Göztepe Tıp Dergisi 24(4):195-197, 2009 **ISSN 1300-526X**

OLGU SUNUMU *Cerrahi*

Desmoid tumors of mesentery: Mesenteric fibromatosis

Özgür EKİNCİ (*), Bülent GÜRBÜZ (**), Özlem OKUR (*), M. Rafet YİĞİTBAŞI (***), Haydar YALMAN (****), Çağrı BİLGİÇ (**)

SUMMARY

Desmoid tumors of mesentery: Mesenteric fibromatosis

Background: *Desmoid tumors are slowly growing deep fibromatosis with aggressive infiltration of adjacent tissue but without any metastatic potential.*

Case Report: *We present a 65 year old male patient with mesenteric fibromatosis of small bowel who has undergone resection of the lesion including some part of small intestine. A huge mass measuring 20*20*20 cm in size in the mesentery of ileum was observed during laparotomy. This mass included 20 cm of ileal segment, begining 60 cm proximally from ileocecal valve. He had a history of an earlier abdominal surgery for pyloric stenosis about 10 years ago. Preoperative evaluation included abdominal ultrasound and computered tomography.*

Conclusion: *Surgery has a key role in management of intraabdominal desmoid tumors.*

Key words: *Desmoid tumors, mesentery, local infiltration, intestinal obstruction*

Anahttar kelimeler: *Desmoid tümör, mezenter, lokal infiltrasyon, intestinal obstruksiyon*

Fibromatosis is the most common primary tumor of mesentery and accounts for approximately 8 % of all fibromatosis. Most cases are sporadic, but some are associated with FAP/Gardner syndrome, trauma or hyperestrogenic states [1-3]. In a patient with a genetic predisposition, tissue injury like previous operation likely to be the cause [2].

Despite their aggressive local infiltration, fibromatosis lack a metastatic potential. Most patients present with asymptomatic abdominal mass. Most mesenteric fibromatosis are large, measuring 10 cm or more [1,4]. Treatment is a multidisciplinary approach but surgery has a key role in management of intraabdominal desmoid tumors. Other methods of treatment; such as steroids, cytotoxic chemotherapy, antiestrogenic agents have variable success [2].

CASE REPORT

A 65 year old male patient with abdominal discomfort was admitted to Göztepe Education and Research Hospital. He had subocclusive symptoms like intermittent vomitting and constipation. His blood count and liver enzymes were in a normal range. His temperature was 37.2 degree centigrade. He had a history of a partial stomach resection due to pyloric stenosis which was 10 years ago. On physical examination, a huge intraabdominal mass which was about 20*15 cm was palpated in suprapubic and left lower quadrant. Abdominal USG revealed a solid mass with well-defined bordes extending from the level of umblicus to the level of bladder, measuring 20*20*15 cm. There was no sign of intraabdominal metastasis.

Multislice spiral abdominal computed tomography (CT) revealed a solid mass, measuring 18*16*19 cm between the level of umblicus and superior border of bladder. Left lateral border was irregular and had fine septations. There was no intraabdominal metastasis. In surgical exploration a jejunojejunal anastomosis (braun anastomosis) located

Geliş tarihi: 28.04.2009
Kabul tarihi: 12.08.2009
Göztepe Eğitim ve Araştırma Hastanesi 3.Genel Cerrahi Kliniği, Op. Dr.*; Asist. Dr.**; Klinik Şefi Doç. Dr.***; Şef Muavini Op. Dr.****

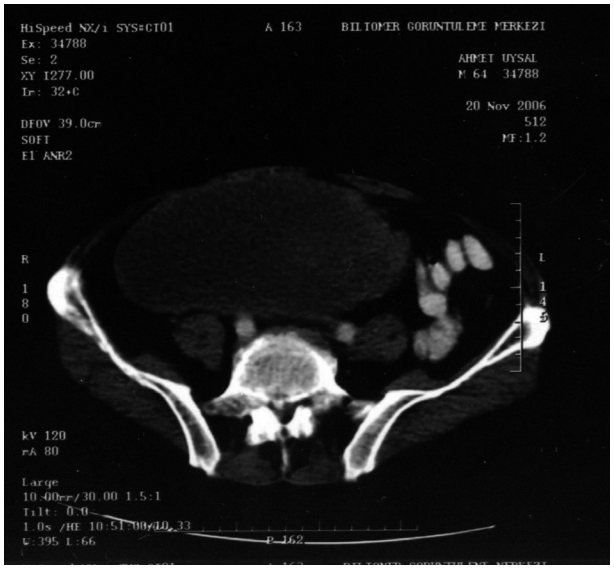

Figure 1. Computerized tomografi of the mass shows irregularity of left lateral wall .

