Costal metastasis: A singular localization of gastrointestinal carcinoid tumor

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A 73-year-old male was referred to our department for the surgical excision of a solid mass of the chest wall involving the 10th left costal arc and diagnosed by Computed Tomography (CT) scan [Figure 1] during follow-up after removal of a gastrointestinal carcinoid. Six years before, the patient underwent ileocolic resection for a neuroendocrine tumor of the distal ileum infiltrating the appendix and involving mesenteric lymph nodes. Four hepatic metastases were also resected during the same intervention (pT3N2M1). Follow-up, consisting of annual 111 Indium octreotide scan (Octreoscan) and liver ultrasound every six months, was substantially uneventful for five years.

On September 2010, the patient occasionally discovered a localized swelling of the left thoracic wall. CT scan confirmed a neoplastic lesion of 6 cm of diameter invading the 10th left rib. The patient did not present diarrhea, flushing, or any other symptom suggestive of “carcinoid syndrome.”

Questions

1. Suspecting a metastatic disease from the gastrointestinal carcinoid tumor, what additional diagnostic examinations are indicated?
2. What is the role of surgery in the treatment of metachronous metastases?

Figure 1: CT scan showing a neoplastic mass invading the 10th left rib
Answers

1. Fine needle biopsy (FNA) offers the advantage of being minimally invasive. In this case, FNA resulted positive for tumoral cells compatible with carcinoid (TTF1 - ck7 negative and chromogranin – synaptophysin positive), providing an affordable cytological diagnosis. To exclude any further lesion, Octreoscan was performed and no other metastatic spot was documented apart from the left costal lesion. Bone marrow biopsy was not considered mandatory in the preoperative work-up.

2. There is a rich medical literature concerning the role of surgery in the management of pulmonary metastases. Different parameters must be evaluated when defining surgical indication: Histology, oncologic stability of the primary tumor, absence of extra-thoracic metastases, number of lesions, disease-free interval, technical resectability and performance status of the patient. However, when dealing with thoracic wall metastasis from extra-thoracic tumors, no therapeutic guidelines are available. We considered in particular three clinical aspects: The solitary lesion, the long disease-free interval (more than 5 years), and histology. As a consequence, we opted for surgical treatment that was achieved by an en-bloc resection of the 9th and 10th left costal arcs with a free macroscopic margin of 3 cm, in order to ensure oncologic radicality [Figure 2]. Chest wall defect was reconstructed with a tailored polypropylene prosthesis.

Course of the patient and histological findings

Postoperative course was regular and the patient was discharged on postoperative day #4.

Histological examination confirmed that the lesion was constituted by neuroendocrine tumor infiltrating bone, adipose tissue, and muscle [Figure 3]. Neoplastic cells were immunopositive for chromogranin and synaptophysin. Nevertheless, the lesion was a metastatic localization, and proliferative rate, evaluated with immunostain for Mib1/Ki67, was extremely low (less than 2%) [Figure 4].

After twelve months, a repeated Octreoscan excluded relapse or further metastatic disease.

Discussion

Extrahepatic metastases from gastrointestinal neuroendocrine tumor (carcinoid) are rare occurrences.[1] In 1867, Theodor Langhans described for the first time the histological features of carcinoid tumor,[2] whereas the first autopic report of two patients with ileal carcinoid tumors was published in medical literature by Otto Lubarsch in 1888.[3]

Carcinoids are rare neuroendocrine tumors. The majority of them are found within the gastrointestinal (55%) and bronchopulmonary tract (30%).[4] Among gastrointestinal carcinoids, the most common localization is the small bowel, followed by the rectum, appendix, colon, and stomach.[5] Small bowel carcinoids most commonly occur in the distal ileum within 60 cm of the ileocecal valve, with a progressive increase of incidence in proximity to the cecum.[6] Given that they do not cause early clinical signs, diagnosis is often obtained at the time of surgery. In particular, carcinoids of the appendix are usually found incidentally by histopathologic examination.[6]
Management strategies include surgery with curative intent (whenever possible) or simply debulking, radiological interventions (by chemoembolization or radiofrequency ablation), chemotherapy and new biological agents, interferon, somatostatin analogs, and peptide-receptor radionuclide therapy.[7]

Surgery remains, however, the mainstay of treatment for primary carcinoid tumors,[4] always including a lymphadenectomy as wide as possible toward the mesenteric artery origin, associated with a prophylactic cholecystectomy. In non-metastatic disease, complete surgical resection in order to obtain microscopic healthy margins is the most important prognostic determinant affecting survival. Up to 75% of patients with carcinoid tumors develop hepatic metastases regardless of the location of the primary tumor.[4] In these cases, surgical excision should be proposed for fit patients when complete resection of the primary tumor is feasible[7] in order to reduce symptoms, improve quality of life, and prolong survival.[5] Careful intraoperative examination is essential, as 20% to 40% of carcinoid tumors of the small bowel are multicentric.[4]

Overall 5-year survival rates range from 75% for local disease to 59% in patients with positive nodes; they decrease to 35-20% if liver metastases are present,[6] although longer survival are sporadically reported.[7]

Extrahepatic intestinal carcinoid localizations are rarely reported. In the medical literature, the only two descriptions of chest wall metastases were reported by Sakamaki et al.[8] in 2005 and Seven et al.[9] in 2006.

In the present case, the patient occasionally noticed a swelling on the left hemi-thorax, since the follow-up control program ended one year before. The lesion was subsequently diagnosed as a carcinoid metastasis by needle biopsy. Considering the long disease-free interval, surgery was proposed with curative intent and consisted of radical removal of the lesion including the two involved costal segments. No postoperative complication occurred.

Costal metastasis six years after the resection of a gastrointestinal carcinoid is an extremely rare occurrence and a standard treatment protocol has not been established.[6] Considering the low proliferative rate of many of these tumors, they may develop distant localization even several years after the first operation. Therefore, an accurate long-term follow-up is essential for a correct evaluation of the natural history of this disease.

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