

CASE REPORTS

MAGNETIC RESONANCE IMAGING FINDINGS IN DESMOID TUMORS

DESMOİD TÜMÖRLERDE MAGNETİK REZONANS GÖRÜNTÜLEME BULGULARI

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SUMMARY: *Desmoid tumors, also known as aggressive fibromatoses, are uncommon, slow-growing, locally aggressive soft tissue lesions. The abdominal wall is one of the most common sites of presentation, but it may be seen in the neck in rare cases. The magnetic resonance imaging (MRI) findings of two desmoid tumor cases, one located in the neck and the other in the abdominal wall, and the value of MRI in the diagnosis is discussed. MRI is the first choice in the preoperative evaluation of neck desmoids due to its known superiority in showing the extent of a tumor and its contrast enhancement.*

Key Words: *Desmoid Tumor, Neck, Abdominal Wall, MRI.*

INTRODUCTION

Fibromatoses are a heterogenous group of soft tissue lesions that have been described using various terms and classifications, but are characterized by a similar histology. The most common type of fibromatosis is the desmoid tumor, also known as aggressive fibromatosis. A desmoid tumor is a benign connective tissue tumor that grows slowly and locally invades adjacent structures like the muscles, tendons, vessels and nerves without causing any distant metastases (1). Common anatomic sites include the extremities, the abdominal wall and the mesentery (1,2). In this article the MRI findings of two desmoid tumor cases, one located in the abdominal wall and the other in the neck, are presented and the value of MRI in diagnosis is

ÖZET: *Agresif fibromatozlar olarak da bilinen desmoid tümörler ender rastlanan, yavaş büyüyen lokal olarak agresif yumuşak doku lezyonlarıdır. Abdominal duvar en sık rastlanan lokalizasyonlardan biridir. Ancak boyun tutulumu nadirdir. Biz burada biri abdominal duvarda diğeri ise boyunda yerleşik iki desmoid tümör vakasında saptanan MRG bulgularını ve MRG'nin tanıdaki değerini tartıştık. Özellikle boyun desmoidlerinde MRG lezyonun uzanımını ve kontrast tutulumunu göstermedeki bilinen üstünlüğü nedeniyle preoperatif değerlendirmede ilk tercih haline gelmiştir.*

Anahtar Kelimeler: *Desmoid Tümör, Boyun, Karın Duvarı, MRG.*

discussed.

CASE REPORT

CASE 1

A 30-year-old woman presented with a painful lump on her midabdomen; she had no previous history of surgery or trauma in her anamnesis. Physical examination and laboratory results were within normal ranges. Ultrasonographic examination revealed a heterogenous isoechoic smoothly marginated area approximately 3.5 × 2.5 × 4 cm in size present as a solid mass located within the left rectus abdominis muscle. The lesion showed lower attenuation than the muscle on CT images. The lesion was hyperintense according to the muscle on T2 weighted images (T2WI), whereas

