

## CONTINUING MEDICAL EDUCATION ΣΥΝΕΧΙΖΟΜΕΝΗ ΙΑΤΡΙΚΗ ΕΚΠΑΙΔΕΥΣΗ

### Medical Imaging Quiz – Case 81

A 45-year-old female patient presented due to frequent urination and abdominal pain. She did not have any underlying disease or history of cancer. Laboratory evaluation revealed none pathological finding; thus, a mass was observed in abdominal ultrasound (US). Magnetic resonance imaging (MRI) was performed and confirmed the underlying pathology (fig. 1).

#### Comments

Desmoid tumors are benign, non-inflammatory fibroblastic tumors with a tendency for local invasion and recurrence post resection. They are sometimes considered a locally aggressive proliferative disease within the family of soft-tissue sarcomas, but metastasis is uncommon. Fibroblastic tumors have abdominal or musculoskeletal presentation. They are rare lesions with an estimated incidence of 3.7 new cases per million population per year, representing only 0.03% of all neoplasms. Desmoid tumors occur in all age groups, but are most frequently encountered between 20 and 40 years of age. They have a women-to-men prevalence 2:1. The term “desmoid” originates from the Greek word “desmos” (δεσμός), meaning band or tendon-like. Some cases have been associated with pregnancy and estrogen therapy and Gardner syndrome. Mesenteric desmoid tumors are seen either sporadically or in association with familial polyposis coli syndrome (FAP): 9–18% of FAP cases may have a desmoid tumor.

Desmoid tumors present as masses, and as such presenta-

tion depends on location. Their exact etiology remains uncertain, although they are frequently associated with previous trauma or surgical incision. On the molecular level, desmoids are characterized by mutations in the  $\beta$ -catenin gene, CTNNB1, or the adenomatous polyposis coli (APC) gene. Frequent locations in the abdomen are the abdominal wall, the root of the mesentery and the retroperitoneum. They are the commonest neoplasms of the abdominal wall and typically appear as homogeneously hypoechoic masses in US. They may have a similar appearance to muscle, may be lobulated and may show vascularity on color Doppler interrogation.

In computed tomography (CT) most desmoid tumors are well-circumscribed masses, relatively homogeneously or focally hyperattenuating when compared to soft tissue on the non-contrast scan and demonstrate enhancement following administration of intravenous contrast. In some cases, they may appear more aggressive with ill-defined margins. MRI is more sensitive to local tumor extension. Their appearance is accounted by their dense cellularity. Typical signal characteristics include low signal intensity in T1 and high heterogeneous signal in T2. Hyperintensity may diminish over time as tumor cellularity decreases and collagen deposition increases. With gadolinium administration desmoids typically show moderate to marked enhancement, hypointense bands may become more apparent because collagen bundles are not enhanced by contrast material. Imaging differential consideration include carcinoid tumor.

Most cases can have relatively good prognosis and watchful waiting is now considered a reasonable option in selected asymptomatic patients. Other management options include: Surgical resection (traditionally used, although recurrence rate is high), non-steroidal anti-inflammatory drugs (NSAIDs) and anti-estrogens can be used to reduce the rate of recurrence and radiotherapy.

#### References

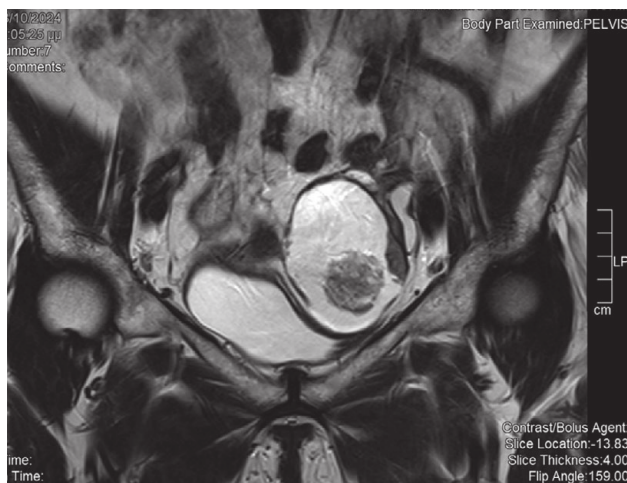
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**Figure 1.** T2 magnetic resonance imaging (MRI) of pelvis reveals a mass with high heterogeneous signal.

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