Case report

Breast metastasis of ilial carcinoid tumor: Case report and literature review
Chakshu Gupta2, Ashok K Malani*1 and Sandeep Rangineni1

Address: 1Department of Oncology, Heartland Regional Medical Center, St. Joseph, Missouri 64506, USA and 2Department of Pathology, Heartland Regional Medical Center, St. Joseph, Missouri 64506, USA

Email: Chakshu Gupta - chakgupta@yahoo.com; Ashok K Malani* - drmalani@yahoo.com; Sandeep Rangineni - sandeep.rangineni@heartland-health.com

* Corresponding author

Abstract

Background: Metastatic breast carcinoids are rare neoplasms. They can be mistaken for primary breast carcinoma both clinically and radiologically, even with known history of carcinoid tumor elsewhere in the body.

Case presentation: We report a case of unilateral breast metastasis from carcinoid tumor of the small intestine in a 52-year-old woman who was successfully treated by lumpectomy and radiation therapy. An extensive review of the literature reveals only a few cases of metastatic carcinoid to the breast from small intestinal primaries.

Conclusion: Clinical suspicion for metastasis should be high in a patient with breast mass and history of known carcinoid elsewhere in the body. Lumpectomy alone may be effective in these patients. Mastectomy and especially axillary dissection could be avoided. Their histological appearance may mimic ductal adenocarcinoma of the breast. However, the distinction is important due to differences in management and prognosis.

Background

Metastatic tumors to breast represent a mere 1–2% of all breast tumors [1,2]. Common tumors that metastasize to the breast include those from the lung, prostate, thyroid, kidney, hematopoietic system, and malignant melanoma [3-5]. In children, rhabdomyosarcoma is reported to be the most common primary source of metastatic breast lesions [6]. In comparison, metastatic neoplasms from gastrointestinal primaries are rare and include the stomach, pancreas, esophagus, and colon. The majority of these tumors are adenocarcinomas [6]. Small intestinal carcinoids metastasizing to the breast have only sporadically been reported in the literature [7]. Metastatic breast carcinoid can be easily mistaken for primary breast carcinoma. This may potentially be detrimental for the patient, especially if the primary surgery is a mastectomy with axillary lymph node dissection. Many times a primary lesion may not be discovered and breast metastasis may be the presenting feature of an occult carcinoid [8,9]. Primary carcinoid tumors of the breast are also reported in the literature, although this view is no longer accepted [7,10,11]. We report a case of metastatic breast carcinoid in a 52-year-old postmenopausal nun along with a review the literature.

Case presentation

A 52-year-old postmenopausal woman, a Nun by profession, presented with complaints of alternating diarrhea.
and constipation, and abdominal distension of several days duration. Her medical history was significant for hypertension, diabetes mellitus, and psoriasis. Physical examination revealed a palpable abdominal mass extending from the pelvis to above the umbilicus. It was firm in consistency and very suspicious for malignancy. Subsequent evaluation for serum tumor markers revealed an elevated CA125 (1017, reference range < 35). Computed tomography (CT) scan of the abdomen showed a large solid midline mass in the pelvis and lower abdomen, extending to above the umbilicus and measuring up to 19 cm. It had multiple areas of low attenuation within it. In addition, several hyperdense lesions were noted in the liver, measuring approximately 1–2 cm in diameter. A third lesion was present in the posterior part of the pelvis, was solid appearing, and possibly involving the right ovary. It measured 10 cm and was noted to be pressing on the inferior vena cava and the right lower ureter causing mild hydrourephrosis.

She underwent a segmental resection of the ileum, along with a total abdominal hysterectomy, bilateral salpingo-oophorectomy, and intraoperative needle biopsies of the liver masses. Histological examination revealed a large carcinoid in the ileum with metastasis to regional lymph nodes. The tumor was present as nests and sheets of uniform cells with focal gland formation (figure 1A). Cells contained modest amount of eosinophilic cytoplasm and monotonous appearing nuclei with vesicular nucleoli (figure 1B). Focal areas of mucosal ulceration, as well as extensive perineural and vascular invasion were present. Metastatic deposits of carcinoid tumor were also found in both ovaries and in the liver.

A 24-hour urine specimen revealed predictably high levels of 5-HIAA (5 Hydroxy Indole Acetic acid) at 45.2 (reference range 0.5–9 mg/24 hours). The patient was followed regularly with 24-hour urine 5-HIAA levels and CT scans of abdomen and chest. Two years post diagnosis; mild progression of her liver disease was noted. The patient was initiated on subcutaneous octreotide (Sandostatin) at a dose of 50 mg/m² twice daily.

