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CT and MRI diagnostics of desmoid-type fibromatosis in familial adenomatous polyposis

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ABSTRACT *AIM: to study of the features of computed tomography (CT) and magnetic resonance imaging (MRI) for desmoid-type fibromatosis (DF) in familial adenomatous polyposis (FAP).*

PATIENTS AND METHODS: the study included 35 patients with desmoid-type fibromatosis (DF) with familial adenomatous polyposis of the colon (FAP). All patients were examined using CT and MRI with intravenous contrast. The site, size, growth pattern, prevalence of DF, features of contrasting and the intensity of the MR signal on T2-weighted and post-contrast T1-weighted were assessed. Twenty-five (71.4%) patients were followed-up, including systemic therapy.

RESULTS: In 21 (60%) patients, only one anatomical zone was involved, when 14 (40%) showed lesions in different anatomical zones. In most cases (33/35, 94.4%), desmoid-type fibromatosis was detected in the mesentery and in root of the small bowel mesentery, including those with combined involvement. Most patients (24/35, 68.6%) were diagnosed with a combination of infiltrative and mass-like form of growth; in 13 (37.1%) mass-like form and in 6 (17.1%) infiltrative form. Twenty-five patients (71.4%), repeatedly re-examined using CT (13/35, 37.1%) and MRI (12/35, 34.3%), in particular during systemic therapy.

CONCLUSION: CT and MRI are the basic methods for detecting DF in FAP, making it possible to determine the pattern of tumor growth, assess its extent of the tumor and the involvement of adjacent organs and structures. In follow-up and evaluation of the response of a desmoid-type fibromatosis to systemic therapy, MRI has greater diagnostic capabilities compared to CT, since it takes into account not only the size of the desmoid tumor, but also the MR signal intensity on T2-weighted and the pattern of the accumulation of a contrast agent on post-contrast T1-weighted with fat saturation.

KEYWORDS: *desmoid-type fibromatosis, desmoid tumor, familial adenomatous polyposis, Gardner's syndrome, computed tomography, magnetic resonance imaging*

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INTRODUCTION

Desmoid fibroids (desmoids, desmoid tumors) are a special variant of mesenchymal tumors that do not metastasize, but are prone to aggressive local growth and recurrence [1,2]. A combination of familial adenomatous polyposis (FAP) and desmoid fibroids (DF), known as Gardner's syndrome, occurs in 10–20% of patients with FAP while one or more desmoid tumors may develop during their lifetime [3–6]. In FAP, desmoids are most often intraabdominal, and with multifocal growth they are often combined with extraabdominal tumors located usually in the anterior abdominal wall [7,8]. Over the past decade, approaches to the DF have changed and include follow-up ('watch and wait'), surgical treatment, systemic therapy (steroids, chemotherapy,

targeted therapy) [9–14]. Imaging, primarily CT and MRI, are important in identifying desmoids determining the extent of the process and the involvement of adjacent organs and structures [11,15–19]. When planning a surgery, this information plays a key role in assessing the resectability of the tumor at the preoperative stage. Radiation methods are of no less importance in detecting relapses of DF, as well as in assessing the tumor response to systemic therapy [14,20,21]. In most publications, the main attention is paid to the imaging of sporadically occurring DF, mainly extraabdominal location (limbs, head, neck, chest, anterior abdominal wall), and the issues of CT and MRI diagnostics of desmoid tumors in FAP are covered only in some works [11,22–25].

AIM

The aim was to study the features of CT and MRI diagnostics of desmoid fibroids in familial adenomatous polyposis.

PATIENTS AND METHODS

The study included 35 patients with confirmed FAP in combination with desmoid tumors of the anterior abdominal wall and mesentery of the small intestine (Gardner's syndrome), in 2009–2021. There were 23 females and 12 males aged 37.3 ± 7.2 (23–57) years. All patients underwent colectomy with J-pouch in 27 cases and excision of the anterior abdominal wall desmoids in 3 cases. After surgical treatment, all patients were checked-up with abdominal CT and MRI with intravenous contrast. In 25 (71.4%) of them the follow-up ranged 2–10 years after surgery. A total of 98 studies were performed on 35 patients: 50 CT and 48 MRI. The desmoid tumors in most cases (30/35, 85.7%) were detected after 2–4 years after surgery. Only in 5 (14.3%) of them desmoids were detected during the initial examination before surgery.

Computed tomography was performed on a "CT Philips Brilliance 64". It included scanning of the abdominal cavity and pelvic organs with a slice thickness of 2 and 3 mm after intravenous bolus injection of a nonionic contrast agent in a volume of 80–100 ml at a rate of 2.5–3 ml/ sec.

