Gezira Journal Of Health Sciences vol.2(2) 2006

EDITORIAL

PSEUDOMYXOMA PERITONEI A RARE CASE

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ABSTRACT:

This is a case report of a 55- year old male patient who presented with features of intestinal obstruction. The obstruction which was relieved surgically by removal of fibrous adhesions was found to be the result of Pseudomyxoma peritonei however, at operation the primary focus of the tumor was not found because of extensive local metastasis. The patient received parental chemotherapy inform of intravenous 5-fluorouracil (6oomg/day 5 times/ week) for 6 month and was discharged from hospital in good condition.

الملخص

هذا تقرير لحالة طبية نادرة لمريض يبلغ من العمر 55 عاماً حضر للمستشفى يعاني من انسداد معوي. تم إفراج الانسداد جراحياً "بإزالة الالتصاق الليفي " ووجد أن الانسداد المعوي ناتج من ورم مخاطوم صفاقي كاذب. كان من الصعب التعرف على البؤرة الأولية للورم أثناء العملية بسبب النقيلة الموضعية المديدة .
تلقى المريض علاج الكيمائي في صورة (5 fluorouracil 600ملغرام/اليوم ، 5 مرات في الأسبوع لمدة 6 أشهر) و خرج من المستشفى بحالة جيدة.

INTRODUCTION

Pseudomyxoma peritonei syndrome first described by Karl F. Rokilansky in 1842 is often a fatal intra - abdominal disease [(1)] .Characterized by mucinous ascites and mucinous tumor disseminated on peritoneal surface; the disease almost always originated from a perforated appendiceal epithelial tumor. Histopathologic assessment of aggressive versus non invasive character of the mucinous tumor has been shown to have an impact on the survival of patients treated with cytoreductive surgery and intraperitoneal chemotherapy [(2)] .The syndrome associated with repeated mucocele, cysadenoma, and low grade carcinoma arising from appendix, ovaries or colon and rare in pleural cavity ([3]) . Metastasis and extra peritoneal involvement are extremely rare events ([4]).

The operative morbidity and mortality are null. The global 5- year s survival rate is 60% (100% in benign diseases and 0% in malignant form. The syndrome is still poorely understood. The origin of the disease is appendix (70-80%) and less frequency the ovary ([5, 6]) .

EDITORIAL

Acute appendicitis is the common presentation 27%. For women the diagnosis was most commonly made while being evaluated for an ovarian mass 39%. Increasingly abdominal girth was the second most common presentation overall 23%, 14% presented with new onset hernia, of which the majorities were inguinal hernia [7, 8] .

The morbidity of treated patients 27% and the mortality is 2.7% (9). Morbidity is principally related to mechanical complications due to ascitis and characteristic of the groups of the secretary cells [10].

Gezira Journal Of Health Sciences vol.2(2) 2006

Abdominal ultrasonography, computer tomography and laparoscopy are of significance in the diagnosis of this disease which is yet confirmed usually during surgery. The treatment is principally surgical, the result of chemotherapy are modest .The disease has slow course and is susceptible to recurrence [11].

CASE PRESENTATION:

A 55 year old male patient was admitted to hospital because of gradual onset abdominal distension, absolute constipation & persistent vomiting 3 weeks prior to admission. His condition was not associated with any febrile illness. He lost weight despite a good appetite. He has no past history of tuberculosis, intestinal schistosomiasis or chronic diarrhea. He was alcoholic and cigarette smoker for the last ten years. Clinical examination revealed a cachexic patient with pallor and gross finger clubbing. The cardiovascular and respiratory systems were normal; however the abdomen was distended with full flanks and multiple abdominal masses. The provisional diagnosis was of intestinal obstruction. Investigations done revealed the following: Hb 8.8 gram/dl, WBC 2100/cml, E.S.R. 110 ml/h, Liver function test, ECG, upper G.I.T Endoscopy and chest X- ray were normal. Ultrasound of the abdomen showed a cirrhotic liver, enlarged paraaortic lymph gland. I.C.T. for tuberculosis was negative. Plain x-ray abdomen showed distended loops of small bowel.

The patient was managed initially conservatively i.e.: nothing by mouth, nasogastric suction, l.V. fluids, ceftriaxone 1 gm I.V. daily and metronidazole infusion. Laparotomy was done 2 days later. The findings at Laparotomy were of mucinous containing masses in the sheets of peritoneum with massive adhesions & distended small bowel loops. The transverse colon was fixed to the anterior abdominal wall frozen abdomen. Fibrous adhesions were relived. No primary lesion can be found at operation. A biopsy was taken. The result of the biopsy showed mainly mucous, mucoid material with scanty abnormal cells, consistent with Pseudomyxoma peritonei. The patient received 5 fluorouracil intravenously (6oomg/day 5 times/ week) for 6 months and was discharged from the hospital in good condition.

