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Mucinous Cystadenocarcinoma of the Appendix*

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원발성 충수선암은 대부분 급성 충수염의 증상으로 내원 충수절제후 우연하게 진단을 내리게 되는 아주 드문 질환이다. 충수선암의 첫 증례는 Berger에 의해 1882년에 보고 되었다. 그때 이 종양이 우 하복부에 촉지된 균상 종괴로 나타나 정확한 발생부위가 충수인지 맹장인지 확실치 않은 것으로 알려져 있다. 문헌고찰에 의하면 1982년까지 충수 암양종(carcinoid)을 제외한 원발성 충수암이 영어로 발표된 문헌에 200예 이하로 보고되어 있다. 저자는 75세 된 여자가 비대한 하부복부 종괴로 내원하여 수술 후 충수 점액성 낭종암으로 진단된 증례를 치험 하였고 또 그 임상적 소견과 혈청 CEA 수치가 특이하여 문헌고찰과 아울러 그 증례를 보고한다. 일반적으로 잘 알려진 점액유종(mucocele)과 점액성 낭종암과의 관계도고찰 하였다.

Introduction

Primary adenocarcinoma of the appendix is not often suspected preoperatively and very seldom, if ever, has been diagnosed prior to surgical exploration. The first case of adenocarcinoma of the appendix was reported by Berger in 1882.[1] This tumor apparently presented as a fungating mass in the right lower quadrant of the abdomen and exact site of origin as to the appendix or the cecal area is not clear.[2] Review of the literature suggests that less than 200 cases of non-carcinoid malignant epithelial neoplasms of the appendix have been reported until 1982.

One patient with mucinous cystic adenocarcinoma who presented with a large abdominal mass without usual symptoms and signs of acute appendicitis is presented. An interesting feature of this patient is that her preoperative plasma carcinoembryonic antigen (CEA) level was 1,400 ug/ml with normal range from 0 to 2.5ug/ml and postoperatively this level gradually came down to a normal level of 1.7 ug/ml in 7 months and stayed within normal range since.

Report of a Case

A 72 year old Caucasian female was admitted to the hospital because of a large abdominal mass. Exact duration was unknown but gradual protuberance of her abdomen was noticed by her family at least for about a year. The patient complained of urgency and frequency of urination, voiding small amount each time and nocturia, getting up approximately six

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times a night. Patient denied nausea, vomiting, abdominal pain, or bowel irregularities. She has suffered from forgetfulness and mental confusion and pertinent history was obtained from her family. Past history was essentially negative for serious illnesses. Father died at the age of 80 in a mental institution and mother died at the age of 50 from carcinoma of the stomach. One sister was suffering from diabetes mellitus and hypertension but seven remaining siblings were reportedly healthy. The patient was on no medication at the time of admission and she did not drink alcohol nor smoke. Physical examination revealed confused and forgetful elderly lady, physically healthy looking without any distress. She was cooperative and followed instructions reasonably well. Temperature was 98.5° F, pulse 108, respiration 20, blood pressure 150/78. A large basket ball size mass was visible and palpable in the lower abdomen reaching just above the umbilicus. Pelvic examination was not conclusive due to large abdominal mass which prevented accurate palpation of the uterus. No hepatosplenomegaly was present and rectal examination was negative and occult blood for stool was also negative.

Hemoglobin was 13.7, hematocrit 43%, WBC count 7,200 with normal differential count. Chemical profile was entirely within normal limit. Plasma assay of carcinoembryonic antigen (CEA) was markedly elevated to 1,400 ug/ml with normal range from 0 to 2.5 ug/ml. Chest X-ray showed elevated right hemidiaphragm but otherwise normal and flat plate of abdomen revealed a large soft tissue density over the lower abdomen but the margin was not well defined. Several calcifications were seen in the pelvis above the urinary bladder. Intravenous pyelogram identified mild dilatation of the right collecting system and right proximal ureter probably due to compression on the right ureter by the mass. Flexible fiberoptic sigmoidoscopic examination was normal up to 35 cm from the anal verge and Pap smear was class I showing no tumor cells.

At operation on September 17, 1980, a large cystic mass was found occupying entire lower abdominal and pelvic cavity and the upper end of the cystic mass reached above the umbilical level. Marked adhesions were present between peritoneum and the mass. Some mucinous material was lost during manipulation. When the entire cystic mass was finally mobilized and freed from all adhesions, it was found to be originating from the appendix. The base of the appendix, about 2 cm in length, was grossly normal. The cystic mass with normal base of the appendix was removed and the peritoneal cavity was thoroughly irrigated removing all mucoid spillage. Exploration of the abdominal cavity showed atrophic ovaries and uterus with numerous fibroids but no evidence of metastasis was found. Postoperative course was uneventful.

