

## Desmoid type fibromatosis of the anterior abdominal wall in pregnancy: A diagnostic dilemma

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### Abstract

Desmoid tumors of the anterior abdominal wall are rare mesenchymal tumors with an unclear etiology. Pregnancy and other hyper estrogenic states are considered as an independent risk factor for sporadic cases. The literature available regarding the assertion of various risk factors, diagnostic approach, treatment modalities are sparse. There are only few anecdotal case reports regarding occurrence of desmoids in pregnancy. We are presenting a 32 year woman at term pregnancy with an abdominal desmoid type fibromatosis resected during caesarean section which was initially misdiagnosed as lower segment uterine fibroid. We are reporting this case for its rarity and the diagnostic and therapeutic challenges it poses to the treating clinician.

### Keywords

desmoids; pregnancy; caesarean

### Abbreviations

FAP: Familial adenomatous polyposis; APC: Adenomatous polyposis coli

### Introduction

Desmoid tumour of the anterior abdominal wall is an uncommon neoplasm. It can occur sporadically or as a part of an inherited syndrome such as Familial adenomatous polyposis (FAP) [1] due to beta catenin gene mutation or Adenomatous polyposis coli (APC) gene [2]. Desmoid tumors occur with a frequency of 2.4-4.3 cases per million per year [3]. The data regarding the incidence in pregnancy is very limited. Typically, sporadic abdominal wall desmoid tumors occur in young women during pregnancy or within a year of childbirth. These indeed elucidate the regulatory role of estrogen in the pathophysiology of the development of desmoids. Deep fibromatosis involving the musculoaponeurotic layer has a relatively rapid growth rate and a higher rate of local recurrence but systemic metastasis is extremely unusual [1].

### Case Presentation

A 32 year old woman presented in labour room at 38 weeks of gestation in labour with the antenatal diagnosis of lower uterine segment fibroid. Her previous obstetric history included a spontaneous abortion at 12 weeks period of gestation and a previous term caesarean section. There was no evidence of presence of fibroid in the previous pregnancies. In the index pregnancy, she was found to

have a hypo echoic mass above the bladder suggestive of fibroid uterus measuring 9\*8\*5 cm during her antenatal ultrasonography along with a single intra uterine pregnancy. On physical examination there was a fixed 10\*10 cm mass occupying the supra pubic area .During labour she had achieved full dilatation but there was no descent of fetal head in spite of good uterine contractions; therefore we proceeded with caesarean section using pfannenstiel incision considering the impacted fibroid as the cause of non descent. On incising the rectus sheath, we encountered a 13\*8\*5 cm solid tumor infiltrating into the rectus muscle and aponeurosis undermining the rectus sheath (Figure 1). The tumor was clearly delineated and resected completely followed by the access into the peritoneal cavity and delivery of the fetus by lower segment caesarean section. There was no other associated gross intra-abdominal pathology. She was willing for concurrent sterilisation so tubal ligation was done after obtaining informed consent. The defect in the abdominal wall was reconstructed. On gross examination of the specimen the outer surface was capsulated with areas of congestion (Figure 2). Cut surface was grey white, homogenous, firm with whorled appearance with no cystic changes. Microscopic examination revealed spindle shaped fibroblasts in a collagenous stroma suggestive of desmoid tumor. The post operative period was uneventful. She was discharged from hospital after suture removal on day 7 as per the treating clinician's discretion.

## Discussion

Desmoid tumors are rare tumors presenting in pregnancy posing a diagnostic challenge. This presentation also infers the occurrence of the tumor in the milieu of estrogen excess establishing the risk factor [4]. Temporal association between development of desmoids and an antecedent history of abdominal trauma or operation has also been noted. The present clinical manifestation was associated with a previous caesarean section. Desmoid tumors usually present with a painless enlarging mass and local pressure symptoms.

We report this pregnancy associated desmoid to add on to the scarce literature available about this condition to impress on the need for anticipating this rare condition in the differential diagnosis of fibroid complicating pregnancies. MRI is the ideal pre-operative diagnostic tool since it provides information regarding the extent of the disease, the location and the relationship to intra-abdominal organs. Furthermore, an MRI prevents a misdiagnosis of fibroid uterus, such as in the case presented [5].

There is paucity of data regarding the management options in pregnancy and labour. There are only few case reports and series and there are no devised guidelines for the management. The main modality of treatment described for desmoids is complete resection with a tumor free margin. Still higher incidence of recurrence has been quoted upto 20-30% in abdominal desmoids [6]. The role of radiotherapy and chemotherapy has been restricted to irresectable tumors [7]. The definitive role for complete resolution of tumor is debatable and their use in pregnancy is questionable. Thus the treatment modality has to be individualised as per the case scenario [8].

Since it is a well known fact that desmoids are hormone responsive tumors and demonstration of estrogen receptors has been noted, use of oral contraceptives and hormone replacement therapy must be avoided. The association with FAP is recognized and appropriate screening measures have to be adapted apart from the treatment of the desmoid tumor.

The association with FAP is recognized and appropriate screening measures have to be adapted apart from the treatment of desmoid tumor. Recurrence in future pregnancies should be studied to know about the role of hormonal influence by clinical examination and imaging.

## Conclusion

In conclusion, though a desmoid tumor is a rare entity, it has to be considered in the differential diagnosis of abdominal tumors in pregnancy.

## Figures

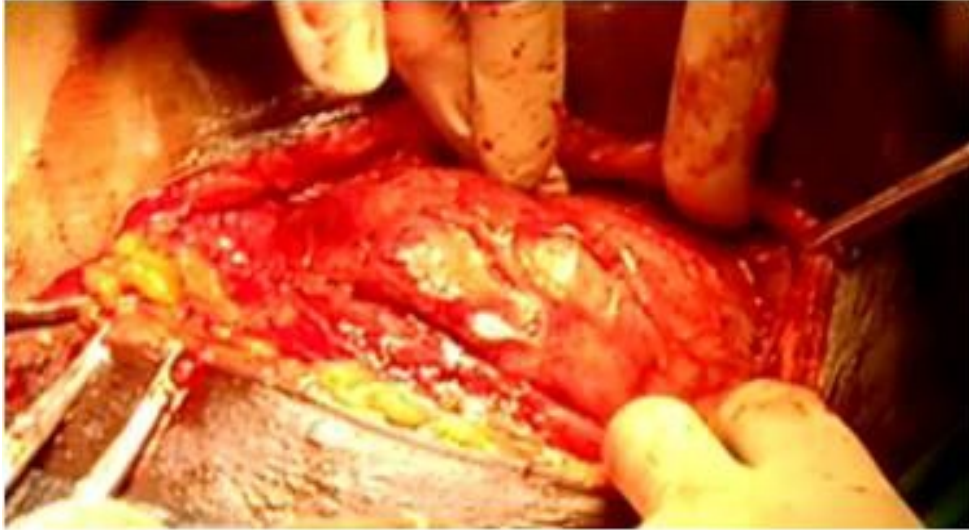


Figure 1



Figure 2

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