CASE REPORT

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CALCIFYING FIBROUS TUMOR OF THE RECTUM - A CASE REPORT

TOMISLAV PAVLOVIĆ[1,2], ROSANA TROSKOT PERIĆ[1,3,4] and SANJA TRTICA[5]

[1]Josip Juraj Strossmayer Unversity of Osijek, Faculty of Medicine, Osijek, Croatia;
[2]Department of Radiology, St.Catherine Specialty Hospital, Zabok, Croatia;
[3]Department of Internal Medicine, University Hospital Sveti Duh, Zagreb, Croatia;
[4]University of Rijeka, Faculty of Health Studies, Rijeka, Croatia;
[5]Department of Radiology, University Hospital Sveti Duh, Zagreb, Croatia

Summary

A calcifying fibrous tumor (CFT) is a benign tumor of unknown etiology. A calcifying fibrous tumor is rare in the intestinal tract. A calcifying fibrous tumor is characterized by hyalinized collagenous fibrous tissue, psammomatous or dystrophic calcification, and focal lymphoplasmacytic infiltrates on histology. Magnetic resonance imaging is the standard method for evaluating the lesions of the rectum, and CFTs should be considered in differentiating the rectal wall tumors. Herein, we report a case of a 68-year-old man with a rectal wall CFT.

KEYWORDS: *magnetic resonance imaging (MRI), calcifying fibrous tumor, rectum*

INTRODUCTION

A calcifying fibrous tumor (CFT) is a soft, rare benign tumor with unknown etiology. A CFT is characterized by the presence of a hyalinized collagenous fibrous tissue, psammomatous or dystrophic calcification, and focal lymphoplasmacytic infiltrate (1). CFT grows in the pleura, extremities, mediastinum, abdominal cavity, lungs, heart, mandible, neck, spine, back, inguinal and paratesticular area, and scrotum (2). CFT was first described in children (3) and rarely grows in the intestinal tract (4,5,6,7). Herein, we describe a case of a patient with a CFT in the rectal wall.

CASE REPORT

A 68-year-old man without any symptoms arrived for a routine check-up. Personal and family history were unremarkable. The digital rectal exam found a mass on the front wall of the rectum with intact rectal mucosa. The laboratory values for leukocytes, erythrocytes, hemoglobin, hematocrit, and liver functions were within the normal range. The serum level of the carcinoembryonic antigen was also within the normal range. We performed the colonoscopy with biopsy. A pathohistological pattern of four pieces of tissue was not conclusive. The patient then underwent a magnetic resonance imaging (MRI) of the pelvis. Radiologists used an MRI protocol with intravenous contrast and rectal distension with rectal gel application. The MRI showed a mass in the lower third of the rectum's front wall at a distance of 5 cm from the anal verge. The mass was well-circumscribed, measuring 25x20x26mm. The tumor formation was low signal intensity on the T2-weighted sequence (Fig. 1), intermediate signal intensity on the T1-weighted sequence (Fig. 2a). The tumor mass showed homogenous enhancement after contrast application (Fig. 2b). On the diffusion-weighted imaging (DWI) sequences

***Corresponding author**: Rosana Troskot Perić, Department of Internal Medicine, University Hospital 'Sveti Duh,' Sveti Duh 64, 10000 Zagreb, Croatia.*
e-mail: rtroskot@gmail.com

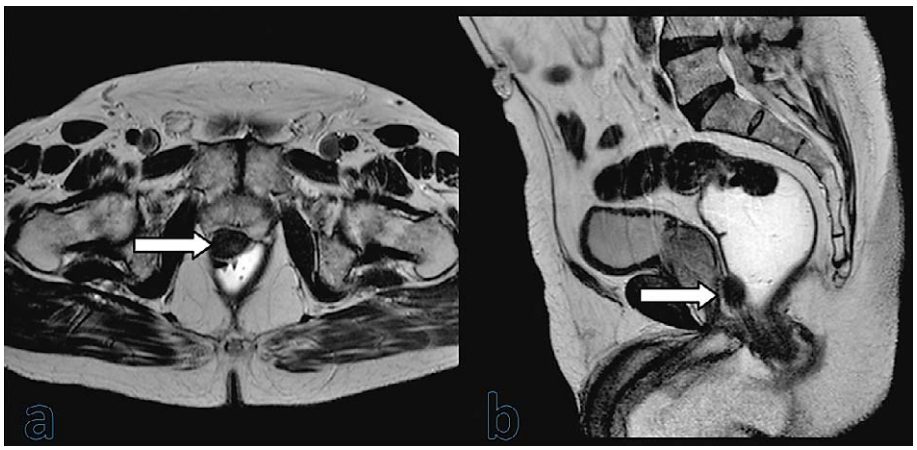

Figure 1. T2-weighted axial (a) and sagittal (b) plane, low signal intensity mass in the rectal wall (shown by arrow)

