Ossifying Fibromyxoid Tumor: A Rare Mesenchymal Neoplasm

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Abstract

The following case study demonstrates a 26 year old male that presented to the dermatology clinic with an enlarging, raised skin nodule located on the superior buttox region. The patient reported it had persisted for two years and had not received prior treatment. He notes a family history of nonmelanoma skin cancer but has had no other dermatological issues in the past. Physical examination revealed a pink, firm and well-circumscribed subcutaneous mass with a prominent follicular pore. It was assumed the lesion was an epidermal inclusion cyst, and surgical excision was performed. Histopathology revealed lobules of epithelioid cells with indistinct cytoplasm in a fibromyxoid hyalinized matrix surrounded by lamellar bone and a collagenous pseudocapsule. Immunohistochemical staining showed moderate immunoreactivity to desmin and mucin but was negative for CD34, S-100, EMA, Actin, and Multicytokeratin. Based on the above findings, a diagnosis of Ossifying Fibromyxoid Tumor was made and given the uncertain biological potential of this lesion, rexcision was performed with negative residual tumor and negative margins on repeat pathological evaluation. The patient was scheduled for close-follow up to survey for recurrence or possible metastasis.

Introduction

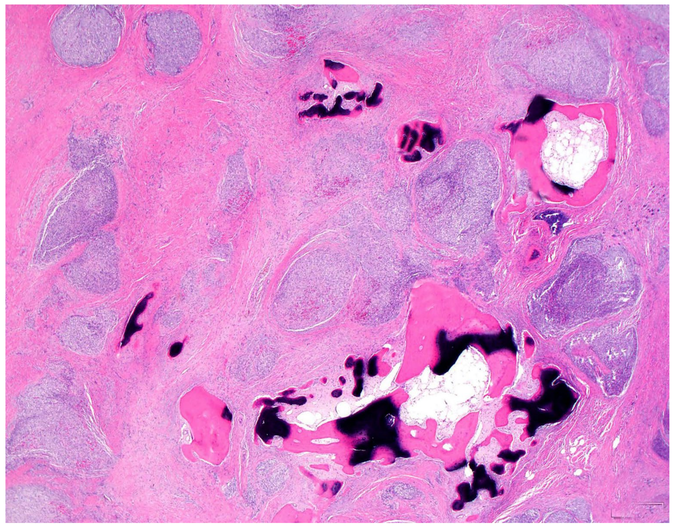
Ossifying Fibromyxoid Tumors (OFMTs) are rare soft tissue neoplasms that develop in the subcutaneous layer of the skin. Histologically, they are characterized by lobulated chords of bland, round cells organized in a fibromyxoid hyaline matrix with a peripheral shell of woven bone. Clinically, an OFMT tends to present as a small, painless mass that persists over years most commonly arising on the extremities. Most OFMTs are benign and can be treated with excision, however local recurrence has been seen in 17% of cases. Furthermore, malignant OFMTs have been identified in 5% of cases and have a metastatic potential of 60%. This case study aims to demonstrate an exemplary presentation, work-up and management of an OMFT in order to shed further light on these rare tumors as well as consider the origin of development in these patients. .