DISCUSSION

EDITORIAL

Pseudomyxoma peritonei occurs more frequently in females. The abdomen is filled with a yellow Jelly, large quantities which are often more or less encysted. The condition is associated with mucinous cystic tumors of ovaries and appendix. The condition arises in one of 2 ways either from a mucocle of the appendix or rupture of pseudomucinous cyst of the ovaries.

It is often painless and there is no impairment of general health for a long time. When the condition arises from the appendix the mass is often more localized but in cases of ovarian origin the whole peritoneal cavity is involved. Although the abdomen is distended with what seems to be fluid that cannot be made to shift should suggest the possibility. It is highly improbable that a correct preoperative diagnosis will be made. At Laparotomy masses of Jelly are scooped out and the primary focus if can be found is removed. Unfortunately recurrence is usual.
Occasionally the condition responds to radioactive istopes which should certainly be employed in recurrent cases. Interferon has been used in the treatment of this condition ([12)].

Recent advances in treatment of pseudomyxoma peritonei included extensive surgery with direct administration of selected drugs into the periotoneal cavity. This permits delivery of high concentrations of drugs directly to abdominal and pelvic surfaces where the tumor is located. This pharmacological advantage is the result of the peritoneum plasma barrier. A major problem in the past with intraperitoneal chemotherapy delivery involved non-uniform drug distribution. This resulted from intestinal adhesions, from tissues closed off by sutures, and from pooling of inraperitoneal fluid at dependent sites. The use of heated intraoperative intraperitoneal chemotherapy (HIIC) after complete dissection of an adhesive process and before anastomoses are completed minimizes the problem. It does not only improve drug distribution but also improves the drug penetration into tissue compared to normothermic drug administration. Common drugs which are in use include mitomycin, 5 fluorouracil and leucovorin ([13)].

CONCLUSION:

Pseudomyxoma peritonei is a locally malignant tumor; preoperative diagnosis is unlikely to be established. In our patient the diagnosis was established at lapratomy. The obstruction was relieved surgically and the patient received 5 fluorouracil posts operatively. His general condition improved and he was discharged from the hospital in good condition.

REFRENCES

1. O, Connell JT, Tomlinson JS, etal. Pseudomyxoma peritoni syndrome is a disease of MUCSexpressing goblet cells. Am J Pathology 2002 Aug; 161

   (2): 551-6

2. Yan H., Pestieau SR, Shmookler BM and Sugarbaker PH. Histopathologic analysis in 46 patients with Pseudomyxoma peritoni syndrome: failure versus success with a second look operation. Mod Pathol 2001 Mar, 1 4 (3): 164-71.

EDITORIAL

3. Peek DF, Beets GL. Pseudomyxoma peritoni in the pleural cavity: report of a case. Dis Colon Rectum 1999 Jan; 42 (1): 113-5
4. Dejong CH, Booster MH, Theuniseen PH, Beets GL, Duin CJ. Ned Tijdschr Geneesked. 1997 Jun 14; 141
5. Angelescu N, Bordea A, Popa E, Constantinescu N, Zodieru 1, Mircea N. Pseudomyxoma peritoni (gelatinous peritonitis. Chirurgia (Bucur). 2001 Sep Oct; 96 (5); 443-51
6. Van Ruth S, Ackerman YI, etal .Pseudomyxoma peritoni: a review of 62 cases .Eur J Surg Oncol.2003 Oct; 29(8):682-8
7. Lo NS, Sarr MG. Mucinous cystadenocarcinoma of the appendix. The controversy persists: a review. Hepatogastroenterology. 2003.
8. Esquivel J.; SugarbakerP.H. British Journal of Surgery .Vol 87, Number 10, October 2000, pp 141414 (5)
9. Sugarbaker PH. Cytoreductive surgery and preoperative intraperitoneal chemotherapy in patient with Pseudomyxoma peritoni syndrome. Eur J Surg Oncol 2001 Apr; 27(3):239-43
10. Curant C, Burn G. Mucoid peritonitis .Apropos of 2 cases reports. Review of the literature. Rev Fr Gynecol Obstet .1984 Oct; 79 (10):641-6
11. Besznyak I, Pommershein F, Toth J. Pseudomyxoma peritoni .Orv Hetil. 1996 Dec 15; 137 (50): 2803-7
12. Bailey and Love s Short practice of Surgery 23rd edition 1007; 2000 Arnold 338 Euston Road London NW1 3BH
13. http:// www. Surgical oncology. Com / pmp. Htm. Sugarbaker oncology associates. Specialty section for the treatment of Pseudomyxoma peritonesyndrome