OLGU SUNUMU (Case Reports)

Inflammatory Fibrosarcoma: A Rare Tumor Involving Retroperitoneum, Ileum And Colon

İnflamatuar Fibrosarkom: Retroperiton, İleum ve Kolonu Tutan Nadir Bir Tümör

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Abstract
An inflammatory fibrosarcoma is a rare tumor with a varied biological behavior, from benign to locally aggressive and occasionally malignant. It generally involves mesentery and retroperitoneal structures, but it rarely can involve the hollow viscera and commonly affects children and young adults.
We report a case of inflammatory fibrosarcoma affecting an unusual age group, involving the ileum and descending colon (a rare site) causing subacute intestinal obstruction. We also discuss its surgical management, differential diagnosis and review of current literature.

Keywords: **Colon; Fibrosarcoma; Intestinal obstruction; Intestine; Small .**

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Özet
İnflamatuar fibrosarkom benignden lokal olarak agresif ve nadiren de malign değişikliliklere kadar uzanan biyolojik davranışlar gösteren nadir bir tümördür. Genellikle mezenter ve retroperitoneal yapılarda görülmesine rağmen nadiren içi boşluklu organları da tutar ve sıklıkla çocuk ve genç erişkinleri etkiler. Sunulan çalışmada subakut ince barsak tıkanıklığına neden olan nadir bir yerleşim yeri olarak retroperiton, ileum ve inen kolonu tutan; atipik yaş grubu içerisindeki inflamatuar fibrosarkom olgusunu takdim ettik. Biz ayrıca bu olgumuzun cerrahi yönetimini ve ayırıcı tanısını güncel literatür eşliğinde tartıştık.

Anahtar Sözcükler: **Bağırsak tıkanıklığı; Fibrosarkom; İnce barsak; Kolon.**

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Introduction

The inflammatory fibrosarcoma entity was first described by Meis & Enzinger in 1991. Tumors of the mesentery, retroperitoneum, and omentum which histologically contain fibroblasts, plasma cells, and histiocytes have been reported under different names in the past, including inflammatory pseudotumour, plasma cell granuloma, myofibroblastoma and inflammatory myofobroblastoma. These previous histological reports of intra abdominal lesions have been compared to inflammatory pseudotumor of the lung and their apparent benign behavior. The similarity extends to the relief of systemic symptoms, including fever, weight loss, and anemia following surgical removal of the mass (1). In fact, inflammatory fibrosarcomas are locally aggressive, potentially metastatic lesions that may lead to the patient's death and therefore should be designated as sarcomas rather than as cellular inflammatory pseudotumors (2).

Case Report

A 67 years old woman presented with a two weeks history of generalised abdominal pain localized mainly in the left iliac fossa.

She was previously fit and healthy and started with generalized feeling of being unwell for four months, recurrent pain abdomen, bloating sensation, loss of weight over 3kg and loss of appetite. There was no history of bladder or bowel disturbance, fever, jaundice or vomiting. Her pain was worse in the last 2 days associated with nausea.

On clinical examination she appeared anxious and sweaty with mild dehydration. Abdominal examination revealed tenderness and guarding in the left iliac fossa with the rest of the abdomen soft and there were no masses palpable. Rectal examination was unremarkable.

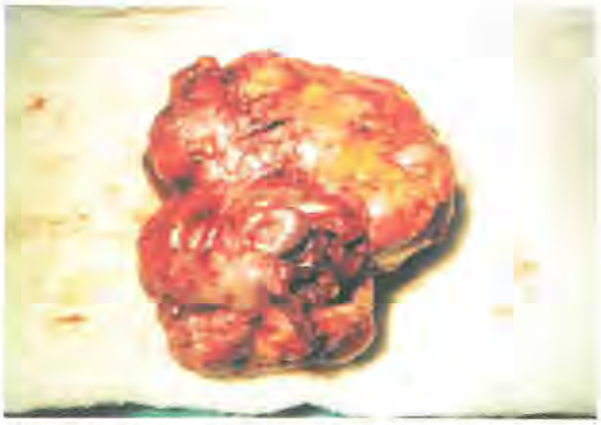
Hematology showed a white cell count of 15.600 and biochemical investigations were within the normal limits. Chest X-Ray was normal with the abdomen plain film showing a few dilated small bowel loops. Urgent ultrasound scan of abdomen confirmed a small collection of fluid in the left iliac fossa. Preoperative Computerized tomography scan showed a small bowel related pelvic mass with 15x12x10cm in dimensions.