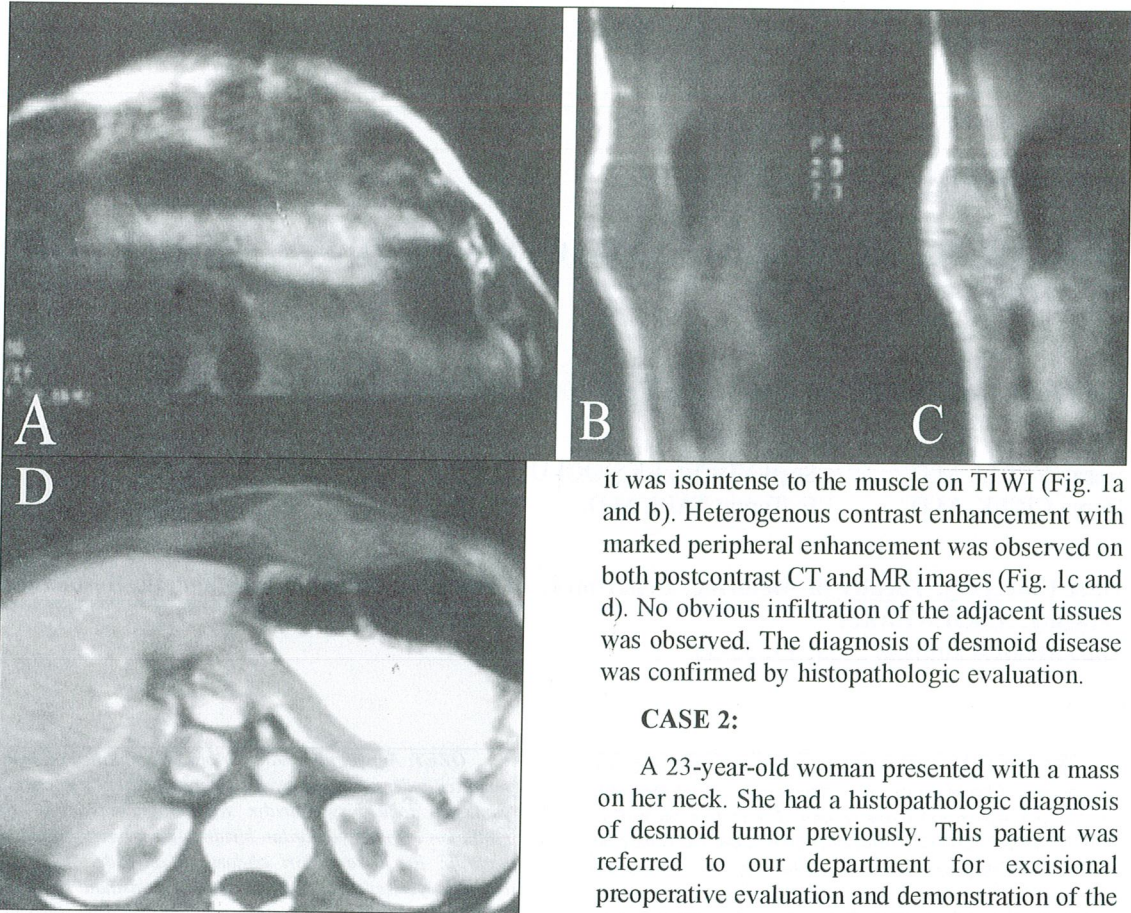


Fig. 1: T2W axial MR image shows a hyperintense mass lesion located within the left rectus abdominus muscle (A). T1W precontrast sagittal image shows that the lesion is isointense with the muscle (B). Postcontrast sagittal MR and axial BT images show the heterogenous enhancement of the mass lesion is most prominent at the periphery (C, D).

it was isointense to the muscle on T1WI (Fig. 1a and b). Heterogenous contrast enhancement with marked peripheral enhancement was observed on both postcontrast CT and MR images (Fig. 1c and d). No obvious infiltration of the adjacent tissues was observed. The diagnosis of desmoid disease was confirmed by histopathologic evaluation.

CASE 2:

A 23-year-old woman presented with a mass on her neck. She had a histopathologic diagnosis of desmoid tumor previously. This patient was referred to our department for excisional preoperative evaluation and demonstration of the extent of tumor growth. MR revealed a mass lesion of $9.5 \times 6.5 \times 4.5$ cm extending from the level of the uvula to the supraclavicular fossa. The trachea was deviated to the left, compressed and narrowed by the lesion (Fig. 2a). Following the administration of IV contrast media, the mass