Magnetic resonance imaging was performed on a 'Philips Achieva' with a magnetic field strength of 1.5 T. A 16-channel receiving and transmitting coil for the Sense XL Torso body was used for scanning. Abdominal and pelvic organs were scanned with intravenous gadolinium containing contrast agent 0.1 mmol/kg.

Bowel cleansing for CT and MRI studies included a diet with the exclusion of gas-forming products 2–3 days before the diagnostic procedure, with the last meal taken at least 6 hours before the procedure.

When analyzing the CT and MRI images obtained, the site, growth pattern, prevalence of desmoid tumor, involvement of adjacent organs and structures were determined. The structure of the DF and the degree of accumulation of contrast agent were evaluated with CT on native (contrast–free) tomograms and after intravenous contrast; with MRI — on T2-WI images and post-contrast T1-WI images with adipose suppression.

RESULTS

In 21 (60.0%) patients, only one anatomical area was affected (mainly the mesentery of the small intestine), and in 14 (40.0%) cases, desmoid tumors were located in different anatomical zones: the mesentery of the small intestine and the anterior abdominal wall, the mesentery of the small intestine and the pelvis (Fig. 1). In the majority of patients (33/35, 94.4%), DF was detected in the mesentery, including simultaneous lesions (Table 1).

Two thirds of patients (24/35, 68.6%) showed *infiltrative-nodal form* of growth of desmoid tumors located in the mesentery of the small intestine and pelvis, in 8

Table 1. *Location and pattern of growth of desmoid tumors according to CT and MRI (n = 35)*

Location	Form of growth				Total
	Infiltrative-nodal form	Infiltrative	Nodal form	Infiltrative-nodal + nodal form*	
Mesentery of the small intestine	11 (31.4%)	5 (14.3%)	3 (8.7%)		19 (54.4%)
Anterior abdominal wall			1 (2.8%)		1 (2.8%)
Retroperitoneal space			1 (2.8%)		1 (2.8%)
Mesentery of the small intestine + anterior abdominal wall				8 (22.9%)	8 (22.9%)
Mesentery of the small intestine + small pelvis	5 (14.3%)	1 (2.8%)			6 (17.1%)
Total	16 (45.7%)	6 (17.1%)	5 (14.3%)	8 (22.9%)	35 (100%)

** In the mesentery of the small intestine, there is an infiltrative-nodal form of growth, in the anterior abdominal wall –nodal form*

of them — in combination with desmoids of the anterior abdominal wall (Table 1). During CT and MRI, this form was characterized by infiltrative growth, without clear margins, tumor spread in the mesentery of the small intestine and/or pelvic cavity with the involvement of small intestine loops (23/35, 65.7%), mesenteric vessels (19/35, 54.4%), pelvic peritoneum (5/35, 14.3%), ureters (3/35, 8.6%), J-pouch

