1. Introduction

Carcinoid crisis is caused by massive acute release of neuroendocrine substances, and patients with carcinoid syndrome are at risk for developing a carcinoid crisis during surgery or other types of intervention. The patients may have sudden change of blood pressure, bronchospasm, facial flushing and even death. Our patient presented with carcinoid crisis 24 h after bland embolization of liver metastases. Pre-procedure administration of octreotide remains the mainstay of preventive therapy for patients undergoing procedures relating to carcinoid tumors. For patients with extensive metastases, adjunct therapy with corticosteroids, cyproheptadine, and glucagon should be considered.

2. Case report

A 75-year-old male was initially found to have multiple liver lesions identified on chest X-ray at time of stress test and echocardiogram in 2005. Ultrasound-guided fine needle aspiration performed in January 2005 showed metastatic carcinoid tumor. He underwent a right hemicolecystectomy in September 2005, followed by subsequent chemoembolization of his liver metastases. In September 2008, an octreotide scan was done which showed probable progression of his metastatic hepatic disease. Subsequently, the patient underwent another chemoembolization in October 28, 2008. He had intermittent problems with anasarca and continued octreotide therapy until September 4, 2009. As his disease progressed, severe edema, respiratory distress due to pleural effusion, and renal compromise as shown by increasing creatinine and protein spillage further complicated his condition. Bland embolization was performed on January 20, 2010, which involved embolization of the main right hepatic lobe. The following day, the patient developed hypotension 48/30 mmHg and erythema of his face. He was subsequently transferred to the ICU. Patient was given octreotide drip 50 mcg/h, lasix 20 mg/h, dopamine, and levophed drip. Morphine was added for pain control and amiodarone was given for his atrial dysrhythmia. Dialysis was initiated for his renal failure. Patient subsequently developed episodes of severe hypotension, altered mental status with delirium. Beta-blockers, vasopressin, and cyproheptadine were added to therapy and octreotide was increased to 150 mcg/h. Morphine was removed to avoid carcinoid crisis. Patient was transferred to hospice for end-of-life care after developing right hydropneumothorax with complete collapse of the right lung.

3. Discussion

Carcinoid tumor arises from the diffuse neuroendocrine cells of the intestinal tract with the unique ability to secrete bioactive amines and peptides. Prevalence of carcinoid tumor is estimated to be two cases per 100,000 persons in the United States (1). Majority of the tumor are found incidentally during other surgical procedures. Carcinoid tumors can cause carcinoid syndrome which is caused by chronic release of neuroendocrine substances such as serotonin, histamine, kallikreins, and catecholamines produced by the carcinoid tumor. The syndrome can be divided into 2 categories: typical and atypical. Typical carcinoid syndrome (TCS) is usually caused by metastatic midgut
carcinoids. Atypical carcinoid syndrome (ACS) is most often associated with metastatic foregut carcinoids. Both TCS and ACS cause flushing but the pattern is patchier in ACS. Additionally, these areas of flushing often exhibit central clearing and increased pruritus when compared with flushing due to TCS. Due to the increased systemic levels of histamine often present in foregut carcinoids, bronchoconstriction, salivary gland swelling, cutaneous edema, and lacrimation may also be present (2). Only 10% of all patients with carcinoid tumor develop carcinoid syndrome (3).

Pre-, peri-, and post-operative care for carcinoid tumor varies. Some advocate octreotide for patients who have carcinoid syndrome and are planning for surgical removal (4), while others recommend octreotide for all patients with carcinoid tumors who are planning for surgery (5). In addition, H1 receptor blockers, H2 blockers, and occasionally, dexamethasone may be needed if symptoms are severe. It is recommended that patients be evaluated with echocardiogram prior to surgery for the presence of carcinoid heart disease. Electrolytes should be obtained in patients with severe diarrhea prior to surgery. Parenteral nutrition may be needed in patients with weight loss and hypoproteinemia especially before major surgery. Niacin deficiency is common since the essential amino acid tryptophan is converted to serotonin in excessive amounts in carcinoid syndrome and needs to be addressed/monitored prior to surgery (6).

Massive acute release of neuroendocrine substances results in carcinoid crisis. Life-threatening hypotension, arrhythmias, bronchospasm, facial flushing, edema, metabolic acidosis, coma, confusion, and death characterize the crisis (7). Carcinoid crisis from bland embolization have been reported to occur peri- or 24 h after procedure (8). Carcinoid crisis can be precipitated by a number of environmental triggers such as stress, alcohol, amine containing food products, medications, and non-environmental triggers such as tumor manipulation during surgery, embolization, or chemotherapy (9). High pre-operative urinary 5-HIAA and carcinoid heart disease are significant risk factors for perioperative complications (10). Treatment of a carcinoid crisis includes fluid expansion and use of 50-500 μg IV octreotide. Current recommendation for treatment of carcinoid crisis with hypotension that develops during procedure is 500 μg IV bolus every 5 min until symptoms are controlled (11). Ketanserin (selective antagonist of the 5-hydroxytryptamine receptor 2, the α1-adrenoreceptor, and the H1-histamine receptor) 10 mg IV bolus can be used to block the actions of the offending mediators in carcinoid crisis (12). Avoidance of catecholamines in the treatment of hypotension is stressed because catecholamines may stimulate tumor cells to release even more serotonin.

Our patient presented with carcinoid crisis 24 h after bland embolization of liver metastases. Pre-procedure administration of octreotide remains the mainstay of preventive therapy for patients undergoing procedures relating to carcinoid tumors. For patients with extensive metastases, adjunct therapy with corticosteroids, cyproheptadine, and glucagon should be considered.

References


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