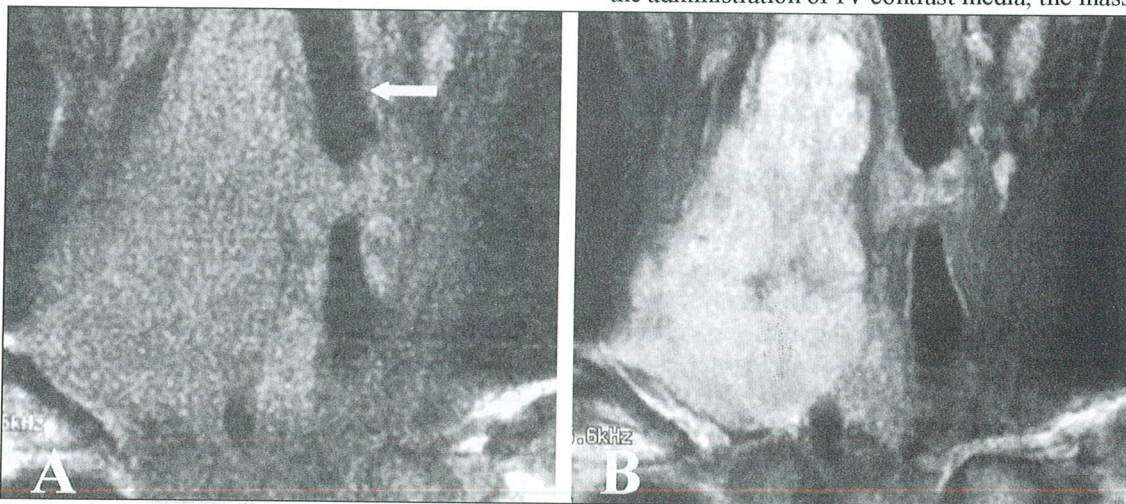


Fig. 2: T1W precontrast coronal MR image demonstrates a mass lesion extending from the uvula level to the supraclavicular fossa. The lesion causes tracheal deviation (arrow) to the left (A). Postcontrast image shows the marked heterogeneous, diffuse contrast enhancement (B).

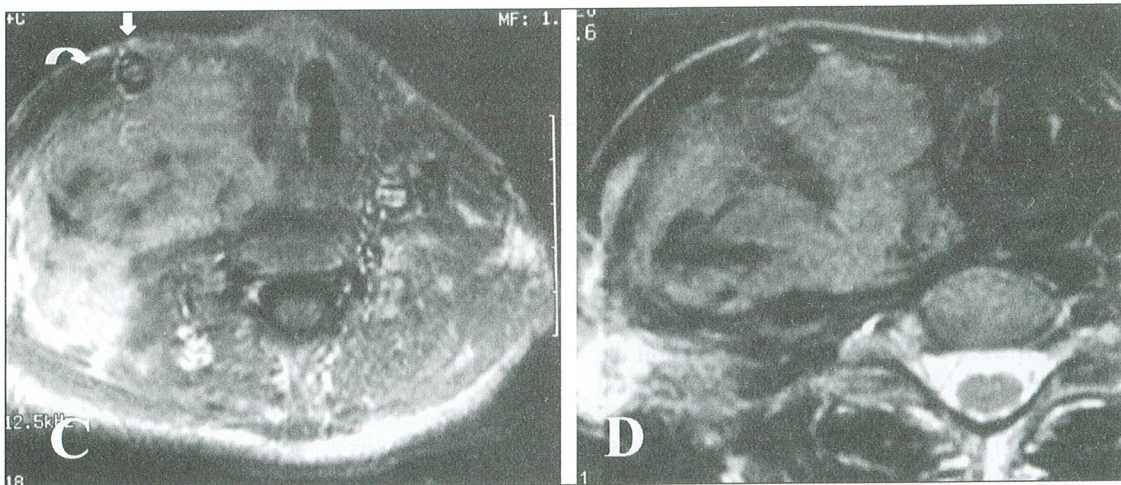


Fig. 2: Precontrast T1W axial images show the displacement of the jugular vein (curved arrow) and carotid artery (arrow) caused by the mass lesion which infiltrates the cutaneous and subcutaneous fat tissue (C). The lesion has a higher signal intensity than the adjacent muscle tissue on the T2W axial image (D).

lesion showed a heterogenous enhancement (Fig. 2b). The margins of the lesion could not be differentiated from the right thyroid lobe, and it displaced the jugular vein and carotid artery laterally and so caused a significant decrease in the calibration at various levels (Fig. 2c). Anterior extensions to the cutaneous and subcutaneous area and posterior extensions to the paravertebral area and multifidus, semispinalis capitis and splenius capitis muscles were observed. The lesion was hyperintense relative to muscle on T2WI (Fig. 2d). No lymphadenopathy was present on the cervical chains bilaterally.

