

Abdominal Aggressive Fibromatosis: Diagnostic and Therapeutic Features

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ABSTRACT

Desmoid tumours are rare soft tissue tumours developed from myofibroblasts. They usually infiltrate into adjacent muscle bundles and are characterised by the absence of metastatic potential. However, a high local aggressiveness is seen with a high rate of recurrence after surgical excision. We hereby attempt to present a series of patients with desmoid tumours and review treatment options and management strategies. Ten tumours occurred in the abdominal wall, five in the mesentery, and one was presacral tumour. The mean diameter of the tumour was 6 cm (range, 2.3-40 cm). Associated Familial Adenomatous Polyposis (FAP) was found in five patients. The margin of surgical resection was healthy in five patients. During follow up period, two patients returned with recurrence of tumours.

Keywords: Desmoid tumours, Oncology, Surgery

Desmoid tumours, also known as aggressive fibromatosis, are rare soft tissue tumours. They represent 3.5% of fibrous tumours and less than 0.03% of all tumours. They occur sporadically or as part of FAP. They are characterised by local malignancy with a high tendency to recur [1,2]. Aim of this series was to report a 10-years experience of 16 desmoid tumours and review some facts with symptoms, investigation, and therapeutic aspects of this rare entity.

CASE SERIES

From the hospital case record, a total of 16 consecutive desmoid tumours (8 patients) were identified over a duration of 10 years from January 2005 to December 2015 [Table/Fig-1].

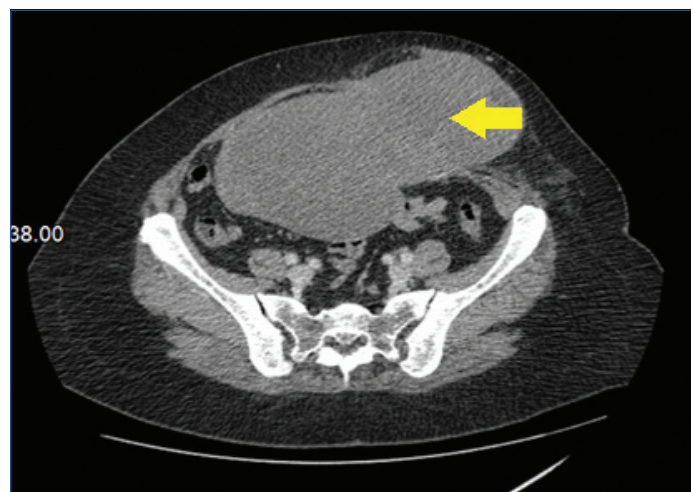
Case	Age (years)	Gender	Number of tumours	Tumour site	Tumour size (cm)	FAP	Treatment	Recurrence
1	42	M	5	Abdominal wall (4)	5	+	Tamoxifen	Stable
				Mesenteric (1)	3			
2	17	F	2	Abdominal wall	8	+	NSAID+ Tamoxifen	Stable
				Mesenteric	6			
3	39	M	1	Mesenteric	5	+	Surgery	-
4	58	M	1	Presacral	2.3	+	Tamoxifen	Stable
5	30	F	1	Abdominal wall	5	-	Surgery	-
6	25	F	1	Mesenteric	7	-	Surgery	-
7	53	F	2	Abdominal wall	6	-	Surgery	+ (4 years)
				Abdominal wall	5			
8	23	F	3	Abdominal wall	5	+	Surgery	+ (2 years)
				Abdominal wall	7			
				Mesenteric	40			

[Table/Fig-1]: Demographic data of treated patients.

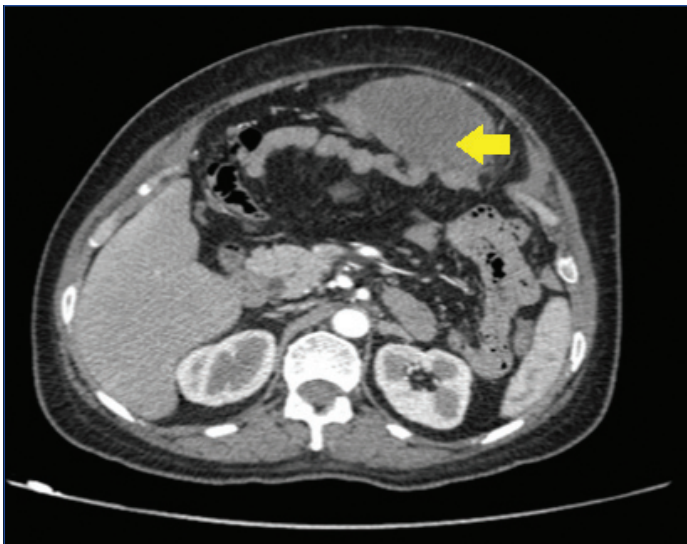
F: Female; M: Male; FAP: Familial adenomatous polyposis; NSAID: Nonsteroidal anti-inflammatory drugs