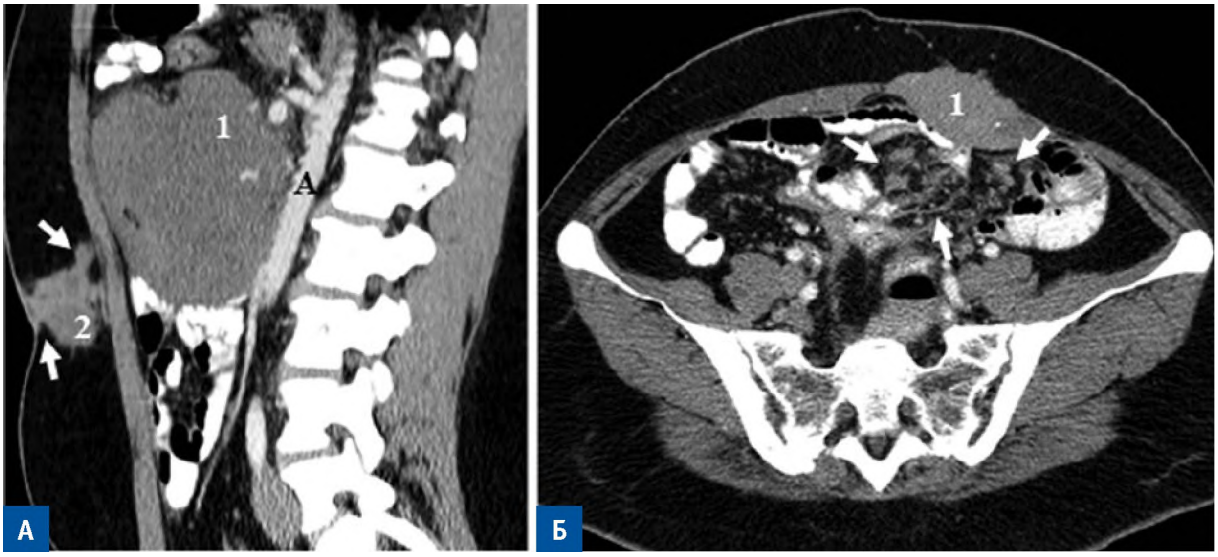

Figure 1. *Computed tomography with contrast enhancement. Combined lesion of the abdominal cavity and anterior abdominal wall. A — tomogram in the sagittal projection of the patient L., 28 years old; 1 — DF of the mesentery of the small intestine with dimensions of 11 × 9.5 cm, adjacent to the abdominal aorta; the structure is homogeneous with a slight accumulation of a contrast agent (A); 2 — DF of the anterior abdominal wall with infiltration of subcutaneous adipose tissue and skin; the structure is heterogeneous with moderate accumulation of the contrast agent (arrows). Б — tomogram in the axial projection of the patient K., 50 years old; 1 — DF of the anterior abdominal wall; diffuse infiltration of the mesentery of the small intestine (arrows)*

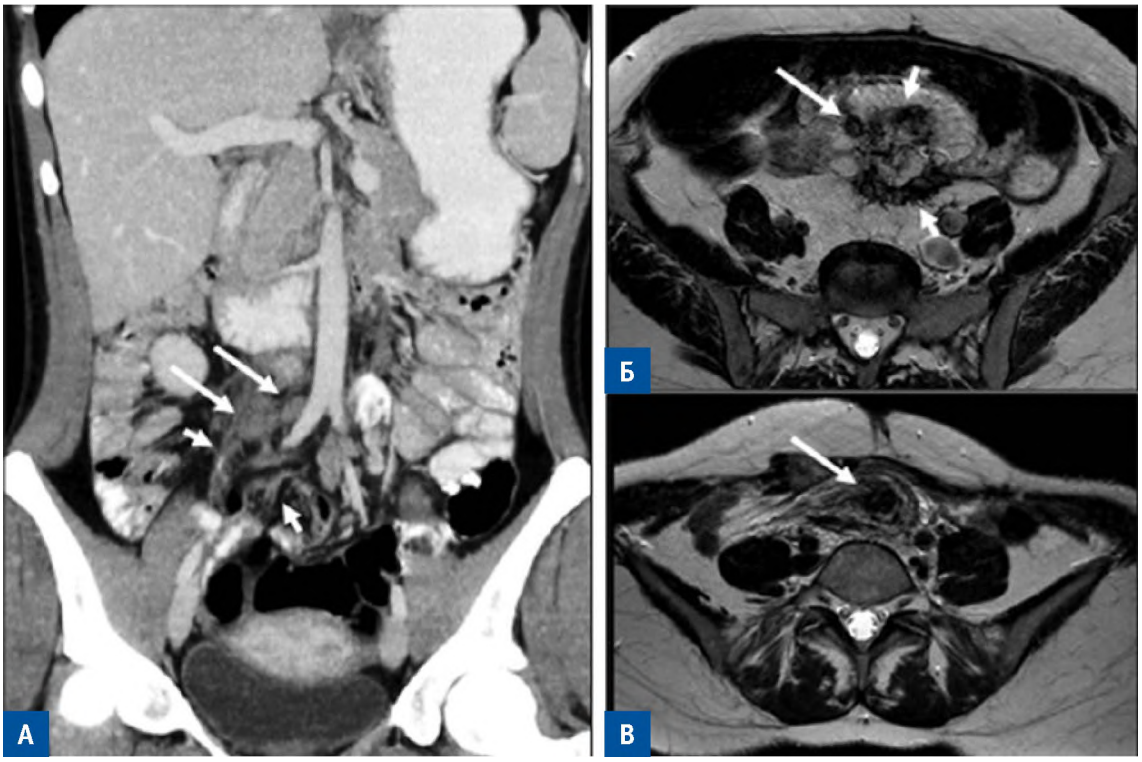

Figure 2. *Combination of infiltrative and nodal form of desmoid-type fibromatosis of the small intestine mesentery. А — CT in the coronal projection of patient Sh., 26 years old; infiltrative (short arrows) and nodal (long arrows) DF components. Б, В — MRI in the axial projection, T2-WI; Б — tomogram of patient S., 47 years old; a mass up to 1 cm (long arrow) against the background of diffuse infiltration of the mesentery involving the loop of the small intestine (short arrows); В — tomogram of patient X, 37 years old; a hypointense mass up to 1.5 cm against the background of diffuse infiltration of the root of the small intestine mesentery*

