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Case Report

Ossifying fibromyxoid tumor of soft tissue: A case report with review of literature

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ABSTRACT

Introduction and importance: The ossifying fibromyxoid tumor of soft tissue is a rare tumor of intermediate differentiation and uncertain lineage that occurs in adults mostly in the extremities and the trunk.
Presentation of case: we present a case of 57 year-old man presenting with a right scapular mass. It was a subcutaneous and painless mass that was largely excised. The diagnosis of ossifying fibromyxoid tumor of the right shoulder was made. The follow up of 1 year was without recurrence and metastasis.
Clinical discussion: The ossifying fibromyxoid tumor of soft tissue is exceptional, microscopic diagnosis and management is challenging, considering the scarcity of the tumor.
Conclusion: More cases and retrospective studies are needed to understand the pathogenesis and to determine optimal treatment regimens.

1. Introduction

The ossifying fibromyxoid tumor of soft tissue (OFT) is a rare mesenchymal neoplasm of intermediate differentiation and uncertain lineage [1]. It was firstly described in 1989 by Enzinger [1–3]. Approximately 300 cases have been reported in the literature until now [1]. It occurs in any soft tissue location, mostly in the extremities and the trunk [2,4].

We report a new case of OFT arising in the right shoulder with a review of literature in order to highlight its clinical, histopathological, and prognostic aspects. We had follow the instruction of 2020 scare guidelines [5].

2. Presentation of case

A 57-year-old man, with no familiar or personal or psychosocial history, presented with a right scapular slow-growing tumor. The physical examination revealed a subcutaneous painless mass measuring 10 cm in diameter. The patient had not drug allergy. He agreed to have an excision of the mass. An orthopedist, with a 15-years experience, underwent the operation without any complications. The patient left the hospital after one week. The macroscopic examination showed an encapsulated, multinodular firm and tan-white tumor containing calcifications. Histologically, the tumor was composed of uniform, round or spindle shaped cells arranged in nests and cords and deposited in a variably myxoid and collagenous stroma with an incomplete ring of lamellar bone at the periphery of the tumor (Fig. 1). Tumor cells had pale nuclei and small amounts of eosinophilic cytoplasm (Fig. 2). Mitoses were absents. On immunohistochemical study, the tumor cells were diffusely positive for S-100 protein (Fig. 3) as well as vimentin and focally positive for desmin. Immunostaining with pancytokeratin, EMA, SMA was negative. The diagnosis of OFT of the right shoulder was made. After 1 year of follow up, the patient is in total remission without recurrence or distant metastasis.

3. Discussion

The ossifying fibromyxoid tumor of the soft tissue (OFT) is extremely rare. It was firstly described in 1989 by Enzinger in a series of 59 cases from the Armed Forces Institute of Pathology (AFIP) [1,2,6]. Its

Abbreviations: OFT, ossifying fibromyxoid tumor; EMA, Epithelial Membrane Antigen; SMA, Smooth Muscle Actin; GFAP, Glial fibrillary acidic protein; EMNST, epithelioid malignant nerve sheath tumors; SEF, sclerosing epithelioid fibrosarcoma.

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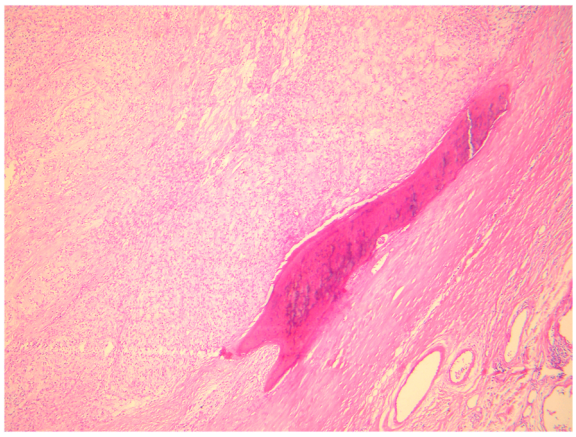

Fig. 1. The tumor is surrounded by a thick capsule and contains an incomplete shell of lamellar bone at the periphery. Tumor cells are arranged in nests and cords in a variably myxoid and collagenous stroma (HE x 40).

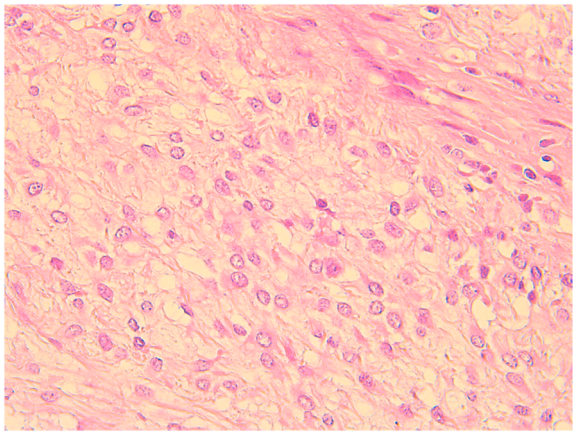

Fig. 2. The tumor is composed of uniform round, or spindle cells with a pale nuclei and a small amounts of eosinophilic cytoplasm (HE x 400).

