International Journal of Innovative Research in Medical Science (IJIRMS)
Volume 08, Issue 06, June 2023,
https://doi.org/10.23958/ijirms/vol08-i06/1684

Case Series

Desmoid Tumors About 3 Cases and Revue of Literature

O. Elatiqi *[1], I. Zinedine [2], O. Aitbenlassal [2], L. Idelkhir [2], MD Elamrani [3], Y. Benchamkha [4]

[1]Assistant Professor, Plastic Surgery Department, Mohamed VI University Hospital, Marrakech, Morocco.
[2]Doctor, Resident, Plastic Surgery Department, Mohamed VI University Hospital, Marrakech, Morocco.
[3]Professor, Plastic Surgery Department, Mohamed VI University Hospital, Marrakech, Morocco.
[4]Professer, Head Master of Plastic Surgery Department, Mohamed VI University Hospital, Marrakech, Morocco.

*Corresponding author: O. Elatiqi; *okelatiqi@yahoo.fr*

Received 07 April 2023; Accepted 25 May 2023; Published 05 June 2023

Abstract

Desmoid tumors or aggressive fibromatosis are rare, recurrent, non-metastasizing tumors, developing from muscle fascias and aponeuroses, before surgery was the main treatment, with the latest recommendations and in view of the high risk of recurrence, the treatment consists of close monitoring, surgery is indicated if there are complications related to the location of the lesion.

We report three observations of three young patients with desmoid tumors, one in abdominal location measuring 20/15cm in diameter invading the muscular wall and the peritoneum, the second at the level of the lower limb in the posterior face of the thigh and the right buttock measuring 30cm/10cm in diameter and the third one is abdominal too measuring about 20cm/15cm of diameter invading the peritoneum in a patient operated for rectal adenocarcinoma with familial adenomatous polyposis. The reason for consultation in the 3 patients was pain.

After multidisciplinary consultations (surgeons, oncologist and radiotherapists) it was decided to perform surgical excision of the tumors, the follow-up is estimated at 25 months for the first abdominal location, 10 months for the location in the lower limb and 2 months for the third case, without noticing any recurrence, no adjuvant treatment was offered to the patients only close surveillance.

Surgery for desmoid tumors is a double-edged sword given the high risk of recurrence which can only be decided after multidisciplinary consultations.

Keywords: ***Desmoid tumors, familial adenomatous polyposis, female, surgery, "wait and see".***

Introduction

Desmoid tumors or desmoid fibromatosis, are rare tumors, witch Arise from fascia's and muscle aponeuroses, they are slow growing, do not give metastasis but locally aggressive, the incidence is very rare in general population 5 to 6 cases per million a year, 5% to 10% of cases are associated with familial adenomatous polyposis [1],previously surgery was the primary standard treatment, in recent years, close monitoring and surgery in case of complications related to its location are the Last recommendations [2], we report 3 cases of 3 female patients operated in our department for complication related to the location of the tumor.

First Case

28 years old female patient, single, with no background, presents a left gluteal mass that appeared 20 months ago in October 2019, gradually increasing in size reaching the lower 1/3 of the posterior face of the left thigh, painful with lameness (**Image 1**)

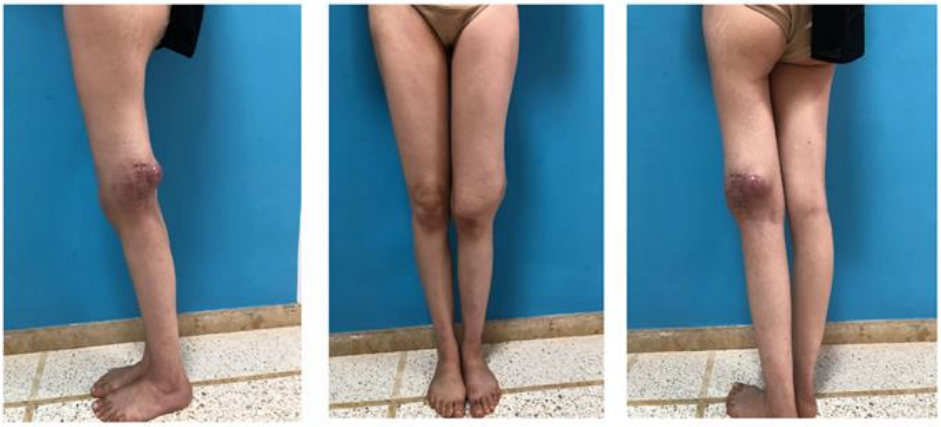

Image 1: Preoperative aspect of the tumor

An IMR was performed demonstrating a polylobed mass of the gluteal soft tissues extended to the posterior and lateral surfaces of the left thigh infiltrating the gluteal muscles and the muscles of the posterior surface of the thigh, arriving in contact with the greater sciatic nerve without edging of separation. (**Image 2**)