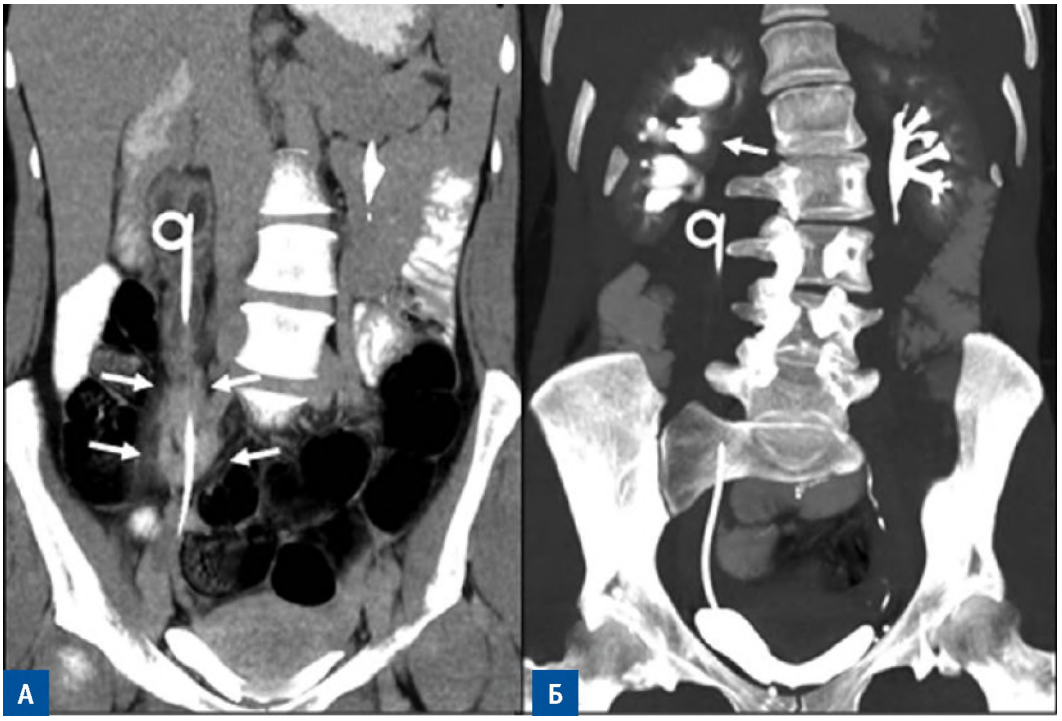

Figure 3. *CT with intravenous contrasting, patient Ch, 36 years old. А — coronal projection; infiltrative growth of DF (arrows) involving the right ureter with a stent placed inside; Б — MIP-reconstruction; hydronephrotic transformation of the pyelocaliceal system of the right kidney (arrow)*