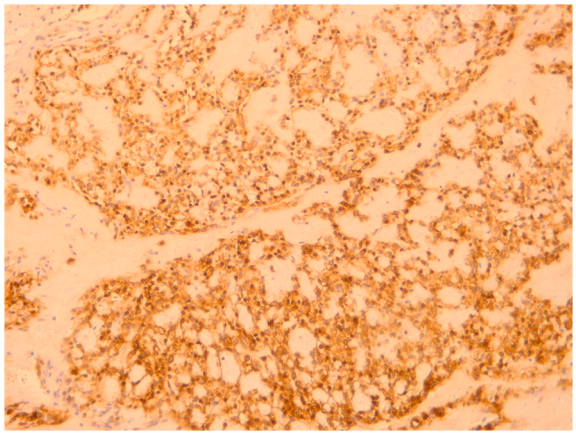

Fig. 3. Diffuse positivity for S100 protein.

pathogenesis is still unclear [7]. Rearrangement of the PHF1, locus at the chromosome 6p21 had recently been observed in 80% of OFT [1,3]. Graham had also reported a deletion of the SMARCB1/INI1, locus at the chromosome 22p12.1 in 71% of cases [1,7,8].

The OFT is a mesenchymal tumor of an uncertain differentiation. Cartilaginous, smooth muscle and nerve sheath origin was suggested but this theory needs to be proven [1,3,6,7].

This tumor arises mostly in adults with male predominance and mean age of 50 years [1,2,6]. It occurs usually in the extremities [1,6]. Less common sites include the trunk, the head and the neck, the oral cavity, the retroperitoneum and the mediastinum [1,3,6,7]. It is an indolent, well-defined, slow growing and firm mass located in the subcutaneous tissue [9]. Location in skeletal muscle or in tendon is rarely seen [9]. Most of tumors range from 3 to 5 cm [9].

The radiographic examination shows a well-defined mass with an in complete ring of peripheral ossification [1,7]. In 60–70% of the case, the Computer Tomography reveals the peripheral "bone shell" [1,7].

The macroscopic finding shows frequently a well-circumscribed, multinodular mass, covered by a thick fibrous pseudocapsule with a tan-white color and a firm to rubbery consistency [1–3,7]. Folpe and Weiss described three subtypes of OFT typical, atypical and malignant. This classification is based on cellularity, nuclear atypia, and mitotic index [1,7,9].

The typical subtype is composed of trabeculae and nest of uniform round to spindle shaped cells surrounded by a fibromyxoid stroma [1,2,6,7,9]. Mitotic rate is usually 1 to 2 per 50 high power-fields [2]. At the periphery of this lesion, incomplete ring of lamellar and metaplastic bone is found in 70% of cases [9]. Satellite nodules have sometimes been observed.

The malignant form is characterized by high mitotic activity (>2 mitosis per 50 high power-fields) and high cellularity [3].

The atypical form is described as a tumor that doesn't fulfill all criteria of malignant or typical subtype [1,4,6].

Immunohistochemically, the tumor cells are positive for S100 protein and vimentin in 60–70% of cases and for desmin in 13% of cases [1,4]. The atypical and malignant forms express less S100 protein [1,4]. Rarely, the tumor can show positivity for CD10, keratin and GFAP [6]. The loss of expression of INI1 has recently been reported with mosaic patterns [1,3].

Cytogenetic analysis using FISH had identified PHF1 arrangement in about 80% of OFT with fusion to EP400 in 44% of cases [1,8] or TFE3 arrangement [8]. In addition, Antonescu et al. had discovered a number of new fusion including ZC3H7B-BCOR, MEAF6-PHF1, EPC-PHF1, CREBBP-BCORL1 and KDM2A-WWTR1 ([10]).

The main histologic differential diagnoses include epitheloid schwannoma, epithelioid malignant nerve sheath tumors, mixed tumor/myoepithelioma of soft tissue, low grade fibromyxoid sarcoma, extraskeletal osteosarcoma and sclerosing epithelioid fibrosarcoma [1,3,6].

The typical subtype of OFT is characterized by a low risk of local recurrence with a rate of 0–12% and no distant metastasis [1,3,4,6]. However, the atypical form has an intermediate malignancy with a local aggressivity and recurrence [1–4,6]. The malignant form is considered as a sarcoma due to its aggressive behavior and its high risk of recurrence and metastasis with a rate of 20–60% [1,11,12]. The common metastatic sites are lung and soft tissue [1,4,6,12]. The local recurrence rate is respectively 0–13% and 0–60% for atypical and malignant OFT [12]. The treatment is based on the wide excision of the tumor [1,7].

4. Conclusion

Ossifying fibromyxoid tumor of soft tissue is a rare mesenchymal tumor of intermediate differentiation and uncertain lineage. Follow up is needed especially for atypical and malignant subtypes.

More cases and retrospective studies are required to understand the pathogenesis of this disease and to determine optimal treatment regimens.

Ethical approval

This study is exempt from ethical approval at our institution.

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Author's contribution

Ahlem Bchir: Drafting, conception, analysis.
Ahlem Bellalah: conception, collecting data.
Nouha Ben Abdeljelil, Manel Njima: acquisition and interpretation of data.
Rym Hadhri, Leila Njim, Abdelfateh Zakhama: revising.

Research Registration number

1. Name of the registry: N/A*
2. Unique identifying number or registration ID:N/A*
3. Hyperlink to your specific registration (must be publicly accessible and will be checked).

Guarantor

Ahlem Bellalah

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Availability of data and material

The authors declare that there are that all data and materials are available.

Declaration of competing interest

The authors have no conflict of interest to disclose.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.102479.

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