



Case Report

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Preference of the Surgeon: The patient or the radiologist? Desmoid tumor

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ABSTRACT

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Our aim is to present diagnosis of desmoid tumor in a patient with recurrent upper abdominal pain and mass with normal ultrasound findings. Fifteen-years-old boy had been operated due to acute appendicitis and intraabdominal abscess four years ago was admitted to our clinic with complaints of right upper quadrant pain and swelling for two years. Many ultrasound were performed but nothing was found. There was 6 centimeter (cm) thickening and heterogeneity of right rectus abdominis muscle with a suspicious mass neighboring the liver at computed tomography (CT) and magnetic resonance imaging (MRI). He was operated and pathology was reported as desmoid tumor. Even basic studies don't reveal any pathology if the patient's complaint goes on surgeon must be aggressive for diagnosis in order not to miss out malignancies.

Keywords:

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Children
Desmoid type fibromatosis
Aggressive fibromatosis

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1. Introduction

Desmoid tumor is musculoaponeurotic tumor also called aggressive or desmoid type fibromatosis. It's a rare neoplasm affects both children and adults (Fletcher et al., 2002). It's a deeply located intermediate malignancy. It effects 2-4 persons per 1 million population per year. It has two peaks: One is between 6-15 years of age and other is between puberty and 40 years of age in women (Faulkner et al., 1995; Buitendijk et al., 2005). It has a very slow progression.

Desmoid tumors can be managed with observation and close follow up according to data obtained from literature. But there is very few information for management of tumor in children (Fiore et al., 2009). Mortality from desmoid tumor in children is rare but

has been reported (Fletcher, 1998). Only a few studies in the literature mentioned difficulties in diagnosis (Posner et al., 1989). We present this case to show how much it is difficult to diagnose a desmoid tumor with very non-specific symptoms.

2. Case

Fifteen-years-old male patient was admitted to our clinic with complaint of right upper quadrant pain and swelling for two years. He was operated two times four years ago, one for appendectomy and one for intraabdominal abscess two weeks after. The patient had been admitted with the same complaint to many different clinics in the last two years. Many blood and urine analysis, X-ray of chest and abdomen was performed. Only liver

enzymes were found to be slightly high. Twenty-three different abdominal ultrasounds were performed but nothing found except suspected hemangioma in one. Patient itself had been insisted on pain and sensation of a mass at right upper quadrant just below the last rib. At erect and lying down position nothing was seen, but when straightening a suspicious lesion can be seen but cannot be palpated due to muscled body of the patient. All laboratory examinations and X-rays were again normal and ultrasound revealed nothing. Because he had two intra-abdominal operations and insisted on complaints, abdominal computed tomography (CT) with intravenous contrast was performed. Nothing was found intra-abdominal but there was thickening and heterogeneity of right rectus abdominis muscle. A mass cannot be ruled out and its neighboring to the liver cannot be clearly identified. Abdominal magnetic resonance imaging (MRI) was performed and approximately 6 cm soft tissue tumor was found. It had nothing to do with liver. It was considered as desmoid tumor according to MRI findings. He was operated and mass was excised approximately 5 cm beyond the tumor margin and en-bloc with fascia and peritoneum due to adherence. He was discharged 3 days after operation. Pathology was reported as desmoid tumor and all margins were free of tumor. KI-67 %1, desmin focal (+), S-100 focal (+), SMA focal (+), CD 114-7 (-), DOG-1 (-), CD 34(-), EMA (-), p53 (-) and weak nuclear staining with betacatein in immunohistochemical analysis. Oncologists decided not to give any chemotherapy or radiotherapy. He has been still free of tumor for the last 6 months.

3. Discussion

There are no guidelines at literature for desmoid tumor in pediatric population. In some cases, desmoid tumor in children can be managed successfully by surgery. But in cases those are really aggressive and rapidly growing, may need all therapeutical choices ie aggressive surgery, chemotherapy and radiation. Many studies have been performed dealing with this problem (Lopez et al., 1990, Faulkner et al., 1995). But in our case we especially insisted on pre-diagnosis part. Patient had been suffering from pain and sensation of mass for two years. He was observed by many pediatricians, pediatric surgeons and radiologist but somehow it couldn't be diagnosed.

There are many patients admitted to hospitals with abdominal pain and sure only very few of them require abdominal CT. Because of long lasting complaint and the patient had abdominal operations two times, we decided to perform abdominal CT. Also one of the

ultrasounds was reported as suspected hemangioma at right upper quadrant which means there is really a problem. We first suspected a pathology originating intra-abdominally due to previous pathologies that is why we perform abdominal CT before MRI. After performing abdominal CT we found that a mass in the anterior abdominal wall. But it was so close to liver that margins between both cannot be clearly identified. We were sure that it was located intramuscularly and didn't have any communication with liver after performing MRI. Actually MRI can be performed before CT but our suspicion was an intra-abdominal pathology at first.

Desmoid tumors are classified as those histologically benign but locally aggressive neoplasms (Faulkner et al., 1995; Maezza et al., 2010). At MRI it was located between muscles and didn't have a communication intra-abdominally. But at operation we found that even the mass itself was located between the muscle it invaded both fascia and peritoneum. It was firmly adhered to fascia and peritoneum that it was impossible to separate them. Also because the tumor itself has an invading nature it's not logical to try to separate them due to risk of microscopic residue. It was shown in the literature that there is a correlation with negative surgical margins and rates of recurrence (Fallen et al., 2006).

Sporadic desmoid tumors are rare. It mostly accompanied with familial adenomatous polyposis (Jones et al., 1986). Especially abdominal desmoids have a tendency to accompany with FAP. Fallen et al. (2006) found this percentage as 67%. In our case it didn't accompany to any other pathology. Both the patient and family didn't have any gastrointestinal complaints.

Abdominal region is the most effected part with a 37-50%. The shoulder girdle, chest wall and inguinal regions are the most prevalent extra-abdominal sites (Reitamo et al., 1982). In our case it was also located at anterior abdominal wall.

Philips et al. (2004) stated at their studies that twenty-one percent of 109 patients developed recurrence after a median follow-up of 39 months. They couldn't show any correlation with recurrence and negative surgical margin or any other variable. Our patient is at 6th month post operatively and hasn't required any other therapy or intervention and has been free of event.

Patients with long lasting complaints especially those make sense must be searched carefully. In order not to miss malignancy we may sometimes examine further. We also must remember that medicine is an art. Even machines don't tell any clues clinicians should be very careful.

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