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RESEARCH ARTICLE

OSTEOCLASTIC GIANT CELL RICH UNDIFFERENTIATED PLEOMORPHIC SARCOMA OF THE VULVA- A RARE CASE REPORT

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ABSTRACT

Background: Undiffrentiated pleomorphic sarcoma of vulva with osteoclastic giant cells is a diagnosis of exclusion. It is an exceptionally uncommon diagnostic finding which was eventually diagnosed in our institute and this required a methodical clinical approach and extensive immunohistochemical markers to rule out other probable diagnostic possibilities at this site. The quick diagnostic approach at our institute was ensued by operation.

Case Report: 53 years old female presented with vulval growth to our institute. Incision biopsy was performed as a routine procedure. Biopsy showed atypical spindle cells with few cells showing rhabdoid morphology and osteoclast like giant cells were also present. Histological findings with aid of immunohistochemistry led to the diagnosis of undifferentiated pleomorphic sarcoma with osteoclast like giant cells. The tumor cells stained intensely for vimentin (a mesenchymal marker), but no immunoreactivity for the Cytokeratin Pan (epithelial cells marker), Leukocyte Common Antigen (lymphoid cells marker), SMA (smooth muscle actin), Desmin (smooth and striated muscular cells marker), S100 (neuronal cells marker), CD34 (to identify vascular tumor), CD99 (synovial sarcoma marker) was detected.

Conclusion: Undifferentiated pleomorphic sarcoma of the vulva is an uncommon mesenchymal neoplasm that presents diagnostic challenges. Therefore, as with all rare tumors, undifferentiated pleomorphic sarcoma should be managed in reference centers to determine whether treatment of choice is surgical excision or some form of adjuvant therapy can have beneficial effects.

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INTRODUCTION

Vulvar sarcomas are histologically heterogeneous non-epithelial malignancies that arise from the connective tissue within the vulva. Undifferentiated pleomorphic sarcoma of vulva with osteoclastic giant cells is a diagnosis of exclusion. It is an exceptionally uncommon diagnostic finding.

Objective

The prevalence, risk factors and clinical course of vulval sarcomas are less well characterized than vulval tumors arising from epithelial tissue. Herein, we report a case of primary undifferentiated pleomorphic sarcoma in a 53-year-old woman; this case highlights a rare and interesting variant of primary vulval sarcoma and the diagnostic difficulty that surgeons and pathologists may encounter.

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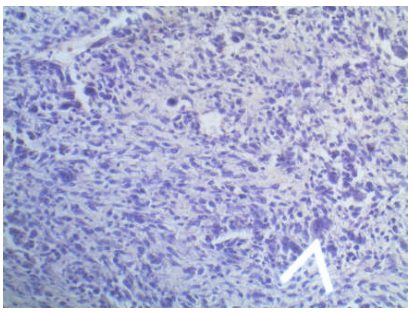
CASE REPORT

53 years old female presented with vulval growth to our institute. Macroscopic examination of the biopsy received elsewhere was described as 15 dark brown soft and friable tissue bits (approx. 3 ml in volume). Incision biopsy was performed as a routine procedure. Biopsy showed atypical spindle cells with few cells showing rhabdoid morphology and osteoclast like giant cells were also present amidst inflammatory cells. Necrotic areas and abnormal mitosis were identified.

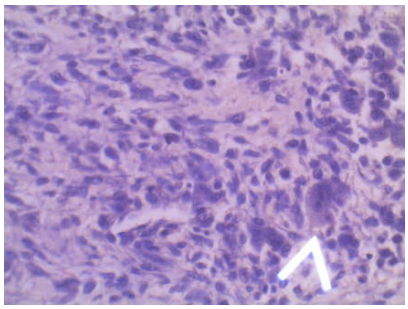
RESULTS

Histological findings with aid of immunohistochemistry led to the diagnosis of undifferentiated pleomorphic sarcoma with osteoclast –like giant cells. The tumor cells stained intensely for vimentin (a mesenchymal marker), but no immunoreactivity for the Cytokeratin Pan (epithelial cells marker), Leukocyte Common Antigen (lymphoid cells marker), SMA (smooth

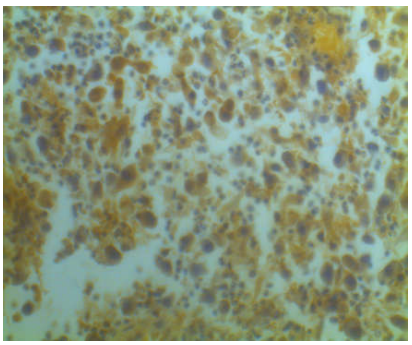
muscle actin), Desmin (smooth and striated muscular cells marker), S100 (neuronal cells marker), CD34 (to identify vascular tumor) and CD99 (synovial sarcoma marker) was detected.



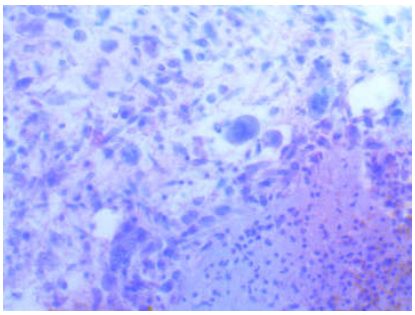
10x view showing osteoclast like giant cells



40x view showing osteoclast like giant cells



Tumor cells showing vimentin positivity



Necrotic Areas with abnormal mitosis

DISCUSSION

After proper histological and immunohistochemical work-up, we arrived at final diagnosis of undiffrentiated pleomorphic sarcoma with osteoclast-like giant cells. IHC positivity with vimentin further confirmed the diagnosis. Sarcomas of the vulva are infrequent neoplasms representing 1–3% of vulvar malignancies. Differential diagnosis of the sarcomas includes leiomyosarcoma, neurofibrosarcoma, reticulosarcoma, MFH, pleomorphic sarcoma, angiosarcoma, liposarcoma and lymhoma. Characteristically undifferentiated pleomorphic sarcoma localized at the genital tract is locally invasive and involvement of the underlying fascia increases the risk of local or distant metastasis. The primary therapy is wide local excision or radical vulvectomy. Metastasis of MFH correlates best with the depth of invasion of the original tumor

Conclusion

Undifferentiated pleomorphic sarcoma of the vulva is an uncommon mesenchymal neoplasm that presents diagnostic challenges. Therefore, as with all rare tumors, undifferentiated pleomorphic sarcoma should be managed in reference centers to determine whether treatment of choice is surgical excision or some form of adjuvant therapy can have beneficial effects.

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