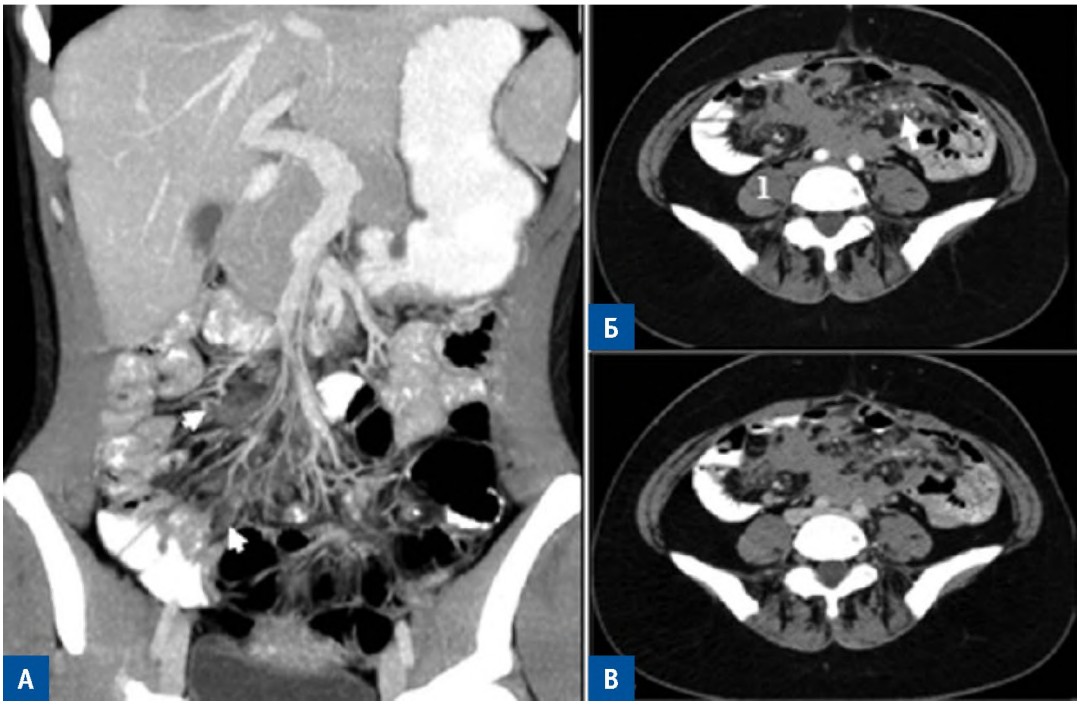

Figure 4. *Infiltrative form of desmoid-type fibromatosis of the small intestine mesentery. CT with contrast enhancement. А — coronary tomogram of patient. K., 26 years old, VIP-reconstruction; infiltrative growth of DF (arrows) along the mesenteric vessels. Б, В — axial tomograms of patient Ts., 50 years old; DF of the mesentery and the root of the small intestine mesentery with the involvement of small intestine loops and mesenteric vessels (arrow); Б — arterial phase; no accumulation of contrast agent, DF density close to muscle density (1); С — venous phase; slight accumulation of contrast agent*

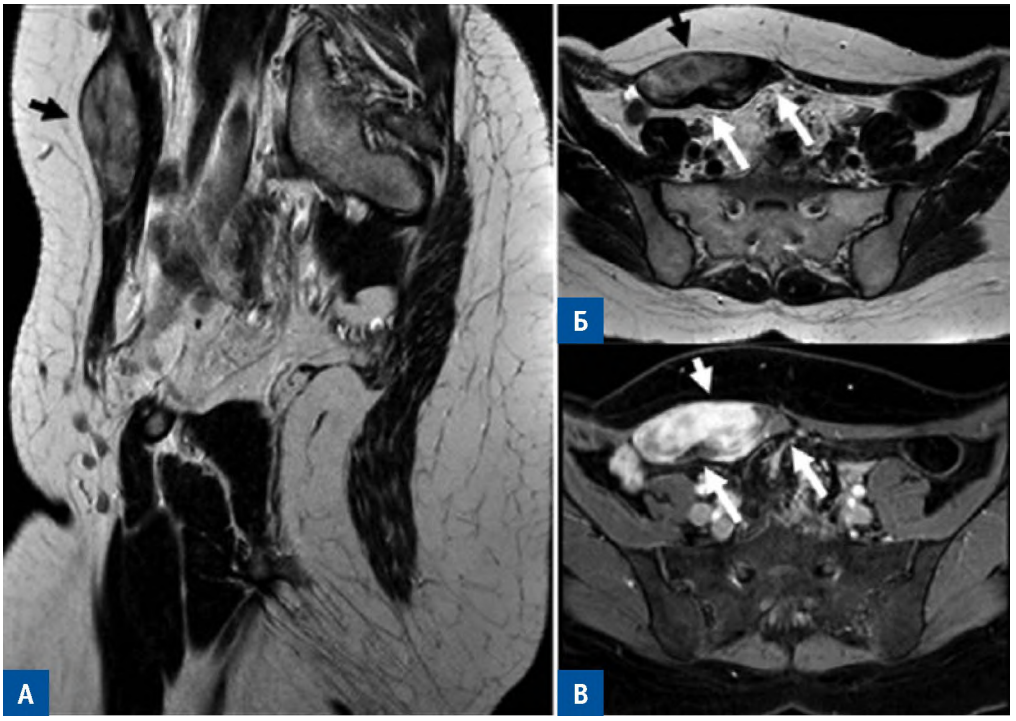

Figure 5. *Nodal form of desmoid-type fibromatosis of the anterior abdominal wall. Magnetic resonance imaging with intravenous contrasting, patient Kh., 37 years old. А, Б — T2-WI in sagittal (А) and axial (Б) projections; the tumor is clearly delimited from the surrounding tissues; the structure is heterogeneous; the hyperintense signal relative to muscle tissue predominates (black arrows) with areas of hypointense signal (white arrows). В — post-contrast T1-WI with adipose suppression; axial projection; clear enhancement of contrast agent in the zone of hyperintense T2-WI signal (short white arrow) and insignificant in the zone of hypointense T2-WI signal (long white arrows)*

