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Uncommon abdominal wall mass in a young boy: Desmoid tumor

Genç bir erkek çocukta ender rastlanan batın duvarı kitlesi: Desmoid tümör

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SUMMARY

Fibromatosis that arises in musculoaponeurotic structures of the abdominal wall is called abdominal fibromatosis, abdominal desmoid tumor (DT), or aggressive fibromatosis. Fibromatoses occur at various anatomic locations with different clinical features. Abdominal fibromatosis tends to occur in young women. Conversely, in children is an extremely rare condition. Herein, we report the case of a 15-year-old boy with desmoid tumor. He was referred to our department because of a growing painful mass in the right lumbar area. There was no history of known trauma or surgery prior to the time. He was active wrestler. Ultrasonography demonstrated a solid and relatively well-circumscribed mass. Magnetic resonance imaging demonstrated a well-defined tumor originating from the abdominal transversal muscle and internal abdominal oblique muscle fascia. The mass was total excised. The histological diagnosis was desmoid tumor. In conclusion, in child patients presenting a painful tumors of the lower abdominal wall as well as DT should be considered. Recurrent sports trauma during wrestling can be a factor for this tumor, in our case.
Keywords: Desmoid tumor, abdominal wall, child

ÖZET

Karın duvarının kas-aponörotik yapılarından köken alan fibromatozislere abdominal fibromatozis, abdominal desmoid tümör (DT), ya da agressif fibromatozis adı verilir. Fibromatozisler farklı anatomik lokalizasyonlarda farklı klinik özelliklerde olabilirler. DT genç kadınlarda daha sıktır. Çocuklarda ise oldukça ender bir durumdur. Bu yazıda 15 yaşında desmoid tümörlü bir erkek olgu sunuldu. Hasta hastanemize sağ lumbal bölgede büyüyen ağrılı bir kitlenin farkedilmesi üzerine sevk edilmişti. Hastanın öncesinde bilinen bir travma ya da operasyon öyküsü yoktu. Aktif olarak güreş yapmaktaydı. Ultrasonografisinde solid ve göreceli olarak iyi sınırlı kitle saptandı. Manyetik rezonans görüntülemede iyi sınırlı, abdominal transversalis kası ile internal abdominal oblique kasının fasyasından kaynaklanan kitle saptandı. Kitle genel anestezi altında total olarak eksize edildi. Kitlenin histopatolojik incelemesinde desmoid tümör tanısı konuldu. Sonuç olarak lateral abdominal bölgede ağrılı kitle yakınmasıyla başvuran çocuklarda DT de ayırıcı tanıda dikkate alınması gereken bir patolojidir. Olgumuzda güreş sırasındaki yineleyen spor travması bu tümörün gelişiminde bir faktör olabilir diye düşünmekteyiz.
Anahtar sözcükler: Desmoid tümör, abdominal duvar, çocuk

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INTRODUCTION

Fibromatosis that arises in musculoaponeurotic structures of the abdominal wall is called abdominal fibromatosis, abdominal desmoid tumor (DT), or aggressive fibromatosis. Despite their aggressive local infiltration, desmoid tumors lack metastatic potential [1]. Aggressive fibromatosis is an often-used term to emphasize its frequently aggressive behavior. DT accounts for 0.03% of all neoplasms and 3% of soft tissue tumors, with a reported incidence of 2-4 individuals per million per year [2]. DT at various anatomic locations with different clinical features. Depending on the site of occurrence they are classified as:

1) Abdominal– in the anterior abdominal wall,
2) Intra-abdominal in the mesentery or pelvis, intraperitoneal or retroperitoneal,
3) Extra-abdominal in the chest, extremities and head and neck region.

They usually arise in the mesentery, omentum or retroperitoneum, while the abdominal wall is very rare [3]. DT tends to occur in young women. Conversely, in children is an extremely rare condition [4]. The exact incidence of DT in children is unknown.

Surgical excision with a clear resection margin has been accepted as the most successful treatment. Chemotherapy or radiotherapy has been suggested as adjuvant therapy in patients with residual tumors, as the potential for morbidity is high after the second operation [4].