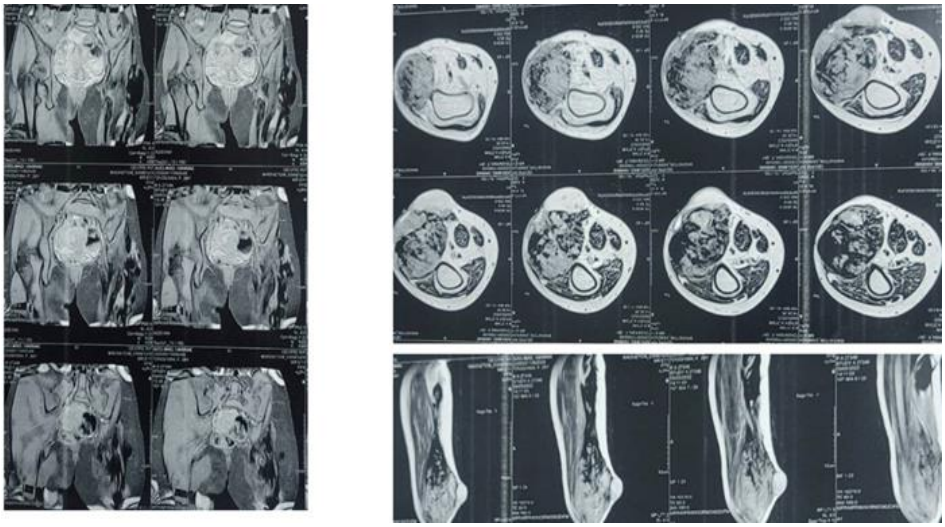

Image 2: MRI of the tumor

The diagnosis of desmoid tumor was confirmed by a biopsy.
The surgery was decided after a multidisciplinary meeting between onco-radiotherapists, traumatologists and plastic surgeons and in view of the invalidity caused by the tumor the surgical excision of the tumor was indicated, initially an amputation was proposed, but the patient refused, the medical team proposed surgical excision of the tumor and if recurrence an amputation, the patient accepted this proposal.

The surgical team consisted of plastic surgeons and traumatologists, the entire tumor was removed, note that the greater sciatic nerve was preserved. (**Image 3**)

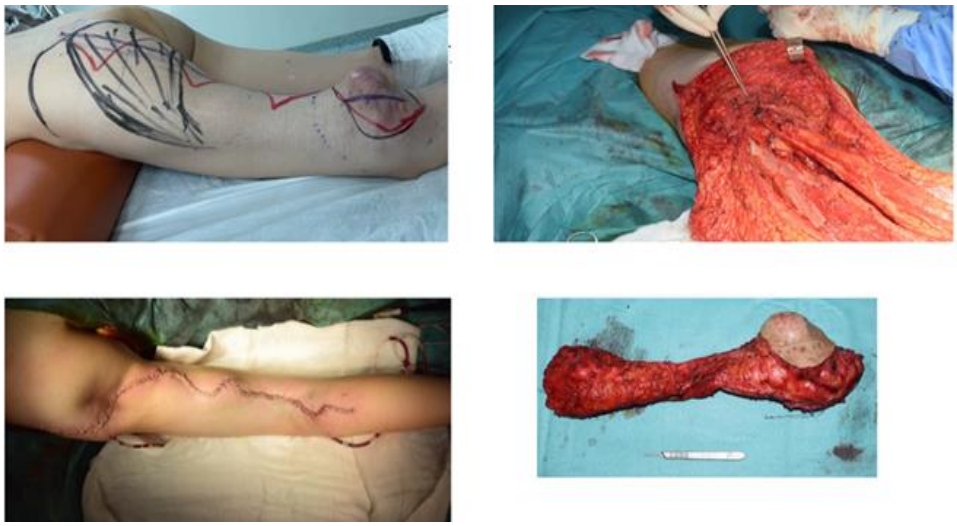

Image 3: Per operative aspect chowing preservation of greater sciatic nerve

Postoperative follow-up was simple, the patient was declared discharged 2 days after surgery, rehabilitation with the wearing of compression stockings was prescribed 10 months after the surgery no recurrence was observed. (**Image 4**)