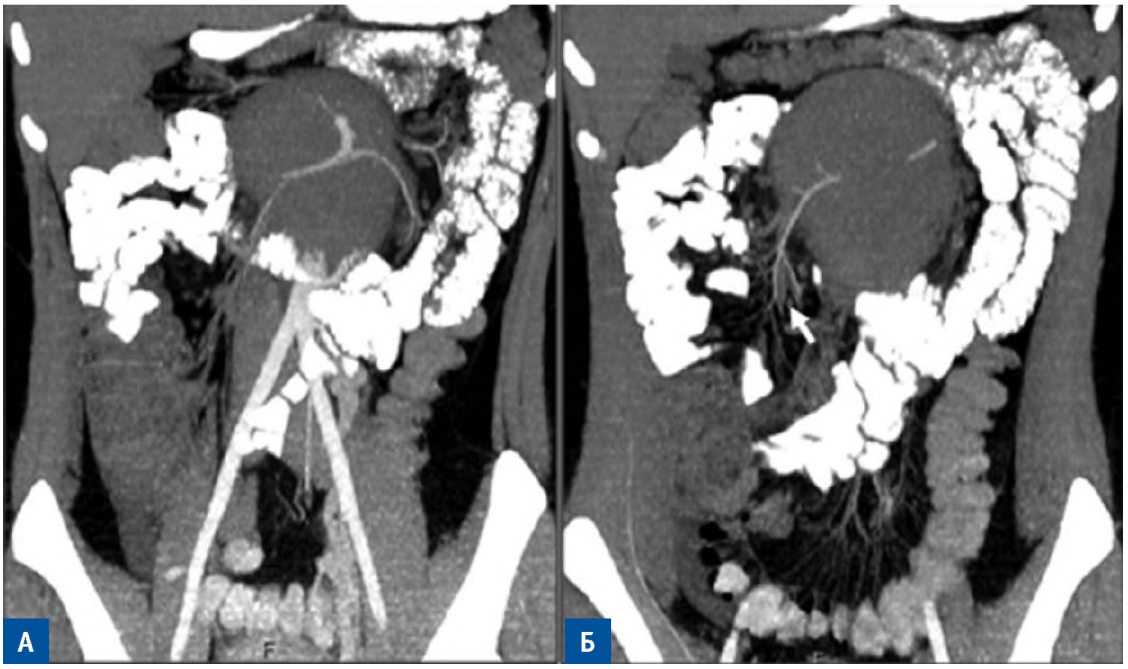

Figure 6. *Nodal form of desmoid-type fibromatosis of the small intestine mesentery. Computed tomography with intravenous contrast enhancement of the patient A., 38 years old, MIP-reconstruction in the coronal projection. А — mesenteric vessels are pushed forward and 'flattened' on the tumor; Б — infiltrative tumor growth along the vessels (arrow)*