DISCUSSION

The Greek word "desmos", which means band or tendon, is the origin of the term "desmoid" and was describe by Muller to define tendon-like tumors (3). Desmoid tumors can be classified as fibromatoses, a group which comprises plantar fibromatosis, Peyronie's disease, Dupuytren's contracture and hypertrophic scars (4).

Fibromatoses are infiltrating fibroblastic proliferations that are locally aggressive and histologically benign neoplasms arising from the fascia or aponeurosis of muscles in any area of the body. The prevalence is around 2-4 cases/million. In addition to the abdominal wall, which is one of the most common sites of presentation (49% of cases), desmoids may be seen in the pelvis, mesentery, retroperitoneum and shoulder, and more rarely in the chest wall,

neck, breast and mediastinum (4-8). The rectus muscle/sheath is a typical location for the desmoid accounting for two-thirds of all abdominal wall locations. Lambroza et al. reported that desmoid tumors constitute the most frequent tumors of the rectus muscle/sheath complex (7). Less commonly, the internal oblique or external oblique muscle may be involved. They are typically seen in gravid, parous women during gestation or more frequently during the first year of childbirth (70% of women are between 20 and 30 years of age), and sporadic cases may also be seen among adults (4,6,9,10).

Some of the possible causes suggested are previous accidental trauma, surgery, hormonal effects especially estrogens and pregnancy (10,11). Increased frequency of tumors has been seen in association with Gardner's syndrome. Molecular studies have confirmed that these lesions are the result of a clonal process, thus establishing that desmoids are neoplasms and not the product of an intense inflammatory fibrous reaction (12). Because of the distinct preponderance among women, hormonal factors, especially in young women, are also considered to be a precipitating factor. CT is the standard technique for evaluating suspected intraabdominal desmoid tumors (13). CT is sufficient to determine the localization of the lesion and its relationship with the adjacent soft tissues. However, the enhancement pattern of the tumor on CT is not typical. It may be peripheral, homogeneous or heterogeneous. Desmoid tumors

do not have specific attenuation values. Abdominal desmoids are usually isointense according to the muscle on T1WI, but they can show different signal characteristics on the T2WI of MRI examinations (14). The signal changes with the age of the tumor. In an aging tumor, as the collagen ratio increases with time, the signal decreases. MRI also enables the evaluation of the contrast enhancement pattern better because of its improved contrast resolution over that of CT (14). However, it is not possible to mention the superiority of MRI to CT in the radiological evaluation of abdominal desmoids during the diagnosis stage. The diagnosis can be confirmed by histopathologic examination. It should be emphasized that during the follow-up stage the presence of high signal intensity on T2W images (signal intensities equal to or greater than water) may be useful in predicting those desmoid tumors that will grow rapidly (14).

Desmoid tumors of the head and neck are rare, fibrous neoplasms with a good prognosis, but significant morbidity. Low-grade fibrosarcomas take place in differential diagnosis. Head and neck desmoids are histologically the same as abdominal desmoids and other extra-abdominal desmoids, but head and neck desmoids are different in that they are usually invasive and spread even to tissues surrounding deep blood vessels and nerves, while they are also difficult to remove completely and they are likely to show recurrence after surgery (2). MRI is the first choice in the preoperative evaluation of head and neck desmoids due to its significant superiority in showing the extent of a tumor and its contrast enhancement.

In conclusion, fibromatosis has to be considered in the diagnosis of abdominal wall tumors and infiltrative tumors of the head and neck region. The efficacy of MRI depends on the localization of the tumor and the stage of the clinical course.

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