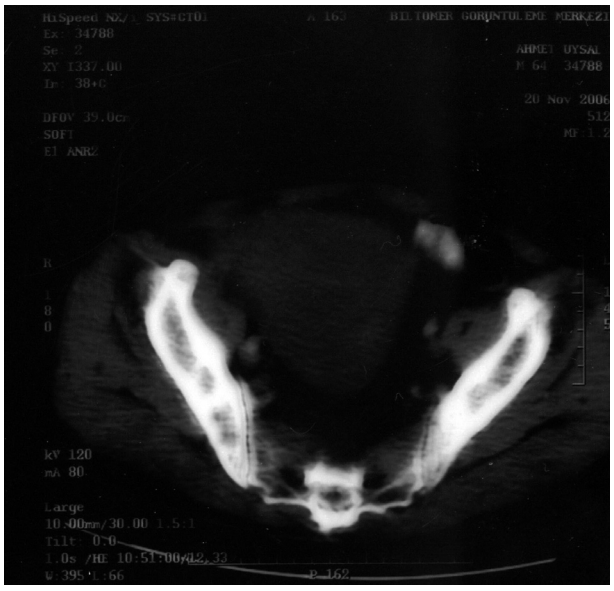

Figure 2. Computerized tomografi of the distal part of the mass.

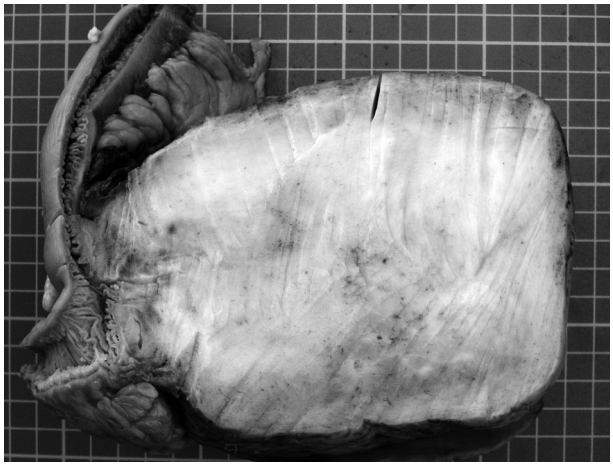

Figure 3. Macroscopic view of the mass.

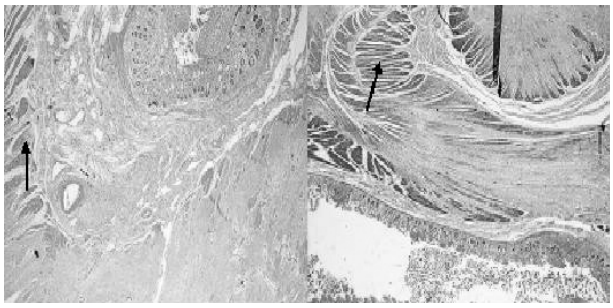

Figure 4. İnfiltration patern of mezenteric fibromatozis (arrow indicates muscle layer) (A→H&E, B→Mason-Trikrom).

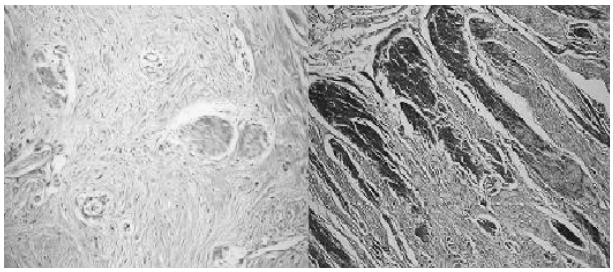

Figure 5. Elongated-spindle shaped fibroblastic infiltration in collagenized stroma (A→H&E, B→Mason-Trikrom).

approximately 15 cm distally from the ligament of Treitz was observed. A huge mass measuring 20*20*20 cm in size in the mesentery of ileum was observed. This mass included 20 cm of ileal segment, begining 60 cm proximally from ileocecal valve. The mesenteric mass was strongly adherent to serozal layer of ileum, so this segment is removed together with the mass. Ileoileal anastomosis was performed. At third day after operation, patient drank water, sixth day patient discharged from hospital. Sixth month after operation a abdominal CT performed and there was no sign of recurrence or intraabdominal metastasis.

