

High-grade Serous Umbilical Adenocarcinoma. About a Case

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Abstract

Primary malignant umbilical tumors are very rare, representing 20%, within umbilical pathologies, and only 0.1% in general frequency. We present a case of a female patient with a history of Primary Umbilical Adenocarcinoma in 2017, untreated and for 2019 extensive umbilical tumor with incisional biopsy that reports: Serous Large Duct Adenocarcinoma. Immunohistochemistry: Adenocarcinoma derived from remnant of the omphalomesenteric duct or yolk duct. She receives preoperative chemotherapy and in May 2020, a Wide Resection is performed, plus reconstruction with Polypropylene mesh. As findings, a 20x20 cm lesion is evident, which involves all the layers of the skin, subcutaneous cell tissue up to the peritoneum, with the omentum involved in the surgical piece. Additionally, cavity-like liver MT lesions are evident. Biopsy reports High-grade Serous Primary Umbilical Adenocarcinoma.

Keywords: Umbilical Adenocarcinoma; Primary; Metastatic

Introduction

The term “Sister Mary Joseph nodule” was first introduced by Hamilton Bailey in 1949 in recognition of the nurse at St. Mary’s Hospital in Rochester, Minnesota, who predicted the presence of intra-abdominal neoplasia by palpating a firm nodule in the umbilical region [1]. Umbilical metastasis is a rare finding in the vast majority of intra-abdominal cancer cases. Primary malignant umbilical tumors are very rare, constituting only 20% of cases and the spectrum includes melanoma, basal cell carcinoma and adenocarcinoma, while metastatic umbilical carcinomas constitute 83% and, in most cases, have their primary in the gastrointestinal tract. Due to the infrequency of the case, we present a patient with a High Grade Primary Umbilical Adenocarcinoma.

Case Report

This is a 47-year-old female patient who is referred to the Oncological Surgery consultation by the General Surgery service in May 2019, in the late postoperative period of Open Cholecystectomy, presenting an increase in volume in the mesogastrium and right flank, painful, interpreted as a lipoma, which is why said service decided to perform its excision under local anesthesia and whose

biopsy B19-48.955 (05/12/19) reports: A 9 cm portion of tissue compatible with Large Duct Serous Adenocarcinoma.

Important Background: Maternal Grandmother Lung Ca (deceased), Maternal Aunt Breast Ca (deceased), Surgical: September 2017 Endometriosis in Abdominal Wall with Biopsy 17B001949 that reports: Soft tissue lesion measuring 10x7x6cms Metastatic Neoplastic Infiltration by Well-differentiated Adenocarcinoma and IHC17-22783: Primary metastatic adenocarcinoma of the digestive tract or pancreatic biliary area, if no primary lesion is located in these sites, the possibility of Primary Adenocarcinoma of the Omphalomesenteric Duct.

April 2019 Open Cholecystectomy (missing biopsy, according to the surgeon of the case, it did not appear malignant, only bent), Lipoma May 2019. Denies Smoking and Alcoholic Habit. Denies digestive problems. Occupation: Household Trades. Gynecological: Endometriosis, treated. I Gesta I Caesarean section. Menarche 12 years, Menopause 46 years.

Extension Studies 2017: MRI of Abdomen and Pelvis without lesions, Bone Scintigraphy without evidence of malignant infiltrative

Immunohistochemical study	Result
Broad spectrum cytokeratin	3+
Epithelial membrane antigen (EMA)	3+
Carcinoembryonary antigen (CEA)	3+
Antigen hep-par 1	Negative
Alpha-fetoprotein (AFP)	Negative
Cytokeratin 20 (CK-20)	Negative
Cytokeratin 7 (CK-7)	3+
Antigen ca 19-9	3+
Thyroid transcription factor-1 (TTF-1)	Negative
Estrogen receptors (ER)	Negative
Progesterone receptors (PR)	Negative
Antigen Ca125	+/-

Table 1: Immunohistochemical panel 17-22783.

disease. Despite the malignancy result in Surgery in 2017, she did not receive any type of treatment, since her treating doctor did not consider it necessary (Reported verbally by the patient).

On physical examination (May 2019): Patient in stable clinical conditions, afebrile, hydrated, tolerated orally. Surgical wound scar on the right flank of the pararectal type with signs of phlogosis of approximately 10 x 10 cm, and a wound with purulent, fetid discharge, fistulized to the skin, with retraction of the same (Image 1). Rest within normal limits.