In view of the diagnosis of pelvic mass, laparotomy was carried out. This revealed a large mass involving retroperitoneum, ileum segment and also neighbouring descending colon in the left iliac fossa. Resection of the mass (Picture 1) and 20 cm ileal segment and an end to end ileo-ileal anastomosis was performed. The postoperative recovery of the patient was uneventful.

The histopathology was initially reported as highly cellular spindle cell tumor, with frequent mitotic activity with the most likely site of origin of tumor being bowel wall and spreading in to the surrounding areas including the mesentery.

Tumor markers SMA, desmin and vimentin (indicative of smooth muscle origin) were positive there by indicating leiomyosarcoma. In view of the positive cytokeratin immunostaining (indicates epithelial origin), a second opinion was sought by pathologists.

A further histological report showed the tumor to have resemblance to an inflammatory pseudo tumor, however the pleomorphism was against. This on final review they concluded infact, was an "Inflammatory fibrosarcoma of the ileum" a rare tumor. At this stage the patient was referred to the oncologists.



Picture 1. Macroscopic appearance of the resected mass.

Discussion

Inflammatory fibrosarcoma is generally seen in children and adolescents (1) (in a retrospective study over 45 years, in the largest series of 38, patients 30 were younger than 21 years). Patients commonly present with non specific symptoms like abdominal pain, anemia, fever, night sweats (2), weight loss, mass, abdominal distension, and diarrhea. Duration of the symptoms can be highly variable ranging from acute onset to years.

Most of the tumors involve the mesentery with only a few closely related to gastro intestinal tract and involving transmurally. Retroperitoneum is the site of involvement in 87% of cases though the mediastinum and omentum can also be involved. Esophagus, bone (4) and pancreas (5) have also been reported as other rare sites. The biological behavior of these tumors varies and it is reported that 37% recur locally with 11% multiple local recurrences and 11% distant metastases (1). It is possible that benign metastatic or multicentric tumors can behave like this however in view of inflammatory fibrosarcoma's potential for local invasiveness and occasional ability to metastasize with consequent mortality it is unlikely that they are similar. Children may have better prognosis than adults, as the biological behavior is borderline or intermediate.

Immunohistochemical stains for keratin, vimentin, desmin, muscle-specific actin, smooth muscle actin, S-100protein, and KP1 (as in our case) are positive. Differential diagnoses of inflammatory fibrosarcoma are inflammatory pseudotumor, xsanthogranuloma, and malignant fibrous histiocytoma. Leiomyosarcoma also closely resembles inflammatory fibrosarcoma, but are easily differentiated by abundant eosinophilic fibrillary cytoplasm, less heavy collagenation, and also by lacking an intense chronic inflammatory component. Inflammatory myofibroblastic tumor and inflammatory fibrosarcoma have many overlapping clinical and pathological features and differ very little forming different ends of same spectrum (2).

Suggested treatment for these tumors is complete surgical excision with removal of multiple nodules, if feasible, and close follow up, as several of these tumors do not recur. Invasion or metastasizes to other sites should be treated by chemo or radiotherapy especially where complete excision is not feasible (6). Chemo and radiotherapy can also give palliation in dysphagia in esophageal involvement (3). Alpha-Interferon has been tried and shown to improve the quality of life in one child.

As an alternative effective therapy for inoperable inflammatory fibrosarcomas chemotherapy including vincristine, actinomycin-D and cyclophosphamide (VAC) could be performed. (8)

In conclusion, we conclude that inflammatory fibrosarcoma is a very rare tumor affecting all ages and various organs of the body. The clinical presentation and biological behavior are varied and pose diagnostic and therapeutic challenge. There is no common agreement on the modality of treatment though surgical excision is the treatment of choice where possible with chemo-radiotherapy or Alpha-Interferon being other options. Each case should be individualized.

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