On macroscopic pathological examination, the tumoral lesion was adherent to mesenteric side of small bowell segment, which was 35 cm in length. On cut surface, tumor had no capsule but a well-

defined borders, located on serozal side of small intestine, and had no relationship with the mucosal layer. On histopathological examination, a fibroblastic lesion, infiltrating seroza, muscularis propria and submucosa was observed. There was no inflammation, and the lesion had densely collagenous stroma, elongated spindle-shaped fibroblasts. Cellularity was mild-moderate, pleomorphism was mild, atypia was mild. No sign of hemorrhage or necrosis was observed.

DISCUSSION

Fibromatosis is the most common tumor of mesentery. Most cases are sporadic, but some are associated with Gardner syndrome, trauma or hyperestrogenic state [1,3]. In our case, the patient did not have Gardner syndrome, but had a previous abdominal operation due to pyloric stenosis. Most commonly tumors are located in the mesentery of small bowel, but they may originate from omentum or retroperitoneum. Most patients present with asymptomatic abdominal mass. Most mesenteric fibromatosis are large, measuring 10 cm or more. Complications may be caused by compression of ureter, small bowel or large intestine [1]. In our case, tumor was 20 cm and the patient had subocclusive symptoms.

CT localizes the tumor and excludes metastasis. Grossly most lesions are fairly well circumscribed. Although microscopically there is typically infiltration into surrounding soft tissues including small bowell wall, the tumor has no metastatic potential [4,5]. Histologically the lesions are composed of cytologically bland spindle-shaped or stellate cells evenly deposited in a densely collagenous stroma. Typically there is variable cellularity, with some areas showing almost complete replacement by dense fibrous tissue [1,3]. The most likely cause appears to be tissue injury in a patient with a genetic predisposition to excessive fibrous growth. Like other forms of fibromatosis, tumors have a propensity for local recurrence. Patients with Gardner syndrome, recurrence rate is 90 % compared to 12 % recurrence in patients without Gardner syndrome [1]. Local recurrence rate is high when the margins are tumor positive. Pharmacological treatment methods such as cytotoxic and non-cytotoxic chemotherapy have different success rates. Non-cytotoxic chemotherapy with hormonal agents, such as antiestrogens, response rate was 50 % among 12 of 20 patients, the others had only stabilization of their disease. Treatment with anti-inflammatory agents such as NSAIDs, the response was 57 %. Biological agents such as interferons 4 of 9 patients had response; (2 with complete response) and the remaining 5 had stabilization of their disease. Cytotoxic combination chemotherapy has a response rate between 17 and 100 %, with a median response rate of 50 % [6].

CONCLUSION

Treatment of mesenteric fibromatosis is a multidisciplinary approach. Non-surgical treatment resulted in diverse and unpredictable outcome and it is considered to be an opportunity in patients with unresectable lesions or for adjuvant therapy. Surgery has a key role in management and radical resection with clear margins is the principle treatment of this tumor entity [1,2,5].

REFERENCES

1. Weiss SW, Goldblum JR. Erzinger and Weiss's Soft Tissue Tumors. Philadelphia: Mosby, 5th ed 2008: 247-50.
2. Sakorafas GH, Nisottakis C, Peros G. Abdominal desmoid tumors. Surg Oncol 2007;16:131-42.
3. Rosai and Ackerman. Surgical pathology. Toronto. 9th ed 2004: 2390-2391.
4. Hartley JE, Chuch JM, Gupta S et al. Significance of incidental desmoids identified during surgery for familial adenomatoud polyposis. Dis Colon and Rectum 2004;47:334-40.
5. Bertagnollia MM, Morgan JA, Fletcher CDM, Rauta CP et al. Multimodality treatment of mesenteric desmoid tumors. Eur J Cancer 2008;44:2404-10.
6. Janinis J, Patriki M, Vini L, Aravantinos G and Whelan JS. The Pharmacological treatment of agressive fibromatosis: Ann Oncol 2003;14:181-190.