Image 4: Image after 10 months of evolution

Second Case

31 years old female patient, married, one child a child born by caesarean section, no other backgrounds, has been complaining for 4 months, a mass in the right iliac fossa rapidly increasing in size and painful, clinically it´s a painful mass of 12cm / 8cm mobile with the superficial plane, and fixe to the deep plane, Biopsy: histological aspect evoking a desmoid tumor. (**Image 5**)

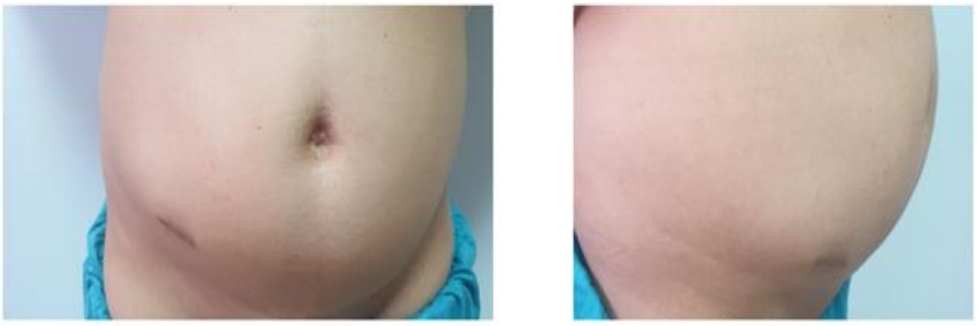

Image 1: Preoperative aspect of the tumor

Abdominal CT: mass of the abdominal wall at the expense of the right internal oblique muscle without fatty border of separation pushing back the peritoneum, evoking more a desmoid tumor (**Image 6**).

