Case Report

Pseudomyxoma Peritonei - A Rare Abdominal Tumor

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Pseudomyxoma peritonei is a rare abdominal tumor with a reported incidence of approximately 1 per million per year. It is invariably fatal as the space within the abdomen is eventually replaced by mucinous tumor. We report one case of Pseudomyxoma peritonei.

Keyword; Pseudomyxoma peritonei, cytoreduction surgery.

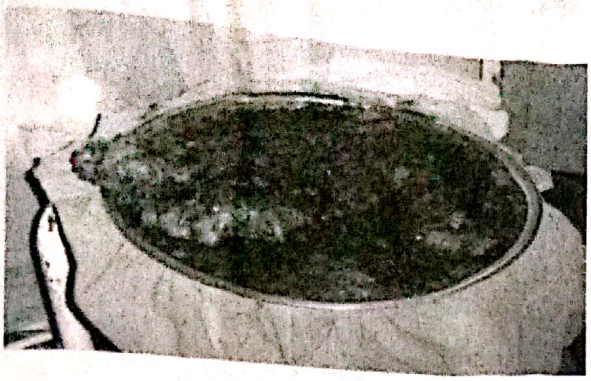
Pseudomyxoma peritonei is a rare disease characterized by copious production of mucinous ascites (picture 1&2).In 1884 W th coined the term PMP in relation to mucinous carcinoma of ovary[1]. In 1901, Frankel described PMP in association with a cyst in appendix (picture 3). It has an incidence of 1 per million per year. Median survival is approximately six years. Five year survival ranging from 53-75%

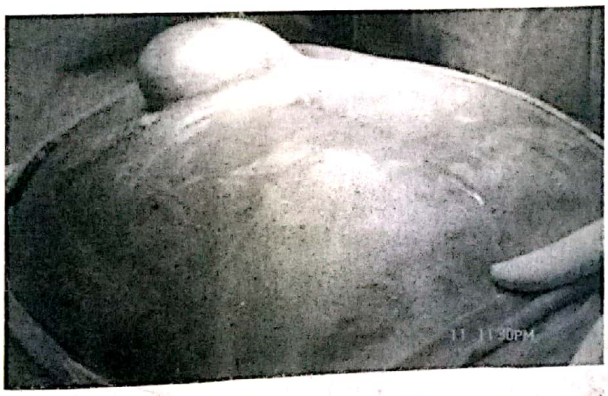
Diagnosis is by cross sectional imaging. CT scan, image guided percutaneous biopsies, and tumor markers; CA-125, CEA, and CA19-9[9]. Controversy still continues as to the etiology of PMP, particularly in women. PMP usually arises from a tumor of appendix which is originally a tumor in the sub mucosa of appendix and it presents as Jelly Belly[3]. Sugar baker and colleagues have tried to clarify the uncertainty by defining the term PMP syndrome be strictly applied to a pathologically and diagnostically homogenous group of cases characterized [illegible] or histologically that are frequently [illegible] al mucinous tumor[6].

[illegible] presented to us (FMH) in [illegible] history of pain abdomen and [illegible] years duration He was [illegible] to us and diagnosed as a [illegible] itonei. Exploratory [illegible] aching hospital in Lahore [illegible] he tumor was extending all [illegible] al distention and pain was [illegible] ission to surgical department [illegible] After detailed history and [illegible] gery was planned. Whole of the [illegible] intestines were studded with jelly [illegible] procedure [illegible] ft and right [illegible] and pelvic [illegible] d junction was [illegible] rs to complete [illegible] Patient started [illegible] day and was [illegible] ent [illegible] ion visits. On six month visit CT scan abdomen and pelvis was done, and it was unremarkable.

Discussion

PMP is a disease characterized by copious production of mucinous ascites. It usually arises from a tumor of appendix and presents as Jelly Belly. PMP is regarded as borderline malignant. It doesn't invade or metastasize, but is invariably fatal as the space required within the abdomen is eventually replaced by mucinous tumor. Lymphatic and hematogenous spread may not occur even in the terminal stages of the disease. Tumor spread in the abdomen is by the redistribution phenomenon. Tumor cells in the peritoneal cavity are surrounded by mucous and moves with the normal flow of peritoneal fluid[7]. Adenomatous epithelial cells surrounded by mucous accumulates at specific abdominal and pelvic sites. Fluid is absorbed and cells are filtered resulting in bulky accumulations. Other mechanism of tumor redistribution is Gravity. There is visceral sparing of PMP but the exceptions are antrum of stomach and pylorus, Ileo-cecal region and rectosigmoid colon. Median survival is approximately 6 years[4]. Mucinous adenocarcinoma from any condition can simulate PMP clinically, radiologically and at operation. As for all tumors, there are four main treatment modalities: chemotherapy, radiotherapy, expectant treatment with symptom control and surgery. Radiotherapy and chemotherapy have a very limited role in the treatment. Intraperitoneal chemotherapy along with cyto-reductive surgery increases the 5 year survival as compared to the results after surgery alone[2]. Mitomycin C and 5-fluorouracil are the main drugs used. The surgical procedure includes stripping of parietal peritoneum and resecting intra abdominal structures at fixed sites that contain tumor laden visceral peritoneum. In our patient cyto reduction was done up to the maximum as it is the main stay of treatment. We did not offer intra-peritoneal chemotherapy to the patient because of lack of facilities for this treatment option. Patient is fine after one year follow up. So, this case report is highlighting the importance of surgery in PMP management to accomplish a complete cyto reduction[8].





Conclusion

Pseudomyxoma peritonei is a rare condition which, though of 'borderline' malignancy is invariably fatal. The optimal treatment is undoubtedly complete tumor excision, by complex surgical peritonectomy procedures, taking on average 10 hrs. Surgery is usually combined with intra-peritoneal, and now intra-operative, chemotherapy. These techniques have a high morbidity and mortality.

References

1. Fann JI, Vierra M, Fisher D et al. Pseudomyxoma peritonei. Surg Gynecol Obstet.1993:177:441-447.
2. Parvaiz A, Amin Al, Howell R, Sexton R, Moran BJ. First one hundred referrals, predominantly of Pseudomyxoma peritonei to a peritoneal surface malignancy unit: operability and early outcome. Rr J Surg (suppl) 2002: 89:3.
3. Esqviel J, Sugarbaker PH. Clinical Presentation of Pseudomyxoma peritonei syndrome. Br J Surg 2000:87:1414-1418.
4. Gough DB, Donohue JH, Schutt AL, Gonchoroff N, Goellner JR, Wilson TO, Pseudomyxoma peritonei; Longterm patient survival with an aggressive Regional approach. Ann Surg 1994: 219:112-119.
5. WitkampAL, De Bree E, Kagg MM, V... Coevorden F, Zoetmulder FA... cytoreduction and intraop... intraperitoneal chemothera... Pseudomyxoma peritonei.Br J S...
6. Sugarbaker PH, Ronnett ... Pseudomyxoma peritone... surg1997:30:233-280.
7. Sugarbaker PH. Pseudom... en... biology is characterized ... Ann Surg 1994; 219; 109-1...
8. Sugarbaker PH, cytoredu... Pseudomyxoma peritonei. Br...
9. Sulkin, TVC, Oneill H, An... Pseudomyxoma peritonei; a review... 2002, 57; 608-613.