue density and peripheral calcifications (Fig. 1). Based on the clinico-radiological findings, a calcified hydatid cyst was suspected and surgical excision was performed. Post-operative recovery was uneventful. There was no recurrence till the last follow up at 9 months.

Grossly, the tumor appeared pearly white with a lobular contour confined by a thin capsule measuring about 40 × 35 mm in diameter (Figs. 1 and 2). Grittiness of the mass on sectioning suggested calcification. On histology, the tumor was well circumscribed by a thick fibrous capsule. Dense fibrous septa extended from the capsule into the tumor, leading to a lobulated appearance (Fig. 3). Lobules consisted of closely apposed ovoid to epithelioid cells. Some areas were less cellular and lobules are made of short spindle cells surrounded by abundant fibro-myxoid matrix (Fig. 5). Foci of chondroid metaplasia were seen (Fig. 6). The cells were arranged in parallel bundles and focally disposed in perivascular whorls. The nuclei were small, oval shaped, non-pleomorphic and vesicular (Fig. 7). Mitoses were few, less than 1 per 10 high power fields (HPF). Thick-walled, hyalinized vessels were present. There was no focal hemorrhage or

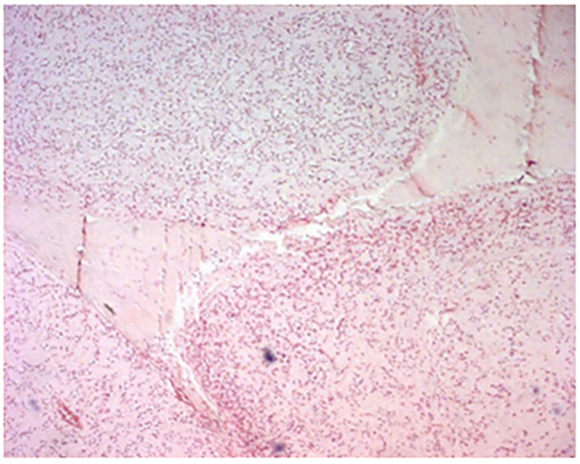

Fig. 3. Fibrous septae giving a lobular appearance (HEx100).

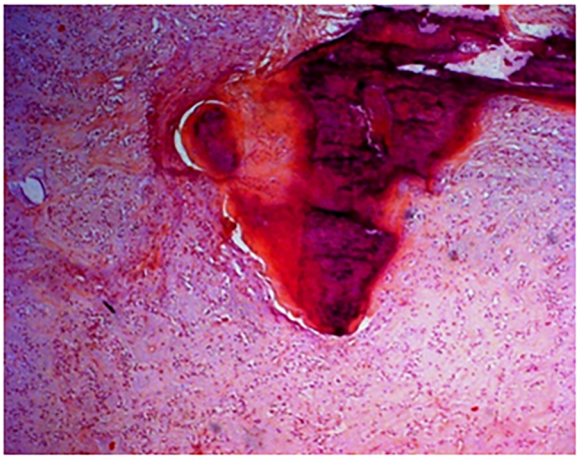

Fig. 4. Fibrous septae that are focally ossified (HEx100).

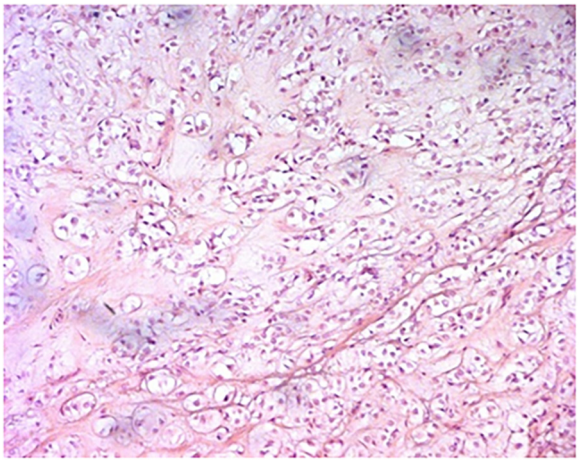

Fig. 5. Abundant myxoid matrix separates nests and cords of tumour cells (HEx200).