CASE REPORT

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Sister Mary Joseph Nodule in an Ovary Adenocarcinoma

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ABSTRACT

Introduction: Sister Mary Joseph Nodule (SMJN) is a metastatic umbilical lesion secondary to a primary malignancy of any viscera, stomach and colon being most common in men, and ovary in women. **Case report:** In this article, we present the case of SMJN in a 54-year old female patient. An urgent diagnostic workup was performed with a computerized tomography of abdominal cavity and pelvis showing an expansive tumorous formation covering uterus with a carcinomatosis of peritoneum. After biopsy, immunohistochemical profile suggested adenocarcinoma of the ovarian origin. The patient was then referred to the Oncology Consilier of Gynecology Department and further continued followed by the Oncology team.

Keywords: Sister Mary Joseph Nodule, Ovary Adenocarcinoma, Cutaneous metastasis.

1. INTRODUCTION

Sister Mary Joseph Nodule (SMJN) is a metastatic umbilical lesion secondary to a primary malignancy of any viscera, stomach and colon being most common in men, and ovary in women (1). However, cutaneous metastasis from ovarian carcinoma is relatively uncommon in clinical practice (2). Here we report a case of SMJN in a 54-year old female patient presented at Dermatology Department, Clinical Center of The University of Sarajevo.

2. CASE REPORT

A 54-year old female patient with an 8-week history of several umbilical nodules resistant to local treatment was referred to Dermatology Department with a diagnosis of SMJN and suspected underlying malignancy. On examination, she had evident abdominal ascites with a presence of nodules in the paraumbilical area on both sides of umbilicus (Figures 1, 2). An urgent diagnostic workup was performed with a computerized tomography of abdominal cavity and pelvis showing an expansive tumorous formation covering uterus with a carcinomatosis of peritoneum (Figures 3, 4). Blood test demonstrated raised tumor markers for adenocarcinoma. Finally, the histopathological result of skin biopsy revealed metastatic deposits of tumor cells intradermally (Figures 5, 6). Neoplastic epithelial cells with pleomorphic nuclei were arranged in papillary structures. Psammoma bodies and microcalcifications were also found (Figure 7A). Immunohistochemistry was performed on formalin-fixed, paraffin-embedded tumor tissue using an immunoperoxide method. Neoplastic cells showed intense diffuse cytoplasmic staining for cytokeratin (CK) 7 (Figure 7B) and CA-125 (not shown), nuclear positivity for Wilm's tumor 1 (WT-1) protein (Figure 7C). Tumor cells were negative for h-Caldesmon (Figure 7D), as well as for CK20, CDX2 and Podoplanin. Morphology and immunohistochemical profile suggested adenocarcinoma of the ovarian origin. These results were indicative of Sister Mary Joseph's nodule. The patient was then referred to the Oncology Consilier of Gynecology Department and further continued followed by the Oncology team. Seven weeks after her initial admission to Dermatology Department she starts with a chemotherapy treatment prior to possible surgical excision of the tumor. Meanwhile, the skin metastases have grown larger and spread with multiple smaller, ulcerated nodules over the abdominal area. However, the patient still remains in relatively stable condition.

3. DISCUSSION AND CONCLUSION

The classic presentation with a Sister Mary Joseph Nodule (SMJN) at the umbilicus from intra-abdominal extension via lymphatics is a well known example of skin metas-

Figure 1. Red nodules in the paraumbilical area with several smaller nodules on both sides of umbilicus

Figure 2. Abdomen distended and tense. Percussion reveals ascites with shifting dullness

Figure 3. CT of abdominal cavity and pelvis showing an expansive timorous formation covering uterus with a carcinomatosis of peritoneum (saggital view)

Figure 4. CT of abdominal cavity and pelvis with signs of tumor propagation to the front (transversal view)

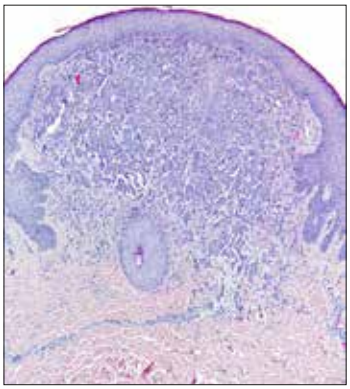

Figure 5. Histopathology showing irregular dermal nests of neoplastic cells underneath intact epidermis (H&E, 40x)

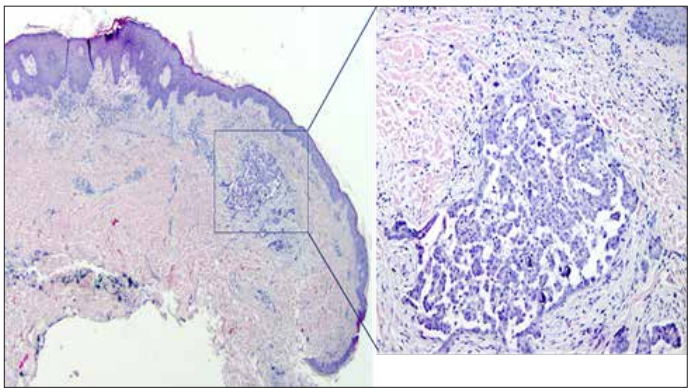

Figure 6. A dermal tumor composed of neoplastic cells forming papillary structures. Psammoma bodies and microcalcification are present (H&E x20, Inset x100)

tases (3). The skin lesion was named after sister Mary Joseph, a surgical assistant to Dr. William J. Mayo, who first noted the association of an umbilical nodule and an intraabdominal malignancy (4, 5). Skin metastasis in form of SMJN from ovarian carcinoma typically occurs in advanced disease with widespread peritoneal involvement, and indicates a poor prognosis (6). The incidence of SMJN is very low, being diagnosed in only 1%-3% of all intra-abdominal or pelvic malignancies (1). In about 75% cases of SMJN with an abdominal malignancy the