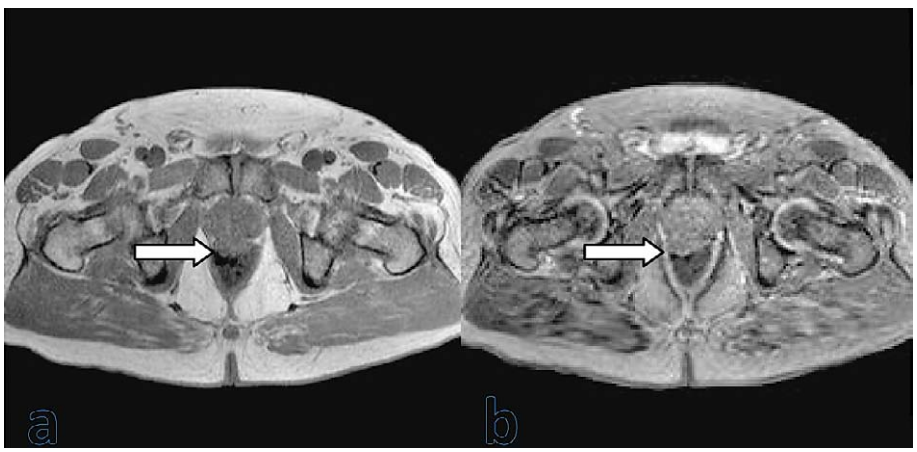

Figure 2. T1-weighted (a), T1 THRIVE post-contrast axial plane image (b) showed homogenous enhancement of the mass (shown by arrow)

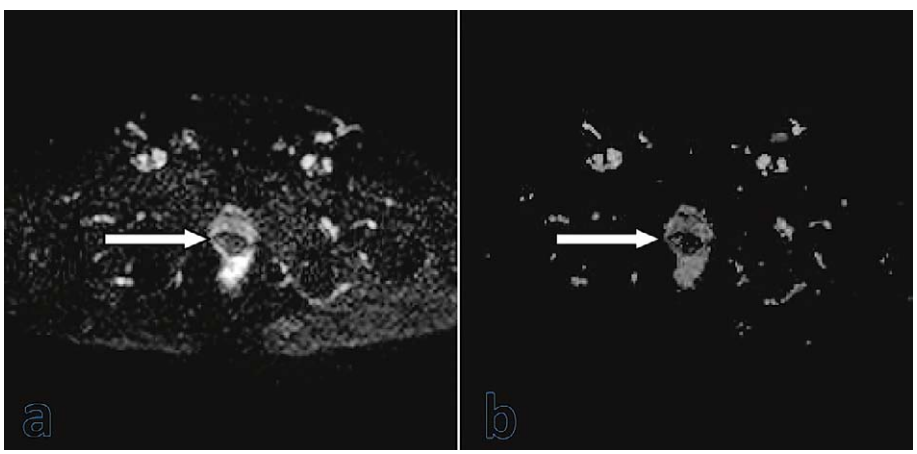

Figure 3. Diffusion-weighted imaging (a) and apparent diffusion coefficient (b) showed the low signal intensity of mass (shown by arrow)

(Fig. 3a) and apparent diffusion coefficient (ADC) mapping (Fig. 3b), the mass was low signal intensity. The MRI examination suspected a fibrous tumor. Based on the findings, the abdominal surgeon suggested the resection. After the operation, the pathology confirmed a calcifying fibrous tumor - the rectal submucosal tumor consisted of fibrous tissue, collagenous tissue, inflammatory cells, and calcification psammoma bodies.

DISCUSSION

A calcifying fibrous tumor is a soft tissue tumor, a rare and benign tumor of unknown etiology. Rosenthal and Abdul Karim described the first cases of CFT as a fibrous tumor with psammoma bodies in two girls, two and eleven years old (3). The pathogenesis of the CFT is not clarified. Some studies connect the CFT to inflammatory myofibroblastic tumors (IMT), which suggests that CFT is the late IMT sclerosis phase (8). Some studies claim a CFT has a different immunohistochemical and histological appearance suggesting that the IMT and CFT are different lesions (9). The presence of a hypocellular, densely hyalinized collagen with psammomatous or dystrophic calcification, a proliferation of fibroblastic spindle cells, mononuclear inflammatory infiltrates, and lymphoid aggregates such as focal plasmocytes, neutrophils, eosinophils and mast cell infiltration characterize a histological CFT (2). Immunohistochemical analysis is useful for the CFT diagnosis. Fibroblasts in the CFT do express factor XIIIa and vimentin, and they do not express S100, desmin, acetone, CD117, CD99, CD31, SMA, CD20, CD32, CD130, CD21, AE1 / AE3, EMA, estrogen receptor, callethrin, progesterone receptor, osteopontin, CAM5.2 and β-catanine (2). Chorti et al. analyzed 157 cases of CFT in 104 articles and found that the mean age of patients was about 34 years, with a higher incidence in women than men (1.27: 1) and a mean tumor diameter was 4.6 cm (2). Im et al. published a case report of a patient with a rectal submucosal tumor and further analyzed six cases of CFTs in the digestive system. They reported that an intestinal CFT appears more often in men (66%), with smaller tumor diameter, and appears as a solitary lesion similar to our case (7).