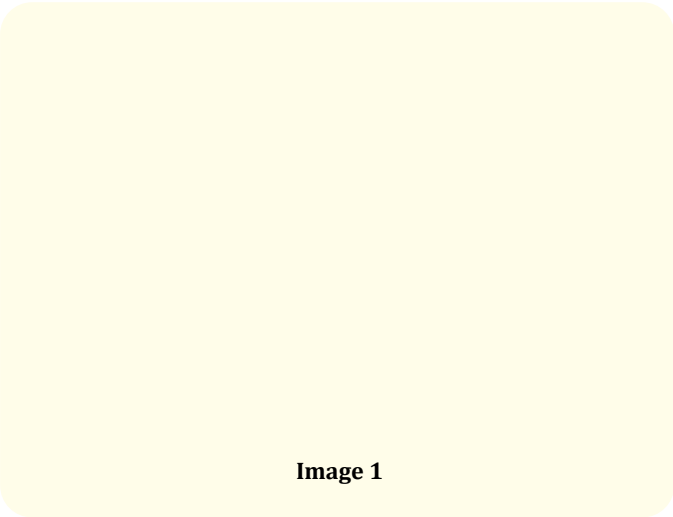


Image 1

Immunohistochemistry study, Abdomen and Pelvis Magnetic Resonance, chest X-ray are requested, and oral and intravenous antibiotic therapy is indicated to decide on the conduct.

05/28/2019 IHC19-24109: Adenocarcinoma derived from remnant of the omphalomesenteric duct or yolk duct.

06/04/2019 Contrast Abdomen MRI: Presence of marked thickening of the abdominal wall at the level of the right flank, with changes in the signal intensity of the muscular planes and of the

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Cytokeratin 7 (CK-7)	3+
Antigen CA 19-9	3+
Thyroid transcription factor-1 (TTF-1)	Negative
Estrogen receptors (ER)	Negative
Progesterone receptors (PR)	Negative
Antigen ca125	Negative

Table 2: Immunohistochemistry panel 19-24109.

subcutaneous cellular tissue measuring 11x8x3.2cms with areas of enhancement later of the administration of intravenous contrast. Normal size liver, spleen and pancreas.

In view of these results, a surgical procedure was decided as soon as possible Wide Resection plus Reconstruction of the Abdominal Wall with Tensor Fascia Lata Vs Polypropylene Mesh.

It is taken to the surgical table in June 2019 but is omitted by the Anesthesiology Service due to high blood pressure figures. It is reassessed by Cardiology without contraindications, but for reasons of inoperativeness of the operating room, the Intervention cannot be rescheduled.

He came back for consultation in July 2019, showing a tumor lesion that compromised the entire flank and right iliac fossa, mesogastrium and hypogastrium, extending to the left flank, approximately 30 x 30 cms, hard, very painful, with persistence of a fistula in a previous scar with a thick fetid serohematic secretion.

In view of the increase in the size of the lesion, a complex reconstruction of the abdominal wall with the possibility of handling it with an open abdomen and without conditions in the hospital for surgery due to inoperative operation of the operating room, it was decided to refer to the Medical Oncology Service, in order to of a preoperative treatment.

In October 2019, Medical Oncology, assesses her and indicates a Gemcitabine plus Paclitaxel scheme, 6 cycles every 21 days. It completes 4 cycles, starting on 12/02/2019 and the last one ending on 03/07/2020.

It is countered by Medical Oncology (April 2020), finding an ulcerated lesion on the skin, with abundant fetid, purulent discharge, which occupies the mesogastrium with the umbilical region, right hypochondrium, flank and right iliac fossa, approximately 15x15cms, with evident reduction in the size of the same towards the left hemiabdomen, but with the novelty of the ulceration in the skin (Image 2).

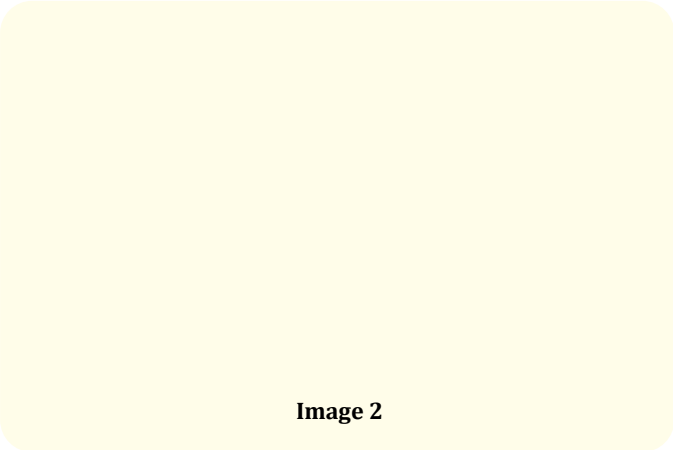


Image 2

In view of these findings, imaging studies (Abdomen and Pelvis CT 20/04/20) (Image 3) are requested and a surgical procedure is proposed Wide Resection plus Reconstruction with Polypropylene Mesh as palliative with the intention of reducing tumor burden.