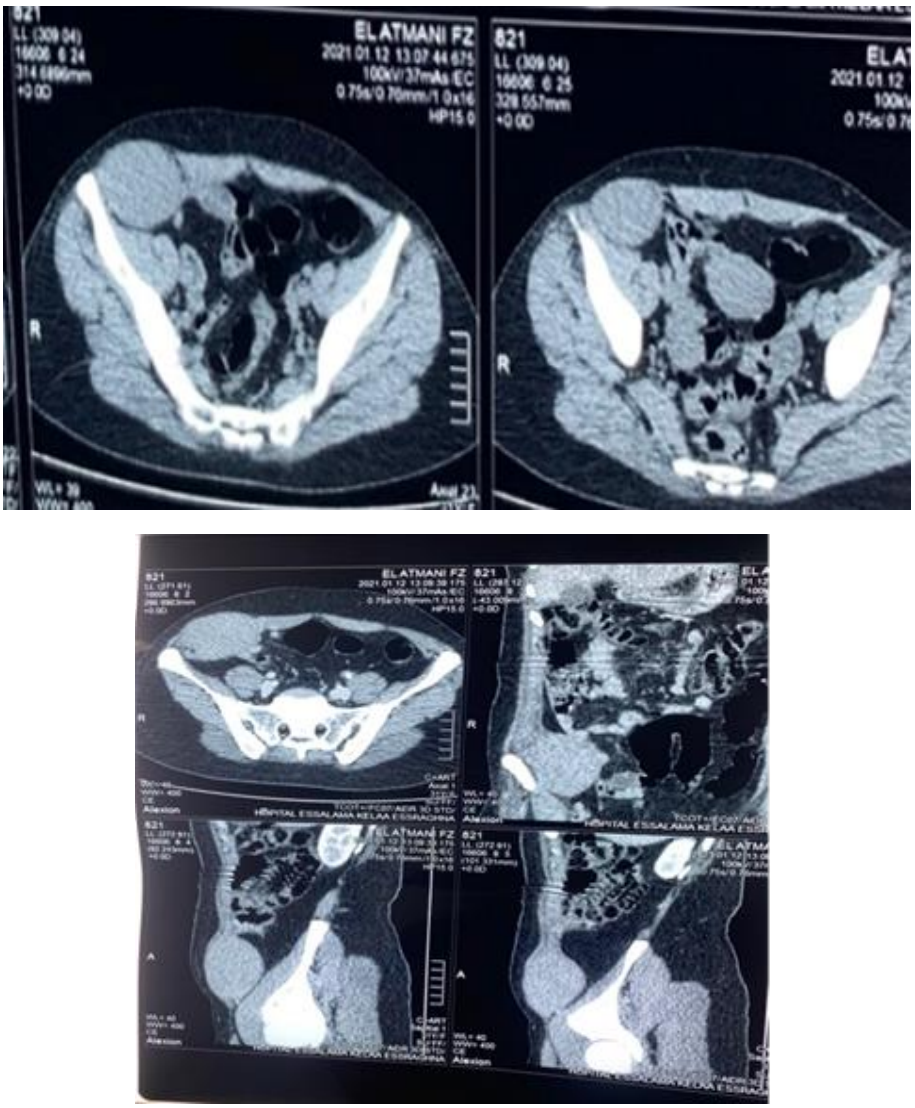

Image 6: Abdominal CT

After a multidisciplinary meeting between onco-radiotherapists, general surgeon, and plastic surgeons and in view of the pain and how quickly the tumor grows, the surgery was decided.

The surgical team consisted of plastic surgeons and general surgeons, the procedure was excision in one piece of the tumor removing a part of transverse muscle and peritoneum invaded by the tumor, closure of the peritoneum, placement of a two-sided plate, then closure of the cutaneous plane. (**Image 7**)

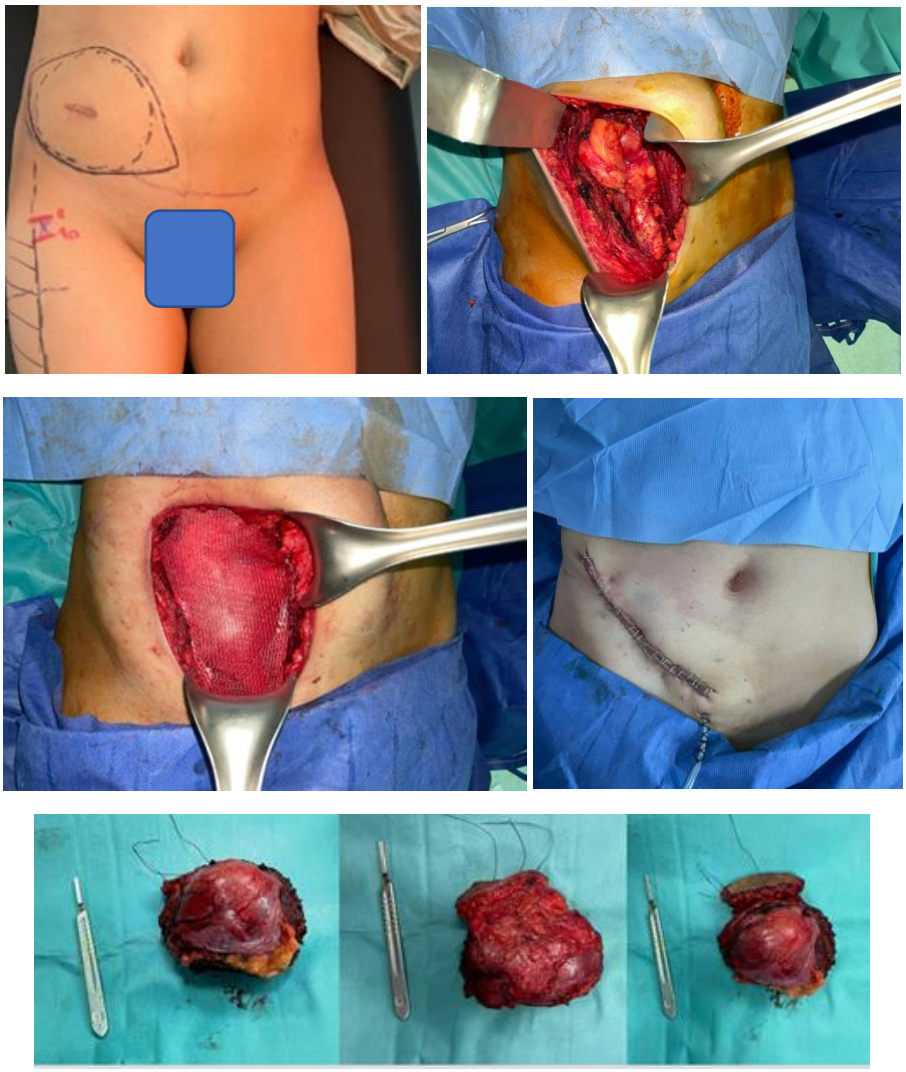

Image 7: Preoperative images

The postoperative follow-up was simple, patient declared discharged on day 5.