CASE REPORT

Herein, we report the case of a 15-year-old boy with DT. He was referred to our department because of a growing painful mass in the right lumber area. There was no history of known trauma or surgery prior to the time that the mass was noticed by the parents. The analyzed blood parameters were within the normal range and the tumor markers were all negative. He was active wrestler. Ultrasonography demonstrated a hypoechoic, solid and relatively well-circumscribed mass. Colour Doppler ultrasonography showed vascular flow within the mass. Magnetic resonance imaging (MRI) demonstrated a well-defined tumor originating from the abdominal transversal muscle and internal abdominal oblique muscle fascia (Fig 1). Axial T2W MRI shows the classical appearance of DT: predominantly hyperintense (cellular) mass with hypo-intense (collagenous) bands. Axial, functional diffusion-weighted sequence does not demonstrate diffusion restriction. The mass was excised under general anesthesia. The postoperative course was uneventful and recovered without any complications or functional defects. The cut surface of the mass was solid and grayish-white without hemorrhage or necrosis. On gross examination, the tumor solid and it sizes were 3×2.5×5 cm and partly circumscribed. Histopathological examination of the specimen showed proliferation of bland-looking spindle-shaped cells with collagen production (Figs. 2-6). There was no necrosis. Mitotic rate was 0-1/10HPF. Ki 67 index was 3%. The histological diagnosis was, as a desmoid tumor.

Postoperative ultrasonography at six months showed no pathology. The patient is currently alive and healthy at three years after the surgery. It has been without tumor recurrence.

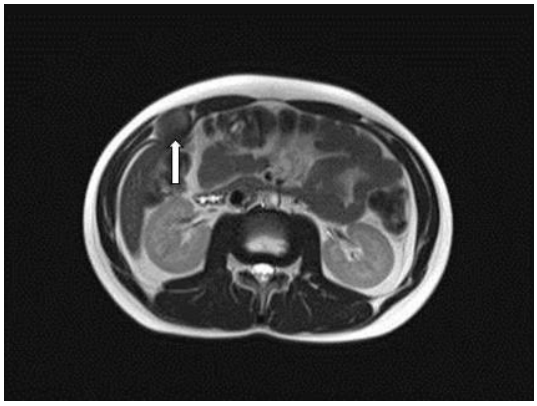


Figure 1. Axial T2 weighted MR image demonstrat a well-defined tumor originating from the transverse abdominal muscle and internal abdominal oblique muscle fascia.

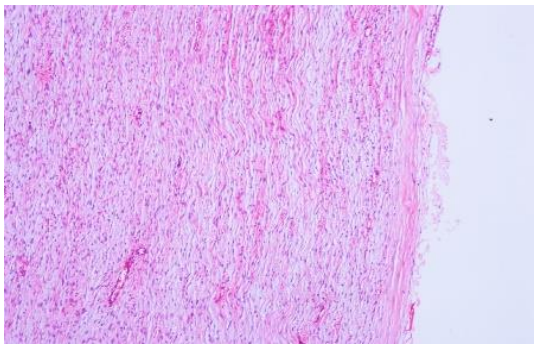
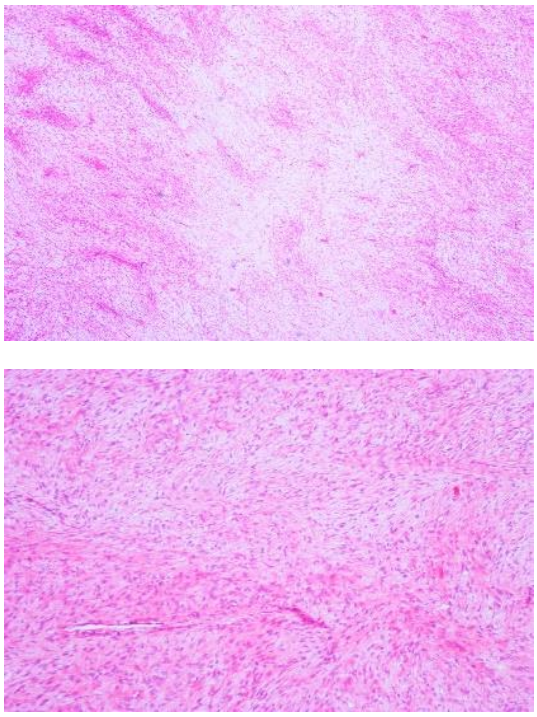


Figure 2. Well demarcated lesion seen in peripherally.



