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

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ARTICLE INFO

Article history:
Received 7 March 2017
Received in revised form 21 June 2017
Accepted 21 June 2017
Available online 22 July 2017

Keywords:
Ossifying fibromyxoid tumor
Case report

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 suggestive 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 immunohistochemistry, 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.

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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 it 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].
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 $4 \times 3$ 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 $38 \times 34$ 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 $40 \times 35$ 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
necrosis. Spicules of bone with focal osteoblast rimming were found in the central zones of the tumor close to fibrous septa (Fig. 4). No glandular structures were identified. Immunohistochemically, the tumor cells were uniformly positive for S100 (Fig. 8) and focally positive for actin smooth muscle and AE1/AE2. Desmin and caldesmon were negative. The proliferative activity as labeled by Ki-67 was less than 1%. Based on these pathological findings, the final diagnosis of ossifying fibromyxoid tumor was made.

3. Discussion

OFMT of soft parts predominantly occurs in adults with a median age of 50 years (range: 14–83 years) with a slight male predilection [2,3]. It most often arises from the subcutaneous soft tissues or skeletal muscles of the extremities (proximal more common than distal) [5]. Classically, OFMT manifests as a small sized (average, 4–5 cm), well-circumscribed painless slowly growing mass [3]. Although occasional symptoms such as paresthesia and discomfort are reported with deeply located tumors. Multifocal presentation is exceedingly rare [2]. Indolent physical manifestation of this tumor is responsible of a longstanding clinical course, ranging from 1 to 20 years or even more.

Radiologically, OFMT appears as a well–limited nonspecific subcutaneous or deeper soft tissue mass [2]. A peripheral shell of bone is seen on CT in at least 60% to 70% of cases [3]. Furthermore, intra-lesional mineralization and calcification may be present [5]. CT features are nonspecific and intra-lesional mineralization or calcification may lead to confusion with several differential diagnoses such as ossifying hematomas, myositis ossificans and calcified hydatid cyst as our patient. MRI features are variable. Myxofibrous stroma is isointense to muscle on T1 and of intermediate to high signal intensity on T2 in contrast while metastatic bone shows low signal intensity on T1 and T2 [3]. These MRI findings helps to rule out malignant neoplasms.

Grossly, OFMTs are well-circumscribed and mostly surrounded by a fibrous or fibro-osseous pseudo-capsule. Cut surface is lobulated, firm and rubbery with presence of myxoid degeneration, at times. A shell of bone is noted in more than 80% of cases [2,3]. Average size of the tumor is about 4 cm; however, some giant nodules >10 cm have been reported [9].

Histologically, the capsule is made of thick fibrous septa that extend into the tumor leading to a vaguely lobulated multinodular appearance [2,3,5]. Underlying the capsule, a complete or at least partial layer of metaplastic woven or lamellar bone is present in most cases. Tumor is composed of uniform round, ovoid, or spindle-shaped cells with vesicular nuclei, discernable nucleoli and eosinophilic cytoplasm. Tumor cells are arranged in nests and cords embedded in a variably myxoid and collagenous stroma. Some lesions are predominantly myxoid with an abundant Alcian blue-positive stroma. Small foci of calcification and metaplastic cartilage may also be seen in the tumor nodules. Most have a rich vasculature, with many vessels exhibiting perivascular hyalinization [6]. According to Folpe et al., tumors characterized by high grade or high cellularity and mitotic rate greater than 2 mitoses per 50 high-power fields (HPFs) were named malignant OFMTs. Tumors deviating from the features of a typical OFMT with insufficient criteria for malignancy, were categorized as atypical subtypes [7,8].

At immunochemistry, a positivity for S100 protein is observed in approximately 90%, of OFMT. Desmin is positive in about half of cases. There is a weak and focal expression of epithelial markers (cytokeratins and EMA) [5]. The loss of INI1 (SMARCB1) in a mosaic pattern has been reported in 30% to 60% [2]. Rare examples (<10%) express smooth muscle actin, CD10, or glial fibrillary acidic protein (GFAP) [9].

Surgical excision is the main therapeutic option, regardless of tumor size, depth or location. Generally, OFMT has good clinical course. Nevertheless, local recurrence is possible and it is reported in 17% to 27% of patients after complete surgical management, decades after initial excision [2,9]. Diffuse hyper-cellularity, high mitotic rate and high nuclear grade seems to be major risks for local
recurrence and metastasis [5,8]. Metastases have been described to the lungs and soft tissues [8].

4. Conclusion

OFMT is a rare soft tissue tumor presenting as an indolent subcutaneous nodule. Complete surgical excision should be performed due to its malignant potential.

Conflict of interest

The authors declare that they have no conflict of interest.

Funding

This study has not received any funding.

Ethical approval

The study was approved by Ethics Committee of Hospital Sahloul.

Consent

Written informed consent was obtained from the patient, and that you we can provide this if the Editor ask to see it.

Author contribution

Study concept or design – MA, HA, NA.
Data collection – HA, NA, AC.
Data interpretation – MA, HA, RG.
Literature review – HA, RG, NA, AC.
Drafting of the paper – HA, RG, AC.
Editing of the paper – MA, AC.

Guarantor

Ahlem bdioui.
Houssen Anamar.

Acknowledgements

None.

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