For 25 months the patient has been under regular surveillance and has not presented any recurrence. (**Image 8**)

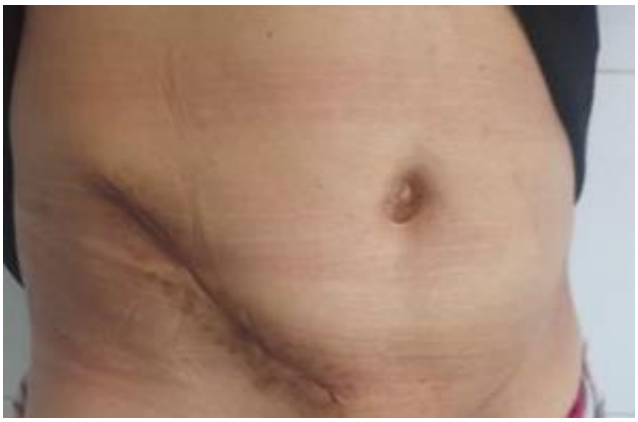

Image 8: Aspect after 25 months

Third Case

28 years old female patient, having familial adenomatous polyposis, operated for rectal adenocarcinoma in 2018, mother, sister and brother who died of colon cancer with familial adenomatous polyposis, after surgery patient noted a superficial para-umbilical mass gradually increasing in size. (**Image 9**)

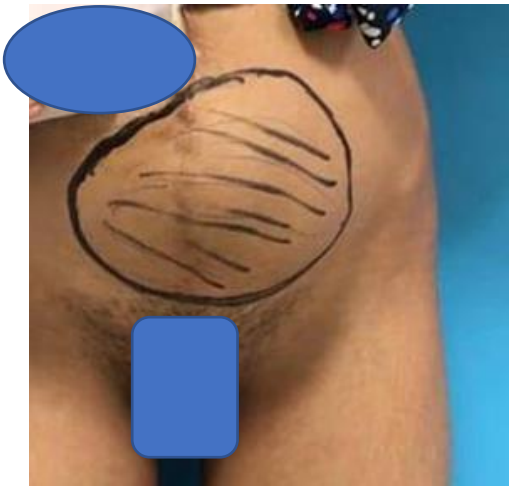

Image 9: Before surgery

Biopsy made in 2019: desmoid tumor.
An abdominal scan was performed showing the presence of a median paraumbilical mass of 19 cm coming into contact with the intestines the iliac vessels, the bladder and the uterus without a fatty separation line.

After a multidisciplinary meeting between onco-radiotherapists, general surgeon, and plastic surgeons, the patient underwent 12 sessions of chemotherapy (imatinib), the evolution was marked by a reduction in size of the tumor by approximately 17%, then the patient was scheduled for surgery. (**Image 10**)

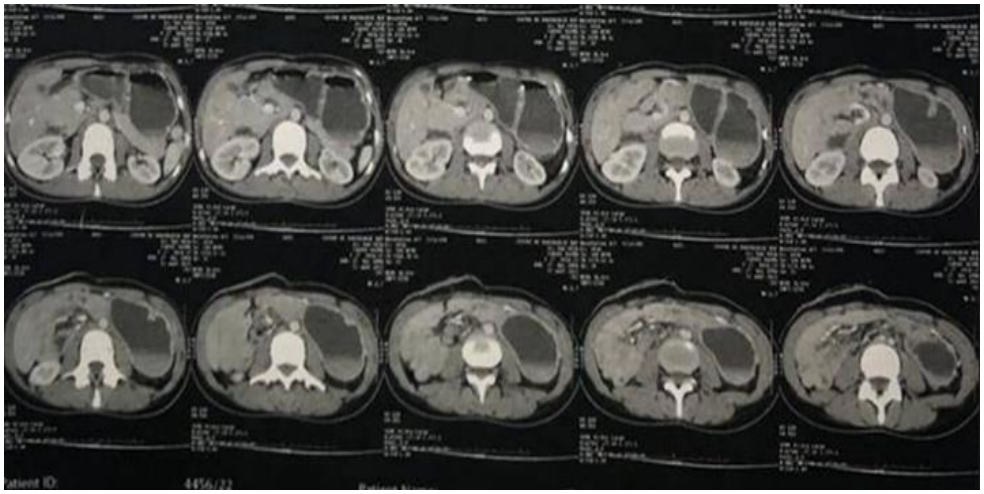

Image 10: Abdominal scan

The surgical team consisted on plastic surgeons and general surgeons, the procedure was excision in one piece of the tumor removing a part of rectis abdominis muscle and peritoneum invaded by the tumor, closure of the peritoneum, placement of a two-sided plate, then closure of the cutaneous plane. (**Image 11**)