Figures 3, 4. Myxoid stroma with spindle cells in storiform pattern (H&E, X25) (H&E, X50).

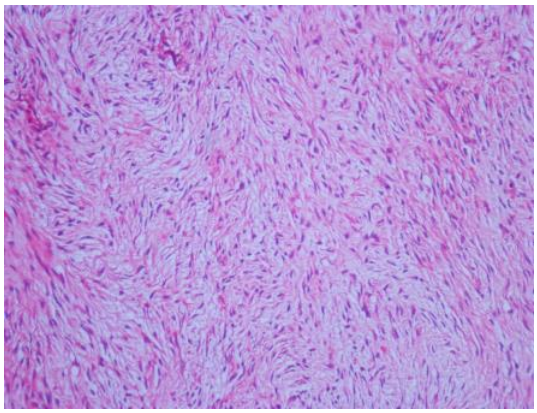


Figure 5. High-power field in figures (H&E, X100).

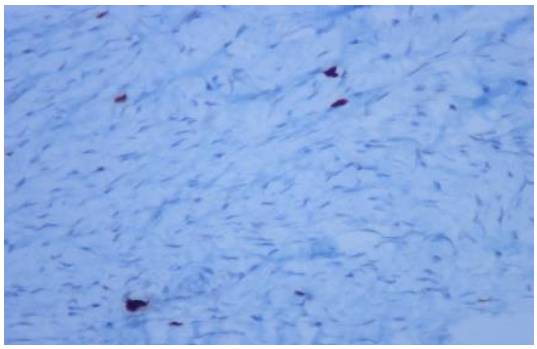


Figure 6. Ki-67 index: 3% (IHC, X50).

DISCUSSION

Soft tissue Tumors of the abdominal wall, though clinically similar, have many distinct histologic subtypes [5]. Desmoid tumors are rare slow growing stromal, benign muscular-aponeurotic fibrous tumors comprised of myofibroblasts with a strong tendency to invade locally and to recur [6]. İmmunohistochemically, myofibroblasts may have a variable phenotype, including those that express;

1. Vimentin (V Type) only; 2. Vimentin, smooth muscle alfa-actin, and desmin (type VAD) 3. Vimentin, smooth muscle alfa-actin (type VA); 4. Vimentin, and desmin (type VD). The pathologic types of our cases was V type [7].

Local recurrence after surgical resection is the major problem and depends on the completeness of the resection. Resection of sporadic intraabdominal desmoid tumor is associated with low recurrence rates [8].

Desmoid tumors usually occur in fertile females and are uncommon during the menopause; during pregnancy an increase in volume occasionally occurs in already existing tumors. This supports the estrogen-stimulated tumor growth hypothesis [9]. In the pediatric population, there is an equal sex ratio, with tumors being predominantly extra-abdominal[2]. Desmoids mainly affect pre-menopausal women and are associated with pregnancy, oral contraceptive use and traumatic events, including surgery, 80% or more develop following colectomy [10]. An antecedent history of trauma at the tumor site was elicited from 28% of patients in series of cases of Lopez at all [11]. Our patient was wrestler. We think that wrestling may be the cause of sports trauma.

In conclusion, some differential diagnoses include other abdominal wall mass as rectus hematoma, soft-tissue sarcoma, lipoma, and hemangioma. Although DT in children are rare, in child patients presenting a painful tumor of the lower abdominal wall as well as DT should be considered, especially children engaged in active sports. Recurrent sports trauma during wrestling can be a factor for this tumor, as in our case.

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