The mean age of patients was 35±14.6 years (17-58 years). The male to female ratio was 3:5. All patients underwent a CT-scan [Table/Fig-2-4] and a colonoscopy. In three cases, fibromatosis was isolated. Associated FAP was found in five patients. Polyposis was discovered by colonoscopies in two patients. In the other three patients, fibromatosis appeared after surgery for FAP with an average delay of two years after surgery. The desmoid tumours were multiple in 4 (50%) cases. A total of 10 tumours occurred in the abdominal wall [Table/ Fig-5], five in the mesentery [Table/ Fig-6,7], and one was presacral tumour. The mean diameter of the tumours was 6 cm (range, 2.3-40 cm). Surgical resection with wide surgical margins was the treatment of choice whenever possible. Excised specimens were all subjected to pathological evaluation.

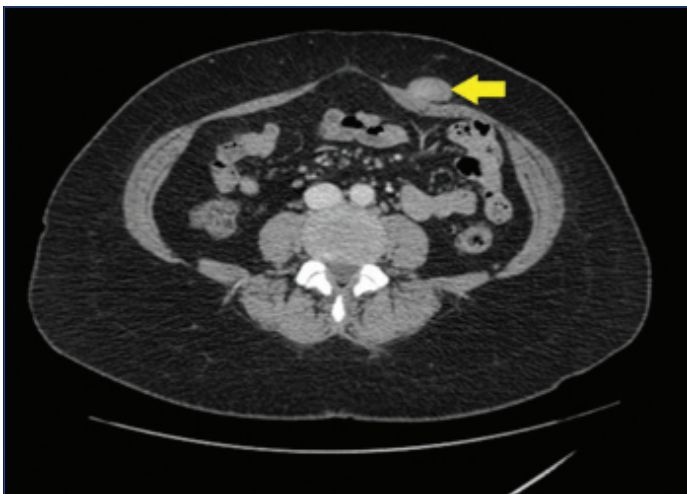
In two cases, treatment given was tamoxifene and/or nonsteroidal anti-inflammatory drugs. The margin of surgical resection was healthy in five patients. Soft tissue reconstruction following resection was required in two cases. No patient had surgery-related complications. During follow up (4.25 years) period, two patients returned with recurrence of tumours. Because of the nonresectable character of the tumour, they had treatment with nonsteroidal anti-inflammatory drugs and tamoxifen.



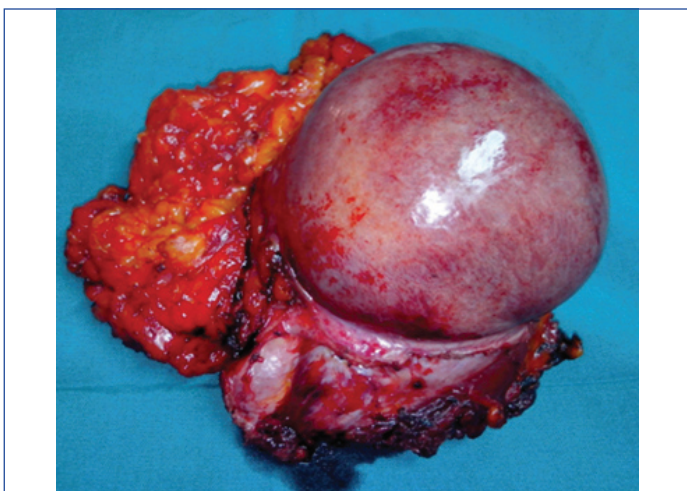
[Table/Fig-2]: Axial CT-scan showing abdominal desmoid tumour.



[Table/Fig-3]: Axial CT-scan showing abdominal desmoid tumour.



[Table/Fig-4]: CT-scan showing a desmoid tumour of the rectus abdominis muscle.