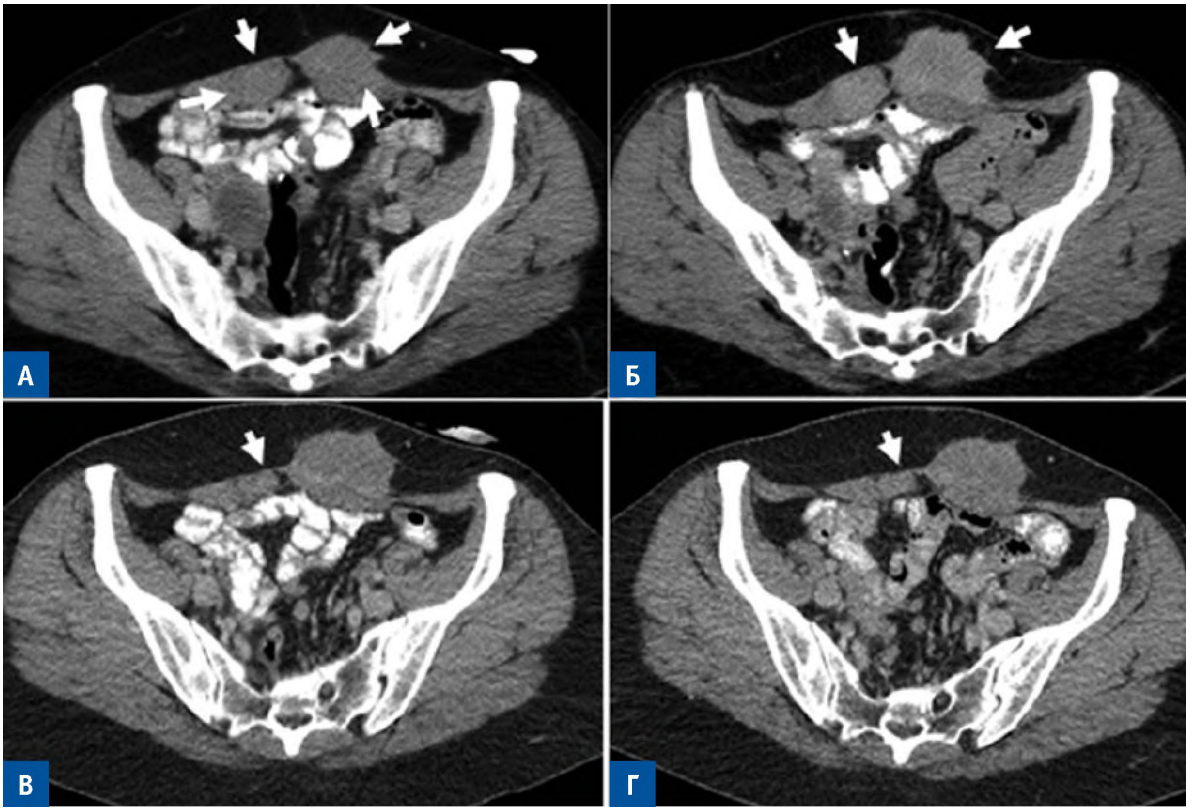

Figure 7. *Desmoid-type fibromatosis of the anterior abdominal wall. CT with intravenous enhancement in patient Y., 36 years old. А — Tumors of the anterior abdominal wall in the area of the postoperative scar (arrows) with dimensions of 3.5 × 2.0 cm and 2.0 × 1.8 cm, 'merging' in density with muscle tissues. Б — after 6 months, there was an increase in the size of the masses up to 4.9 × 3.1 cm and 3.0 × 2.2 cm, respectively, and increased accumulation of the contrast agent compared to the muscles. В — 6 months after chemotherapy (methotrexate, vinorelbine), the tumor on the right (arrow) decreased by more than two times; the size of the second tumor did not change. Г — after 6 months of CT images did not change significantly*

(2/35, 6.7%), uterus (2/35, 6.7%), ovaries (1/35, 2.8%) (Fig. 2). Involvement of the ureters required stenting in two cases due to the hydronephrosis (Fig. 3). Against the background of infiltrative changes, nodal formations were detected in the amount from 1 to 9, with sizes from 1.5 to 5 cm.

The infiltrative form was detected in 6 (17.1%) patients and was located in the mesentery of the small intestine, accompanied by infiltrative involvement of the loops of the small intestine (6/35, 17.1%), mesenteric vessels (2/35, 6.7%) and pelvic peritoneum (1/35, 2.8%) in combination with infiltrative pelvic desmoid (Fig. 4).

The nodal form of desmoid tumor growth was revealed in 13 (37.1%) patients, in 8 of these cases in combination with infiltrative and nodal DF of the mesentery of the small intestine. In 9 (25.7%) cases, nodal formations were detected in the anterior abdominal wall, in 3 (8.6%) cases — in the mesentery of the small intestine and in 1 (2.8%) — in the retroperitoneal space. The number of nodal formations ranged from 1 to 4, sizes from 2 cm to 25 × 10 cm (Fig. 5). Two-thirds of the patients had tumor invasion into adjacent tissues, in the remaining patients the tumor nodes were clearly separated from the surrounding tissues. In one case, a large (12 × 10 × 9.5 cm) desmoid tumor with displacement and compression of mesentery vessels was detected in the mesentery of the small intestine, which required removal of the tumor (Fig. 6).