Image 3

The surgery was performed on 05/21/2020 with room in the ICU and central line catheter, whose findings were: Ulcerated lesion of approximately 20x20cms that compromised the entire abdominal wall of the right hemiabdomen (Image 4), omentum with isolated nodule-like lesions which were resected for biopsy, ascitic fluid, intestinal loop integrity and colon, stomach and pancreas were not evidenced.

An extensive solid liver lesion is palpable in segments II, III, IV and V, hard, highly suggestive of TM. Spleen enlarged. Rest of the cavity without injuries. Solitary nodule on the skin of the hypogastrium, hard, which was resected. Cavity washing, fixation of greater omentum to iliac crests with 3-0 vycril, placement of polypropylene mesh to correct image defect [5], placement of portovac ¼ drain and synthesis by (Image 6). She presented a favorable post-

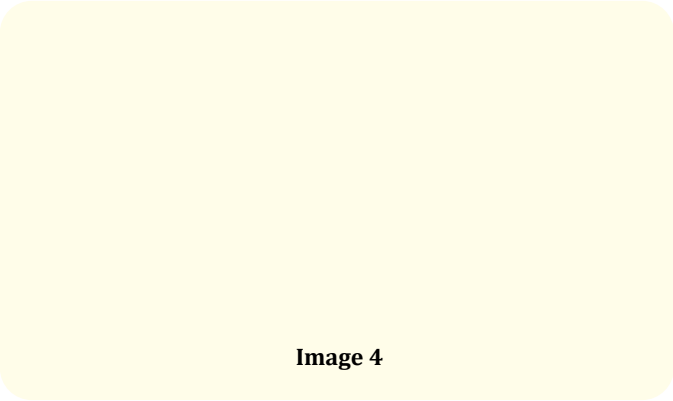


Image 4

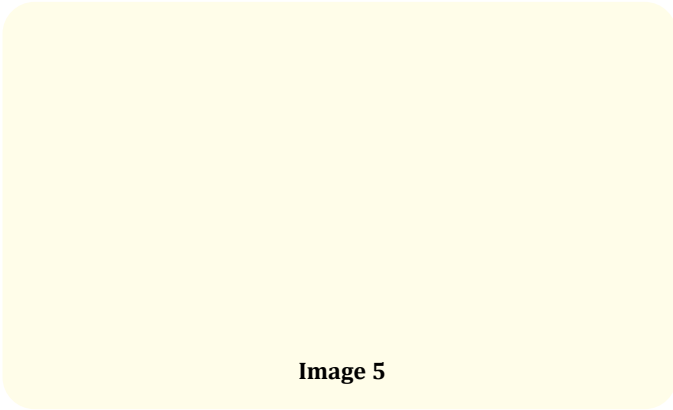


Image 5

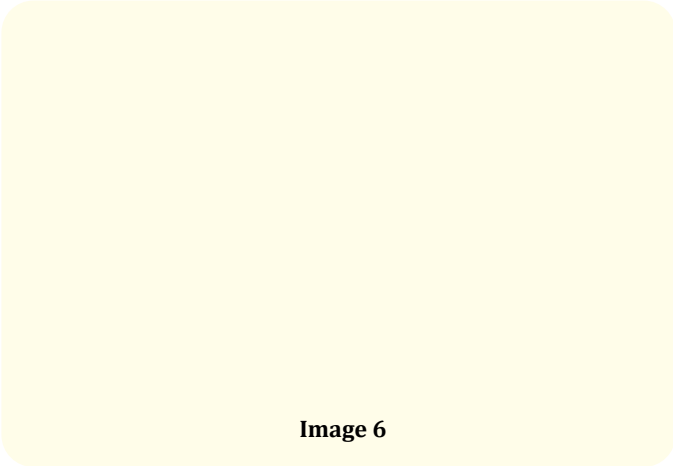


Image 6

operative evolution, without complications and was discharged 24 hours later due to the Covid19 Pandemic.

The result of a 20-30-TV Biopsy is received, which reports (Image 7): Container 1 with tissue formation measuring 16 x 13 x 8 cms, with an adhered omentum fragment of 10x6cms and an irregular tumor lesion of 14 x 13 x 8 cms, compatible with High Grade Primary Serous Umbilical Adenocarcinoma, well differentiated, widely infiltrating subcutaneous and striated muscle cell tissue. Vascular, blood, lymphatic and perineural tumor embolization. Deep surgical resection margins with evidence of neoplastic infiltration.

Package 2 tissue formation measuring 6 x 4 x 1cms epiploic tissue with mesothelial reaction and areas of recent hemorrhage with no evidence of neoplastic cells.

Image 7

Package 3 Skin Losange measuring 1x0.6 cm identified as a nodule in the hypogastric region: Moderate to poorly differentiated High Grade Adenocarcinoma, infiltrating, incompletely resected in extension and depth, which seems to correspond to local TM.

She is referred to the Medical Oncology service and receives systemic treatment, which ends in December 2020. Currently a patient in disease-free control.