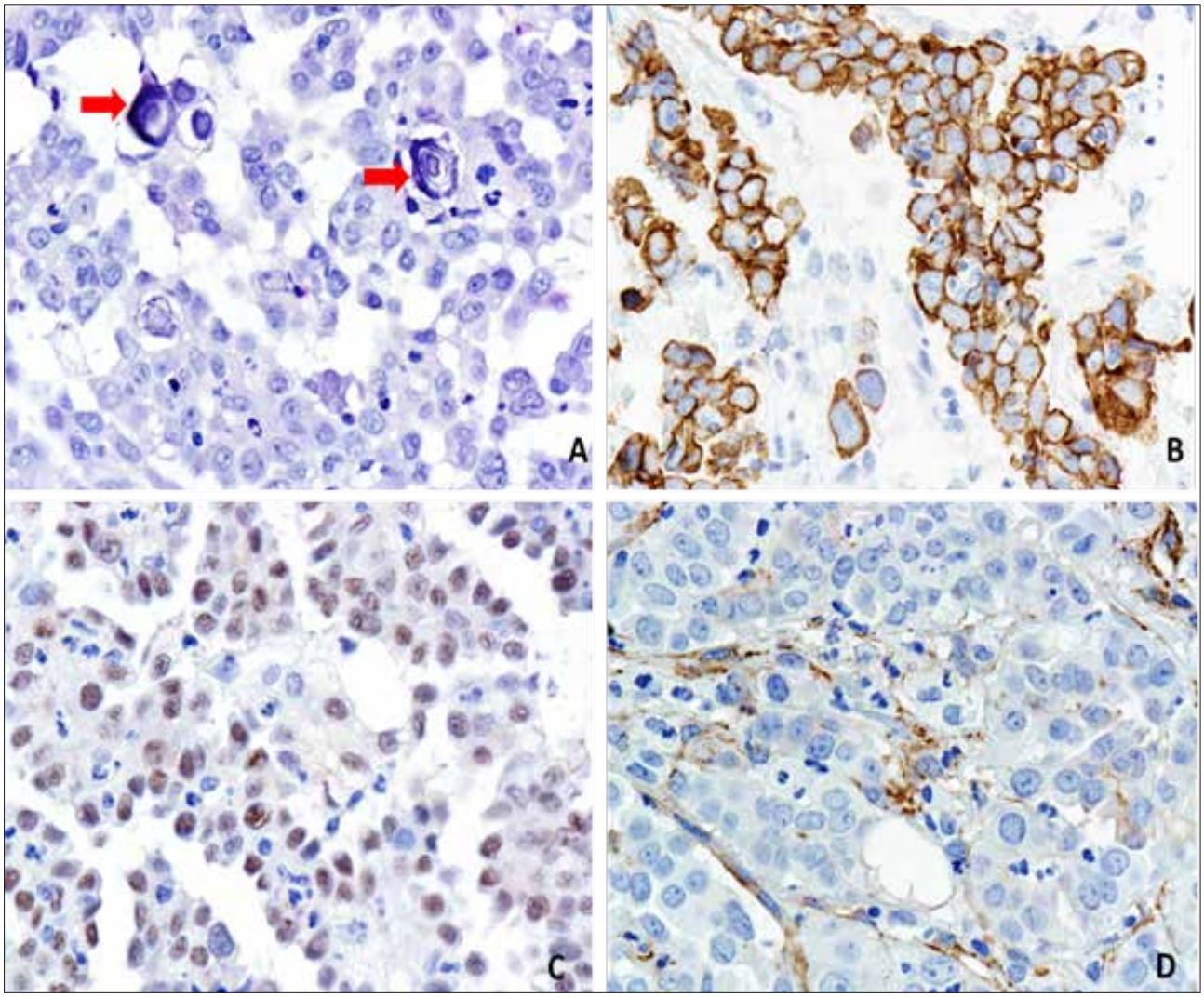

Figure 7. Original magnification x 400 of adenocarcinoma found from umbilical nodule. A) Hematoxylin-eosin stain shows adenocarcinoma cell infiltration. Psammoma bodies are shown with red arrows. B) Tumor cells showing positive cytoplasmic staining for cytokeratin (CK) 7. C) WT-1 immunoreactivity is nuclear. D) Tumor cells are negative for h-Caldesmon

Figure 8. Seven weeks after diagnosing SMJN, the metastases have grown larger and spread with multiple smaller nodules

histological type is adenocarcinoma and in over 55% of cases the origin is from the digestive tract. Gynecological origin is the second most common etiology, with ovarian cancer being the most common, 34% cases (7, 8). Several mechanisms for pathogenesis of cutaneous metastases have been proposed; direct spread from the underlying growth, accidental implantation following surgery, and contiguous spread of tumor cells through the lymphatics. The most common mode of spread of ovarian carcinoma to the skin is retrograde, from involved proximal lymph nodes (9, 10). Metastases from the ovary and the uterus are seen in the skin of the lower abdomen, the groin, or the upper thigh (11). Presentation is typically in the form of rapidly growing, mobile, round or oval nodules of firm or elastic consistency and they may be ulcerated (12). The lesions have also been reported to be variously colored: white, bluish violet, brownish red and so on, with or without pruritus. In diagnosing, a biopsy of the umbilical nodule provides a convenient way of obtaining tissue sample for histological confirmation of the disease (13). Imaging with CT and /or MRI scan will establish the extent of malignancy (14). The therapeutic approach to SMJN is based on appropriate management of the primary tumor, if it has been identified (15).

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