She remained stable for three and one half years, when she discovered a lump in her left breast. Clinical examination revealed a firm mobile mass in the breast suspicious for tumor. Mammograms were significant for an irregular, marginated mass of 2.2 × 2.0 cm in the upper inner quadrant of the left breast. An ultrasound confirmed these findings, revealing a hypoechoic, irregularly marginated mass highly suspicious for malignancy. The patient underwent lumpectomy of the breast mass for a definitive diagnosis. The gross specimen had a tan grey firm nodule of 2.5 × 1.5 × 1.5 cm with smooth borders within otherwise unremarkable fatty tissue. Histological examination revealed a 2.5 cm nodule containing infiltrating cords and nests of cells morphologically similar to her original ileal carcinoid tumor. The larger nests demonstrated an acinar pattern with rosette formation. The cells contained round to oval nuclei with a fine reticular chromatin pattern (Figure 2A and 2B). Necrosis was absent and mitoses were rare. Focally margins were positive for tumor. A partial mastectomy was performed to remove residual tumor. The patient received adjuvant radiation therapy to the affected breast. She has had no recurrences and has had stable liver disease (metastatic carcinoid) for 5 years following her breast surgery.

**Discussion**

Carcinoids are slow growing neoplasms derived from enterochromaffin cells and are thus neuroendocrine in nature. They arise most commonly in the gastrointestinal or respiratory tract [12]. Carcinoid syndrome occurs in approximately 5% of patients with intestinal carcinoids and manifests as episodes of diarrhea, abdominal pain, and flushing [13]. They typically occur in the presence of hepatic metastasis because the liver can no longer metabolize the polypeptides (including serotonin and substance P) produced by the tumor cells. This phenomenon is only observed in intestinal carcinoids due to the venous drainage of this organ system. When the primary tumor is extraintestinal, carcinoid syndrome may be produced without hepatic metastasis [8].

Carcinoid tumors are now considered at the well-differentiated end of the spectrum of neuroendocrine carcinomas. By definition, typical carcinoids have a bland morphology, lack necrosis, and have less 2 mitoses per 10 high power fields. However, carcinoids are malignant neoplasms and retain the capacity to metastasize. Morphological features of neuroendocrine carcinomas do not predict
carcinoid tumors of the breast demonstrate only argy-
carcinoid tumors of the small bowel, whereas "primary" 
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carcinoma in-situ). 
Organoid nests of cells with rosette-like structures can 
mimic the pattern of solid or cribriform DCIS (ductal car-
carcinoma at a different site in the patient [16]. Misdiagnosis of breast carcinoid as primary ductal carci-
cinoma was also in the differential diagnosis. In our patient, clinical suspicion of met-
It was discovered that the patient had a primary carcinoid 
and were subjected to mastectomies [16]. The 
carcinoid were initially considered to have primary breast 
man 
carcinoid. The appendix, duodenum, pancreas, lungs, and 
ovaries were other primary sites, occurring with equal fre-
quency in their review [8]. Metastatic breast carcinoids 
may present clinically as single or multiple well-circum-
scribed lesions, with a firm consistency. In several pub-
lished reports, they were clinically interpreted as fibroadenomas, or uncommonly as medullary or muc-
inous of ductal carcinoma [4,14,15]. In a review by Fish-
man et al, 8 of 13 (61.5%) patients with metastatic breast 
carcinoid were initially considered to have primary breast 
carcinoma and were subjected to mastectomies [16]. The 
diagnosis of metastatic breast carcinoid was made after 
review of the histology of the mastectomy specimen after 
it was discovered that the patient had a primary carcinoid 
at a different site. In our patient, clinical suspicion of met-
astatic carcinoid was high, although a primary breast carci-
oma was also in the differential diagnosis. Misdiagnosis of breast carcinoid as primary ductal carcino-
ma has also been reported even with a prior history of 
carcinoid tumor at a different site in the patient [16]. 
Organoid nests of cells with rosette-like structures can 
mimic the pattern of solid or cribriform DCIS (ductal car-
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Argentaffin and argyrophil stains are usually positive in 
carcinoid tumors of the small bowel, whereas "primary" 
carcinoid tumors of the breast demonstrate only argy-

A modified radical mastectomy was performed in the 
majority of cases reported in the literature, as the clinical 
diagnosis was that of a primary breast carcinoma. 
Lumpectomy has been performed in only a few cases 
[6,7,13,19]. Large series of such tumors are lacking in 
the literature and we cannot recommend a more specific man-
agement approach. Large series of such tumors are lacking 
in the literature and we cannot recommend a more spe-
cific management approach, however, axillary lymph 
ode node dissection may not be necessary. Estrogen receptors 
may be positive in carcinoid tumors, however, none have 
been reported positive in metastatic breast carcinoids to 
our knowledge. HER-2/neu may be positive and has been 
described in at least one report of metastatic carcinoid to 
the breast [7]. Role of chemotherapy and radiotherapy is 
also unclear.

In conclusion, metastatic carcinoid tumors to the breast 
can mimic primary breast carcinoma. In patients with a 
breast mass and a known history of carcinoid tumor, one 
should have a high index of suspicion for metastatic dis-
ease. A lumpectomy should be offered to these patients 
rather than modified radical mastectomy as first line ther-

Competing interests
The author(s) declare that they have no competing inter-

Authors’ contributions
AKM was involved in clinical management of the patient 
following her surgery. CG reviewed the pathology of the 
patient. SR was also involved in the post-operative onco-
logic management of the patient. All three were involved 
in review of radiology films, medical records, literature 
search, manuscript preparation and critical review of final 
manuscript. All authors approved the final version of the 
manuscript.
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