

Angiomyxolipoma of transverse colon - A case report

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We report the clinicopathological findings of the first case of angiomyxolipoma of the colon. A 15-year-old female presented with acute abdomen. She was admitted as a case of intestinal obstruction and a palpable mass per rectum. Intraoperatively, intussusception of the cecum into the ascending and transverse colon was found. After reduction, a large polypoidal globular and soft mass, which was pedunculated, was felt in the transverse colon. Histological examination of the resected specimen revealed a tumor mass arising from the submucosa of the colon, showing a branching filiform pattern. Angiomatous vascular proliferation was seen in the mature lipomatous tumor tissue with areas of myxoid stroma. To the best of our knowledge, this is the first report of the clinicopathological presentation of angiomyxolipoma of the colon and gastrointestinal tract.

Key words: Angiomyxolipoma, transverse colon, intussusception

Transvers kolonun anjiomiksoliomu: Olgu sunumu

Kolonun anjiomiksoliomu olan ilk vakanın klinikopatolojik bulgularını rapor ediyoruz. 15 yaşında bir kız hasta akut karınla başvurdu. İntestinal obstrüksiyon ve rektumda palpe edilebilen bir kitle olarak yatırıldı. Ameliyat sırasında, çekumun çıkan ve transvers kolona intussusepsiyonu bulundu. Redüksiyon sonrasında transvers kolonda büyük polipoid küresel saplı bir kitle hissedildi. Rezekte edilen materyalin değerlendirmesinde kolonun submukozasından köken alan, dallanan filiform patern gösteren bir tümör saptandı. Matür lipomatöz tümör dokusunun miksoid stroma ve anjiomatöz vasküler proliferasyon içerdiği görüldü. Bildiğimiz kadarıyla bu, kolon ve gastrointestinal kanalın anjiomiksoliomunun klinikopatolojik prezentasyonunun ilk raporlandığı ilk yazıdır.

Anahtar kelimeler: Anjiomiksoliom, transvers kolon, intussusepsiyon

INTRODUCTION

Lipoma is the most common neoplasm of the body, and several subtypes of lipoma have been described. These have been classified according to their location and the presence of other tissue elements. Angiomyxolipoma (AML) is a recently identified very rare variant of lipoma that consists of an admixture of adipose and myxoid elements with numerous dilated vascular structures, as first described by Mai et al. in 1996 (1). It should be differentiated from other subtypes of benign and malignant lipomas. Here, we report the case of a 15-year-old female who presented with acute intestinal obstruction. The histopathologic findings after re-

section of the mass from the colon showed alternating nests of myxoid and adipose tissue containing dilated blood vessels, which was consistent with AML.

CASE REPORT

A 15-year-old female presented with pain with distention of the abdomen for five days. She gave a history of straining at defecation due to constipation for the last six months. Per abdomen examination showed tenderness and distention throughout the abdomen. Bowel sounds on auscultation were absent. The rectal examination revealed a

mass protruding in the rectum from above. The upper extent of the mass could not be reached. Blood investigation showed neutrophilic leukocytosis of moderate grade. X-ray examination showed features of intestinal obstruction and multiple air-fluid levels.

On opening the abdomen, a colocolic intussusception was found. The intussusception was reduced completely, and yet a mass could be felt filling the transverse, descending and sigmoid colon (Figure 1, the mass was removed via the transverse colon). Grossly, no lymph nodes were found to be enlarged. A segmental resection of the colon with an end-to-end anastomosis was performed, bearing in mind the young age of the patient. The postoperative period was uneventful. The patient has been followed for the last nine months with no recurrence.

Gross Examination

The specimen of the large intestine measured 8 cm in length with a tumor mass removed via a 4 cm incision in the middle. On opening the segment, the tumor was seen attached to the colonic mucosa by a broad stalk and projected out for about 15 cm in a branching filiform pattern. The mass measured about 4 cm in width at the base and about 2 cm at the middle of each of the four finger-like branches (Figure 2, the formalin-fixed specimen). It was pale yellow in color and soft in consistency. The cut section revealed tortuous blood vessels branching in the lipomatous greyish yellow soft tissue.

Microscopy

Histopathological examination revealed a tumor mass arising from the submucosa of the colon, showing a branching filiform pattern. On microscopy, mature lipomatous tumor tissue with areas of myxoid stroma composed of myxoid tissue with spindle and stellate cells, without significant atypia or neoplastic cells, was found. Scattered throughout the myxomatous background were several blood vessels of varying sizes. The section from the colon revealed congestion with mild inflammation (Figure 3, microscopic findings and inset showing high power field).

A histological diagnosis of AML arising from the submucosa of the large intestine was made. Immunohistochemical localization of CD34, vimentin, S-100, and smooth muscle actin (SMA) showed positive reactions (Table 1).

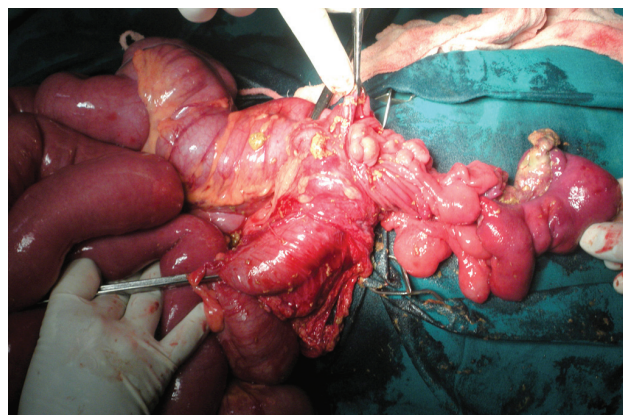


Figure 1. Intraoperative picture of the mass arising from the submucosa of the transverse colon, which was removed via the colon lumen by a small incision. This mass, approximately 20 cm long, traversed from the transverse colon to the rectum, causing intussusception.