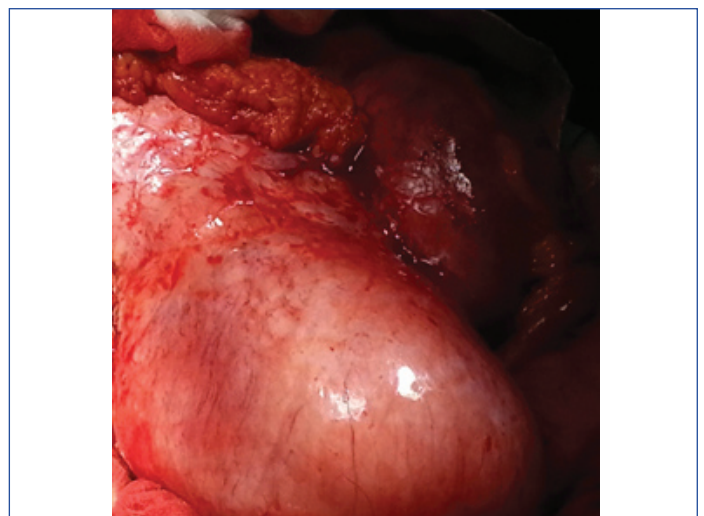
[Table/Fig-5]: Gross appearance of a resected desmoid tumour.

DISCUSSION

Desmoid tumours are soft tissue tumours which develop from myofibroblasts and infiltrate into adjacent muscle bundles. They are proliferations of well-differentiated fusiform conjunctive cells associated with abundant clusters of collagen. They are characterised by the absence of metastatic potential. However, a high local aggressiveness is seen with a high rate of recurrence after surgical excision. They occur sporadically or associated to FAP [1]. Overall annual incidence is 2.4 to 4.3/100,000 inhabitants. They can appear at any age with a predilection for the third and fourth decades of life. In the present case series, the mean age was



[Table/Fig-6]: Voluminous intraperitoneal desmoid tumour of the mesentery.



[Table/Fig-7]: Intraoperative view of a voluminous desmoid tumour.

35 years. Female predominance had been reported [2], consistent with the present case series (3 men and 5 females). Frequency in the FAP varies according to the authors between 4 to 32% [3,4]. In the present case series, the association of FAP was found in five patients because the center to which the patients presented is a reference center in the treatment of polyposis.

According to the anatomical site, desmoid tumours can be divided into extra-abdominal fibromatosis (58%); intra-abdominal fibromatosis (8-15%) and fibromatosis of the abdominal wall [1,2].

Three aetiopathogenic factors are often incriminated: trauma, hormonal influence and genetics [2]. Macroscopically, it appears as a well circumscribed hard mass, elongated in the axis of the muscle. Histologically, there is proliferation of fusiform cells corresponding to fibroblasts and myofibroblasts arranged in long divergent beams without nuclear anomaly. The main distinguishing feature with fibrosarcoma is the absence of necrosis. However, it is essentially the immunohistochemistry which makes it possible to plead for the myofibroblastic origin of these tumours. It also makes it possible to differentiate desmoid tumours from Gastrointestinal Stromal Tumours (GIST) by showing a negativity of CD-34 and S-100, whereas they are positive in GIST. Similarly, the positivity of anti-desmin and anti-alpha-actin antibodies make it possible to eliminate the fibrosarcoma for which it is generally negative [5].

Desmoid tumour has no specific symptoms. It is a firm, painless lesion that usually progresses slowly. Its size varies from 1 to 30 cm. Desmoid tumours are often hypoechogenic on ultrasound.

They usually have a homogeneous appearance with a tissue density on CT scan. An hypo or iso-intense aspect in T1 and a variable signal in T2 are usually noted on MRI. This examination makes it possible to well appreciate the extension and tumour resectability [6,7].

Positive diagnosis is made possible by histological examination. We must search for a FAP with a systematic colonoscopy [8]. As for the present patients, a polyposis was discovered by colonoscopies in two patients.

Treatment is not strictly codified [9]. Different therapeutic modalities can be used: surgical resection, radiotherapy, anti-inflammatory agents, hormonal treatment and chemotherapy. Surgery with healthy resection margins remains the ideal therapeutic option for these tumours. The resection must be complete without being mutilating. Adjuvant treatments, dominated by hormonal therapy and radiotherapy, are indicated in the event of recurrence or an unresectable tumour [10] as in two cases of the present case series.

CONCLUSION

Desmoid tumours are rare mesenchymal neoplasm with local aggressiveness. Despite the absence of metastatic potential, these tumours remain doubttable due to their invasive potential

and their high rate of recurrence. Although, histologically identical to other desmoid tumours, abdominal forms are particular by their locations, occurrence circumstances and therapy. Treatment is not strictly codified. Surgery remains the reference treatment. In case of inoperable or recurrent lesions other therapeutic alternatives remain possible with modest efficiency.

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FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Sep 05, 2017**
Date of Peer Review: **Nov 09, 2017**
Date of Acceptance: **Nov 23, 2017**
Date of Publishing: **Apr 01, 2018**