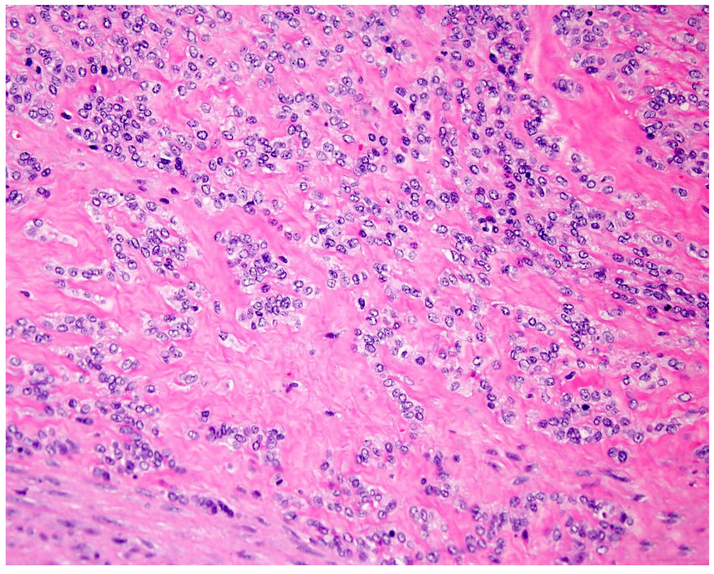
Clinical Presentation

A 26-year-old male presented to our dermatology clinic for a chief complaint of a skin lesion located on his left inferior lateral lower back. He reported that the lesion was enlarging, painful, and was hard to the touch. Nothing had improved or worsened the lesion. He received no treatment for this lesion in the past. Physical examination revealed a 2.3cmx2.0cm painful subcutaneous nodule. Initial assessment of the skin lesions was an epidermal inclusion cyst, and the patient elected to undergo surgical excision. During surgical excision, it was noted that the lesion had a different consistency than an epidermal inclusion cyst which raised concern for an odd neoplasm. The specimen was submitted for dermatopathological evaluation.

Specimen Collection and Pathology Report

The mass was cystic and was composed predominantly of sheets of vague lobules of epithelioid cells, with indistinct cytoplasm. They were surrounded by a "partial shell" of lamellar bone as well as a collagenous pseudocapsule, set within a fibromyxoid to hyalinized matrix. The lesional cells were moderately immunoreactive for Desmin. The diagnosis of an Ossifying Fibromyxoid Tumor was therefore given based on histopathological evaluation. Although the features did not meet criteria for malignancy, there was some uncertainty of biologic potential and thus conservative re-excision to ensure complete removal was advised.





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Discussion

- As seen in the above case scenario, OFMTs are a unique mesenchymal tumor that can mimic other dermatological disorders and go undiagnosed in many patients. One of the most classic staining markers for OFMTs is S-100, which has been identified on average in 75% of cases. Another common marker is Desmin, identified in about 25% of cases. Additionally, some studies note neurofilament staining presence in over 80% of cases. Given the pathology report above for this case, the lesion did not meet malignant criteria in light of cytoarchitecture or immunohistochemical markers. However, typical OFMTs do have metastatic potential of which has been found in 4% of cases. Additionally, these lesions can be persistent and bothersome for many patients. As was performed in this case, local excision with negative margins is the typical treatment for OFMTs. Non-malignant OFMTs have a recurrence rate of about 8%, and thus close post-surgical surveillance is necessary so that new lesions can be identified and treated.

Conclusion

OFMTs represent a group of mesenchymal neoplasms of unknown origin with certain features used to aid in identification. However, classic immunohistochemical markers such as S-100, desmin and others employed may not always be expressed. Furthermore, other tumors share immunological and cytoarchitectural features with OFMTs such as myoepithelial neoplasms, epithelioid schwannomas, and fibromyxoid sarcomas to name a few. Thus, strong clinical suspicion must be utilized in combination with the diagnostic tools available. Although rare and usually benign, certain OFMTs can possess histologically malignant features and have the potential to metastasize and be life threatening. Close clinical surveillance is crucial in these patients not only to identify and treat recurrences, but to also monitor for signs of metastatic lesions.

References

1. Folpe AL, Weiss SW. "Ossifying fibromyxoid tumor of soft parts: a clinicopathologic study of 70 cases with emphasis on atypical and malignant variants." Am J Surg Pathol. 2003 Apr;27(4):421-31.

2. Kempson, R. Ossifying fibromyxoid tumor. Stanford School of Medicine. 2005. http://surgpathcriteria.stanford.edu/softmisc/ossifying_fibromyxoid_tumor/printable.html.

3.Gullett A. Ossifying fibromyxoid tumor. PathologyOutlines.com website. http://www.pathologyoutlines.com/topic/softtissueossifyingfibromyxoid.html.

4.Cody S. Carter and Rajiv M. Patel (*2019*) Ossifying Fibromyxoid Tumor: A Review With Emphasis on Recent Molecular Advances and Differential Diagnosis. Archives of Pathology & Laboratory Medicine: December 2019, Vol. 143, No. 12, pp. 1504-1512. https://www.archivesofpathology.org/doi/full/10.5858/arpa.2019-0371-RA

5.Fletcher, C. Pathology and Genetics of Tumors of Soft Tissue and Bone. Lyon: IARC Press, 2002.

6. Sharma, K et al. "Ossifying fibromyxoid tumor (OFMT) - A rare cause of a painful thumb." *International journal of surgery case reports* vol. 7C (2015): 93-5. doi:10.1016/j.ijscr.2014.10.099