Discussion

Malignant tumors in the umbilical region represent more than 10% of neoplasms that affect the skin of the anterior abdominal wall. This anatomical region harbors numerous vascular and embryological connections with the abdominal organs, which favors the appearance of metastases derived from different visceral tumors [2]. However, primary umbilical tumors account for only 20% of malignant tumors in that location, and very few cases have been reported to date.

Most adenocarcinomas of the umbilicus are secondary and are called Sister Mary Joseph’s Nodule. Primary umbilical adenocarcinoma may arise from a pre-existing endometrioma, the coelomic mesothelium, or an embryologic remnant of the umbilicus. The two umbilical embryological remnants are the yolk-intestinal tract (omphalomesenteric) and urachus [3].

Secondary navel metastases in men can originate in the stomach, colon and rectum, pancreas, lung, or prostate. In rare cases, Meckel’s diverticulum adenocarcinoma can spread to the umbilicus. In women, secondary metastasis can originate in the ovary, endometrium, colon or rectum, stomach, pancreas, cervix, or breast [4]. Other possibilities are primary serous carcinoma of the peritoneum with spread to the umbilicus.

In our patient, the absence of any other tumor, which could be the primary source, and the histological findings have indicated that the lesion is a primary serous adenocarcinoma. The presence of preexisting endometrioma or gynecological origin of the tumor has been excluded, mainly given by immunohistochemical studies. The patient was postmenopausal without gynecological symptoms at the time of the current disease.

Umbilical tumors can present as a nodule of varying size, which can be painful and sometimes ulcerated. There may also be an abscess underlying the tumor. The most frequent histological type are mainly adenocarcinomas; although other histological types have also been described, such as sarcomas, melanomas, basal cell carcinomas and less common basal cell carcinomas [5,6].

The glandular epithelium is not normally present in the umbilical area, but it can appear derived from the metaplasia of the squamous epithelium or from the glandular embryological remains derived from the omphalomesenteric duct and the urachus [7].

The mechanism of metastasis from the primary tumor to the umbilicus is unclear; However, several hypotheses have been proposed. The first is that the spread is by peritoneal infiltration (the most common route) or by arteries, veins, or lymphatic channels. Another is that dissemination occurs through embryonic structures such as the urachus, the round ligament of the liver, the remnant of the intestinal yolk duct, or the obliterated vitelline artery [8-15].

There is a connection between the liver and the navel, through the venous and lymphatic channels and these channels seem to serve as bridges for this [9]. Understanding the aforementioned, the common association between hepatic and umbilical metastases could suggest the hypothesis that the tumor spreads from the primary (navel) to the liver, through the portal system and then through the lymphatic and/or venous channels, is extend to the navel.

However, they are all hypotheses and it is not yet clear whether the spread of the umbilical tumor precedes the spread of the liver or vice versa.

Therefore, an umbilical tumor with histological features of adenocarcinoma, the main problem remains to determine the origin. Immunohistochemistry can guide the diagnosis, but it is essential to complete a series of tests to rule out the existence of an extra-umbilical primary tumor (thyroid ultrasound, abdominal ultrasound, abdominal computed tomography, colonoscopy, chest X-ray, etc.) [10].

The treatment of primary tumors of the umbilicus is surgical conduct with radical criteria. During surgery, all abdominal organs should be examined to rule out the existence of a primary tumor that had not been previously detected [11,12].

In addition to the complete resection of the tumor; It is necessary to monitor these patients, since a relapse in the umbilical area has been reported in the literature, as well as the subsequent appearance of liver metastases and lymphadenopathy in the inguinal region [13,14]. This could indicate the need for adjuvant treatment with chemotherapy or radiotherapy in certain patients [15].

Due to its poor prognosis, the treatment of both primary and metastatic umbilical tumors is controversial. The use of preoperative chemotherapy is not ruled out but it will never replace radical surgery. Radiation therapy is described as the treatment of choice in many cases.

Conclusions

Primary umbilical tumors are very rare and there are few cases reported in the literature.

Primary umbilical adenocarcinoma may arise from a pre-existing endometrioma, the coelomic mesothelium, or an embryologic remnant of the umbilicus.

The mechanism of metastasis from the primary tumor to the umbilicus is unclear. Several hypotheses have been proposed, either that the spread is by peritoneal infiltration by arteries, veins or lymphatic channels or that the spread occurs through embryonic structures such as the urachus, the round ligament of the liver, the remnant of the intestinal yolk duct or the Obliterated yolk artery.

If there is an association between the primary umbilical tumor and liver metastases as occurred in our patient. The tumor spreads from the primary to the liver, through the portal system and then through the lymphatic and/or venous channels.

The key treatment is radical surgery. Adjuvant chemotherapy and in some cases radiotherapy is indicated.

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