Radiology differential diagnosis of the CFT in the digestive system includes the gastrointestinal stromal tumor (GIST), desmoid, solitary fibrosis tumor (SFT), leiomyoma, schwannoma, and inflammatory myofibroblastic tumor (IMT).

Gastrointestinal stromal tumors present as solid masses, well-circumscribed, and may cause the narrowing of the lumen. Some lesions might have central necrosis. On MRIs, they tend to have low signal intensity on T1-weighted and high signal intensity on T2-weighted; the post-contrast enhancement is homogenous in the solid variety and the ring pattern in the case of a necrotic center (10). Large-sized lobulated contours increase in size on follow-up imaging, and the presence of hematogenous metastases are signs of malignancy; at the time of presentation, more than 50% of all GISTs have evidence of metastatic disease (10).

Desmoid tumors may manifest as a pelvic mass and may occur at any age. These tumors are not encapsulated with coarse white trabeculae that simulate scar tissue; at histologic analysis, desmoid tumors consist of elongated spindle-shaped cells separated by dense collagen bands (11). These tumors are low signal intensity on T1-weighted MR images and have a variable signal intensity on T2-weighted images (12).

A solitary fibrosis tumor is a mesenchymal tumor that appears mostly in the pleura, extremely rare in the pelvis, and usually occurs in the fifth decade. These tumors have intermediate signal intensity on T1-weighted images, and variable signal intensity on T2-weighted images shows that the low signal intensity on T2-weighted images suggests fibrous tissue and collagen stroma (13).

A leiomyoma is a rare tumor in the gastrointestinal tract. Tumors appear as a well-defined soft tissue intensity mass, the heterogeneous intermediate signal intensity with high signal intensity areas on T1-weighted sequences, and intermediate to high signal intensity on T2-weighted sequences (14). Schwannomas are peripheral nerve tumors arising from Schwann cells of the nerve sheath, rarely located in the pelvis. Schwannomas are considered to be benign tumors with the possibility of malignant transformation. On MRI, these tumors have homogeneous low signal intensity on T1-weighted images and heterogeneous slight high signal intensity on T2-weighted images (15).

An inflammatory myofibroblastic tumor (IMT) is a very rare mesenchymal tumor, affecting

various organs, most frequently the lungs. An IMT is a spindle cell proliferation of disputed etiology, with a distinctive fibroinflammatory and even pseudosarcomatous appearance. Tumors appear as a heterogeneous mass. The appearance is slightly low on MRIs on T1-weighted images and mixed intensity on T2-weighted images. Enhanced-MRI shows an enhanced tumor (16). A DWI is routinely used in protocols for MRIs for the pelvis, allowing tissue characterization. High signal intensity on a DWI with simultaneous low signal intensity on ADC maps is usually associated with malignancy (17). A similar pattern is seen in some benign lesions. DWI findings must be correlated with standard T1-weighted and T2-weighted images as well as with contrast-enhanced MRIs to avoid potential pitfalls. In our case, a low signal in DWIs and the low signal of ADC maps indicate that the tumor is a benign lesion, and the low signal in T2-weighted images further indicates a fibrous tissue and collagen stroma as a benign lesion. A proctoscopy with biopsy is the most important method for diagnosing rectal tumors, but this procedure cannot determine the exact intramural extension of the tumor and cannot accurately differentiate submucosal and intramural tumors from extramural tumors. Transrectal ultrasound and MRI allow for evaluating the intestine wall thickness, which helps to characterize these tumors better. Recognizing these masses' existence and their clinical and imaging features is crucial for clinicians to diagnose and adequately manage these conditions accurately.

CONCLUSIONS

A calcifying fibrous tumor in the intestinal tract is very unusual and clinically unexpected. An MRI is a useful method for evaluating the rectum wall process, and a CFT should be considered in the differential diagnosis of rectal submucosal tumors.

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Sažetak

KALCIFICIRANI FIBROZNI TUMOR REKTUMA: PRIKAZ SLUČAJA

T. Pavlović, R. Troskot Perić i S. Trtica

Kalcificirani fibrozni tumor (CFT) je benigni tumor nepoznate etiologije. CFT u crijevnom traktu je vrlo rijedak. CFT karakterizira prisutnost hijaliniziranog kolagenskog vlaknastog tkiva, psamomatoznih ili distrofičnih kalcifikacija i fokalnih limfoplazmocitnih infiltrata. Prikazujemo slučaj 68-godišnjeg muškaraca s CFT-om u stijenci rektuma. Magnetska rezonancija je korisna metoda za procjenu procesa stijenke rektuma, CFT treba uzeti u obzir u diferencijalnoj dijagnozi tumorskih procesa stijenke rektuma.

KLJUČNE RIJEČI: *magnetska rezonancija (MR), kalcificirajući fibrozni tumor, rektum*