Adenocarcinoma of Appendix Presenting as Flank Fistula

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Discussion

Neoplasm of appendix account for less than 1 % of all gastro-intestinal malignancies [1]. Only about 250 cases of appendicular adenocarcinoma have been reported since Berger first described it in 1882 [2]. Adenocarcinoma of the appendix is usually seen in the 6th to 7th decade with a male preponderance. The most common presentations of appendicular adenocarcinoma includes appendicitis, palpable abdominal mass, intestinal obstruction and pseudomyxoma peritonei [3]. Unusual presentation includes haematuria due to bladder infiltration [4], direct invasion of ascending colon noted on colonoscopy, hydronephrosis, retroperitoneal abscess, vaginal bleeding [5] and cutaneous infiltration [6]. This unusual presentation result from the propensity of appendicular mucinous adenocarcinomas to perforate and to form fistulae to adjacent structures Figs. 1, 2 and 3.

Adenocarcinoma of appendix is most frequently perforating tumour of gastrointestinal tract [7]. This is due to anatomical peculiarity of appendix which has an extremely thin subserosal and peritoneal coat and the thinnest muscle layer of the whole gastrointestinal tract.

There are four major histological subtype of primary appendicular adenocarcinoma. They are Cystic, Colonic, Carcinoid and adeno carcinoids. Carcinoids are most common, constituting nearly 90 % of the primary tumors of appendix. Macroscopically they produce mucin filled cystic dilatation of the appendix. Malignant tumour normally spreads intraperitoneally through lymphatic. Hematogenous spread is rare [5].

In addition to the risk of perforation, mucinous adenocarcinoma of appendix have peculiar tendency for fistula formation. Many of unusual presentations reported for primary appendicular carcinoma are the results of fistula formation into the adjacent viscera such as the urinary bladder, bowel or vagina as well as extraperitoneally into retroperitoneal tissues or directly to the skin surface. Extraperitoneal spread is associated with relatively good prognosis by preventing the development of peritoneal carcinomatosis [8].

Adenocarcinoma of appendix unless confined to organ needs aggressive approach for treatment. Surgery, i.e. right hemicolectomy is primary treatment of cystadenocarcinoma of appendix when faced with an intra-operative diagnosis. Post-operative diagnosis after appendicectomy requires second surgery in form of right hemicolectomy [9]. Once pseudomyxoma peritonei secondary to the tumour occurs, patient requires aggressive surgical debriement of all deposits on peritoneum besides right hemicolectomy and excision of neighbouring compromised organs [9]. Chemotherapy and radiotherapy have also shown to be helpful [10, 11]. Similarly,

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Fig. 1 Clinical photo showing fistula in right lumbar region

tumours with extraperitoneal extension requires excision of the involved tissue and fistulous track besides right hemicolectomy [5, 8].

Insidious nature of the extraperitoneal rupture of mucinous adenocarcinoma of appendix without any preceding signs of appendicitis may lead to misdiagnosis of the source and nature of infection. Common diagnosis such as tubercular infection, diverticular disease or perirenal abscesses come in consideration. If left untreated or wrongly treated the abscess point at abnormal site such as in our case. The number of operative procedures performed are increased with increase in morbidity and mortality.

Fig. 2 Contrast enhanced CT scan of abdomen showing ceecal mass with extension into parietal wall

Fig. 3 CECT Abdomen showing loculated fluid collection in right paravertebral region extending into adjacent parietal wall

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