Figure 2. The formalin-fixed specimen, 15 cm x 4 cm, with a broad stalk projected out as a branching filiform pattern.

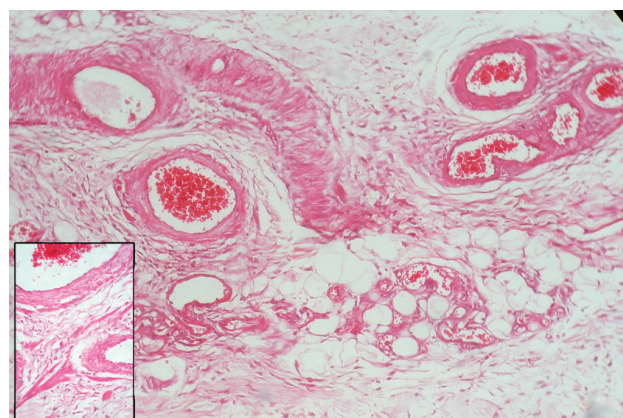


Figure 3. Microscopic picture of the angiomyxolipoma. On microscopy, mature lipomatous tissue with areas of myxoid stroma is seen.

Table 1. Clinical features and immunohistochemical study of angiomyxolipoma in the literature

Pt. No.	Ref. No	Sex/Age (yrs.)	Location	Duration	Immunohistochemistry					
					CD34	Vimentin	S-100	SMA	Desmin	Hmb45
1.	1	M/32	Spermatic cord	3 mos.	NP	+	(l)	+	(v)	-
2.	2	M/57	Scalp	NI	+	(s)	NP	+	(v)	NP
3.	3	M/50	Back	3 yrs.	+	(v, s)	+	(s, l)	+	(l)
4.	4	F/60	Thigh	4 mos.	+	(v, s)	+	(s, l)	+	(l)
5.	5	M/66	Scalp	NI	+	(v, s)	+	(s, l)	+	(l)
6.	6	M/44	Arm	7 yrs.	+	(v, s)	+	(s, l)	+	(l)
7.	6	M/57	Wrist	2 yrs.	+	(v, s)	+	(s, l)	+	(l)
8.	7	M/43	Subungual area	1 yr.	NP	NP	NP	NP	NP	NP
9.	8	M/38	Gluteal area Extremity	3 yrs.	+	(v, s)	+	(s, l)	+	(l)
10.	9	M/69	Hip	3 yrs.	+	(v, s)	+	(s, l)	+	(l)
11.	PC	F/15	Transverse colon	--	+	(v, s)	+	(s, l)	+	(l)

SMA: Smooth muscle actin. M: Male. NP: Not performed. S: Spindle cells in myxoid area. L: Lipocytes in lipomatous area. V: Vessel walls. NI: Not informed or not specified. F: Female. PC: Present case.

DISCUSSION

A number of variants of lipoma have been described, including synovial, parosteal, intraosseous, lumbosacral, and thymolipoma, as well as spindle cell/pleomorphic, chondroid, angio-, fibro-, myxo-, chondro-, osteo-, and myolipoma. AML is a very rare benign neoplasm with characteristic histopathologic and immunohistochemical features. AML is a rare variant of lipoma, first described by Mai *et al.* in 1996 (1).

Ten cases of AML, or vascular myxolipoma, have been described previously (Table 1) (1-9). The patients' ages ranged from 32 to 69 years, averaging 51.6, and most were male. All the cases to date have been treated surgically with no recurrence. The present case is the youngest to be reported (15 years). The location has generally been in the subcutaneous tissue, although one was located on the spermatic cord and one in the subungual region. The present case is the first case to be located in the gastrointestinal tract.

The histopathologic features of AML are characteristic and include an admixture of a myxoid area with relatively few cells, mature adipose tissue and numerous thin- and thick-walled vessels. On electron microscopy, Mai *et al.* (1) identified spindle cells with fat vacuoles (so-called 'preadipocytes') in the transitional areas between the myxoid and lipomatous components.

A cytogenetic study of AML revealed chromosomal aberrations involving translocations t(7;13)(p15;q13) and t(8;12)(q12;q13), which are similar to those found in ordinary lipoma, spindle-cell/pleomorphic lipoma, and myxoma (4), but the primary origin of the AML remains to be determined (9).

Immunohistochemical studies in the present case showed that the spindle cells of the myxoid areas expressed vimentin and CD34, but did not express SMA, desmin, or S-100 protein. The mature adipocytes were immunoreactive for S-100 protein and the blood vessels were positive for vimentin and SMA (9).

The main differential diagnosis for AML is myxoid liposarcoma, as they share many histological features, and myxoid liposarcoma has to be ruled out for prognostic and therapeutic reasons. Similar to AML, myxoid liposarcoma also has a myxoid stroma, lacks mitosis, may be well circumscribed by fibrous tissue, and lacks significant cellular atypia. However, the data supporting a benign tumor in this case include the absence of lipoblasts and absence of a plexiform capillary pattern (3).

In conclusion, AML is a very rare benign adipose tumor, and as in all cases described, including ours, surgery is the curative treatment, with no known recurrences.

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