The gross specimen was a 22×12×7cm, oval, pale tan, smooth, glistening mass arising from appendix which had a weight of 800 gm. The actual size was bigger than this due to some spillage and suctioning through a tear in the cyst during surgery. A mucoid irregular shaped excrescence 9×4×3.5cm was protruding from one portion of the mass. The cyst wall was ranging up to 0.3 cm in thickness and lined by mucoid material and friable, amorphous, pale yellow material. In one section of the wall there was a poorly circumscribed, friable, granular, calcified mass 2.5 cm in greatest dimension. The base of the appendix connected to the mass measured 1.5 cm in diameter and 2.0 cm in length and the lumen appeared distended by mucus and mucoid material.

Microscopic examination showed primarily mucinous material with occasional islands of epithelial cells which are mucin producing and showed significant cytologic atypia and invasion. Sections of the base of the appendix showed a marked proliferation of the mucosa and long papillary fronds lined by abundant mucin producing cells showing various degrees of cytologic atypia. There was only very focal invasion of the wall by the neoplasm in these sections.

Further operation for right hemicolectomy was recommended but patient's family refused. Plasma CEA levels postoperatively were:350ng/ml on September 23, 1980, 3.5 on December 22, 1980, and 1.7 on April 6, 1981. CEA levels afterwards remained within normal limit.

At the time of this report this patient showed no sign of any tumor recurrence or metastatic disease and other than her mental status and obesity which she has acquired since the operation, her physical condition was satisfactory.

Discussion

Tumors of the appendix are uncommon and primary adenocarcinoma of the appendix occupies only 6% of all neoplasms of the appendix.[2] The incidence of primary adenocarcinoma of the appendix has been estimated as 0.08% of all appendices removed surgically[2] and it comprises 0.2 to 0.5% of all gastrointestinal neoplasms.[4] There has been considerable confusion regarding the classification of these tumors. Usually these tumors are classified into three types:[5] carcinoid type [90%], cystic type [8.5%], and colonic type [1.5%]. The carcinomas of the cystic type has been known as malignant mucoceles. The term mucocele refers to a lesion of the appendix with a dilated lumen filled with mucus. In 1973, Higa and his associates recommended elimination of the term "*mucocele*" since it referred only to the presence and effets of accumulation of mucus in the lumen of the appendix and they reclassified "*mucoceles*" into focal or diffuse mucosal hyperplasia, mucinous cystadenoma, and mucinous cystadenocarcinoma.[5,7] Arahna and Reyes presented a new classification of primary epithelial tumors of the appendix after reviewing 19 primary neoplasms of the appendix seen between the years 1950 and 1978 at the Hines V.A. Medical Center which was more or less similar to the classification of Uihlein and McDonald mentioned above. Arahna and Reyes also recommended that the term mucocele should be discarded.[6] Only one out of 19 patients with primary neoplasms of the appendix had mucinous cystadenocarinoma in their series.[7]

The majority of patients with adenocarcinoma of the appendix appear to have symptoms and signs of acute appendicitis and the carcinoma was found only incidentally after appendectomy. Very rarely patient may show signs of right colonic obstruction or palpable mass. For treatment, appendectomy alone is sufficient for carcinoid of the appendix where there is no gross evidence of local metastasis and when the tumor does not involve the cecal wall and is less than 2 cm in size.[5,10] For mucinous cysadenocarcinoma of the appendix which is confined to the mucosa of the tip of the appendix, simple appendectomy alone may be sufficient but many feel that this is not an adequate operation. Arahna and Reyes recommend appendectomy and right hemicolectomy for all adenocarcinomas of the appendix, whether they are of the mucinous or colonic type; all adenoacanthomas; and for all carcinoids of the appendix that are 2cm or greater in size, or have evidence of gross local metastasis.[6] The patient reported here was treated with appendectomy alone. Right hemicolectomy was recommended after the tissus diagnosis was obtained but the patient's family refused. So far the appendectomy alone seems to have served to cure the patient although longer follow up will be needed for definite answer.

The relationship between this patient's mucinous cystadenocarcinoma and markedly elevated CEA levels pre and postoperatively and finally coming down to the normal level 7 months after the resection is not clear. It is generally known that CEA is not useful as a screening test for cancer and of limited clinical value when obtained preoperatively in patients who have carcinoma of the large bowel, but the CEA is of some help for postoperative follow up of patients and the prognosis is usually poor if an elevated preoperative level does not fall to normal within three weeks.[11] Review of the literature regarding the mucinous cystadenocarcinoma of the appendix does not mention any

relationship with CEA levels. CEA is an acid glycoprotein present in the glycocalyx of the fetal digestive system that is rapidly metabolized, probably by the liver. Serum concentrations usually fall to undetectable levels 2 to 14 days following removal of a tumor that is producing the antigen.[12] Clinical evaluation of serum CEA assays indicates that the antigen has broad distribution in neoplastic disease and also occurs in significant levels in some non-neoplastic disorders.[11]

The significance of very high level of CEA in this patient and very slow disappearance from the serum awaits further experience with same type of patients.

Summary

A very rare case of mucinous cystadenocarcinoma of the appendix in a 72 year old Caucasian female is presented. The patient presented with a large abdominal mass and the diagnosis was made after exploratory laparotomy. Appendectomy was the only treatment but the patient is alive and well without any evidence of tumor recurrence or metastasis almost three years after the operation.

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