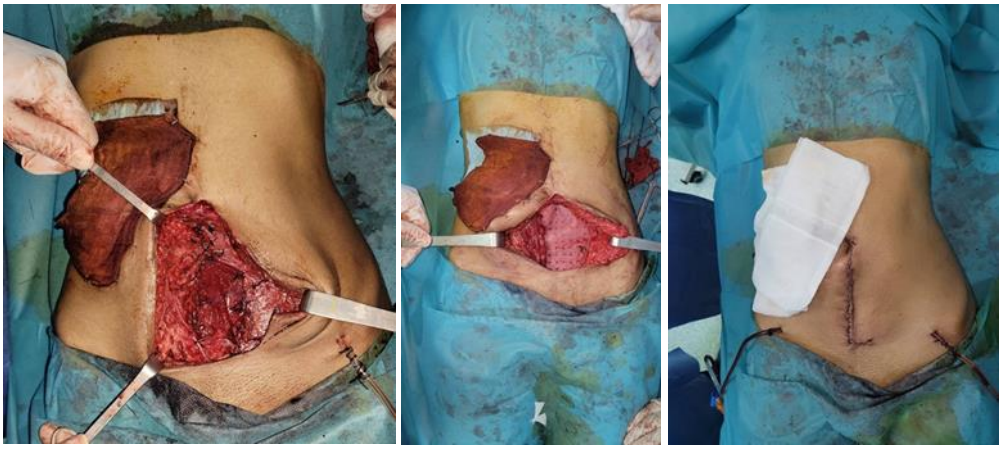

Image 11: Preoperative aspect

The postoperative follow-up was simple, patient declared discharged on day 5.

2 months post operative, patient is under very close and regular surveillance.

Discussion

Desmoid tumors or fibromatosis tumors are a rare monoclinal fibroblastic proliferation, clinically unpredictable (slow or quick growth), with tendance to recurrences bat never give metastasis, the

incidence is about 5 to 6 cases per million of population per year, the peak age is between 30-40 years, much frequent in females than men, 5 to 10% arises in the context of familial adenomatosis polyposis FAP [1,3].

Desmoid tumors affect soft tissue in all sites, classified as intra-abdominal (abdominal wall, peritoneum, mesentery...) or extra-abdominal (trunk, shoulder, neck, limbs...) [1,3-5].

Risk of recurrences is much higher with patients with FAP (44% vs 25%) and mortality also (14% vs 0%) [4].

Desmoid tumors are caused by multifactorial etiologies: genetic, endocrine, physical factors [6,7] pregnancy specially in sporadic cases (21% vs 0% in FAP) and previous surgery more in FAP cases than the sporadic ones (64-83% vs 13% respectively)[7-9].

The tumor growth is variable it can be a very aggressive growth form (10%), or a prolonged stabilized one (50%) or a spontaneous resolution (10%) [2,10,11].

The treatment of Desmoid tumors is complex, including surgery and medical treatment as radiotherapy, systemic treatment like Nonsteroidal Anti-Inflammatory Drugs, hormone therapy (tamoxifene or toremifene), chemotherapy (vinblastine or vinorelbine) in one of our cases imatinib was prescribed and we have noticed a reduction of the size of the tumor.

Before 2000 surgery was the mean treatment based on resection of the tumor with wide margins [12,13] in the last decades multiple studies showed a high risk of recurrences however was the margins [14], that's why active treatment including surgery or medical treatment is indicated only in certain cases: intra-abdominal localization with complications specially patients with FAP and desmoid tumors, and for large tumors in location where progression could become life-threatening [2], in our 3 cases surgery was indicated in front of complications in relation with location, growth and pain caused by the tumor.

Based on the last recommendations close observation is now considered as an approach "wait and see approach" especially for asymptomatic patients. [15,16].

Conclusion

Desmoid tumors are very aggressive, if they are asymptomatic the best thing to do is to observe them "wait and see" approach, this can be one or two months after diagnosis and every 3 pr 6 months, only treatment is indicated for complicated cases.

Conflict of interest

The author declares no conflict of interest regarding the publication of this paper.

Funding

The study is not funded

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