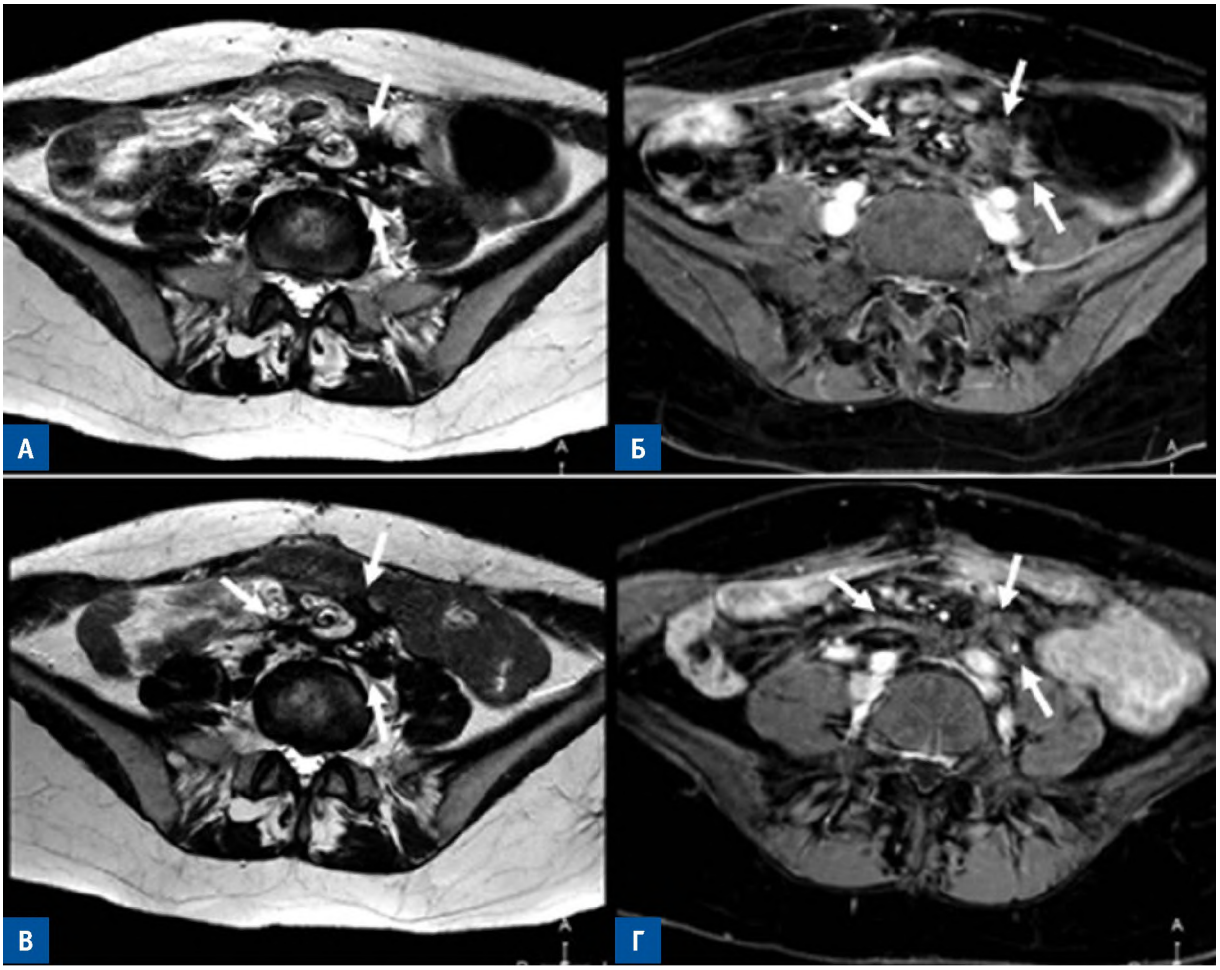

Figure 8. *MRI with intravenous enhancement in patient D., 33 years old. Axial projections. A, B — T2-WI; Б, Г — post-contrast T1-WI with adipose suppression. Infiltrative DF of the root of the small intestine mesentery; hypointense MR signal (A) with a slight accumulation of contrast agent (Б) (arrows). When examining 12 months later, the area of the lesion, the intensity of the MR signal on T2-WI (B) and post-contrast T1-WI (Г) did not change (arrows)*

Twenty-five patients (25/35, 71.4%) were re-examined using CT (13/35, 37.1%) and MRI (12/35, 34.3%), including after systemic therapy or during treatment.

Computed tomography revealed an increase in the size of desmoid formations in 5 patients (with the appearance of a new node in one case), and subsequent imaging during systemic therapy showed downsizing of tumors in three of them (Fig. 7). In the remaining 8 patients, under follow-up, the size of the DF remained the same (5 patients) or smaller (3 patients). CT density and structure of formations, the nature and degree of accumulation of contrast medium did not change throughout the follow-up in most patients. Only in two patients, the increase in desmoid tumor size was accompanied by the appearance of heterogeneity of the structure and a moderate increase in the accumulation of contrast agent.

With MRI in 7 patients, the picture remained the same throughout the entire follow-up: the size of the DF did not change, which was accompanied by the preservation of a hypointensive signal on T2-WI, the absence of contrast or a slight accumulation of contrast agent on T1-WI with adipose suppression (Fig. 8). In two cases, the size of the DF decreased, which was combined with a decrease in the intensity of the MR signal on T2-WI and on post-contrast T1-WI with adipose suppression. In three cases, an increase in the size of the tumor was revealed, while an inhomogeneous structure

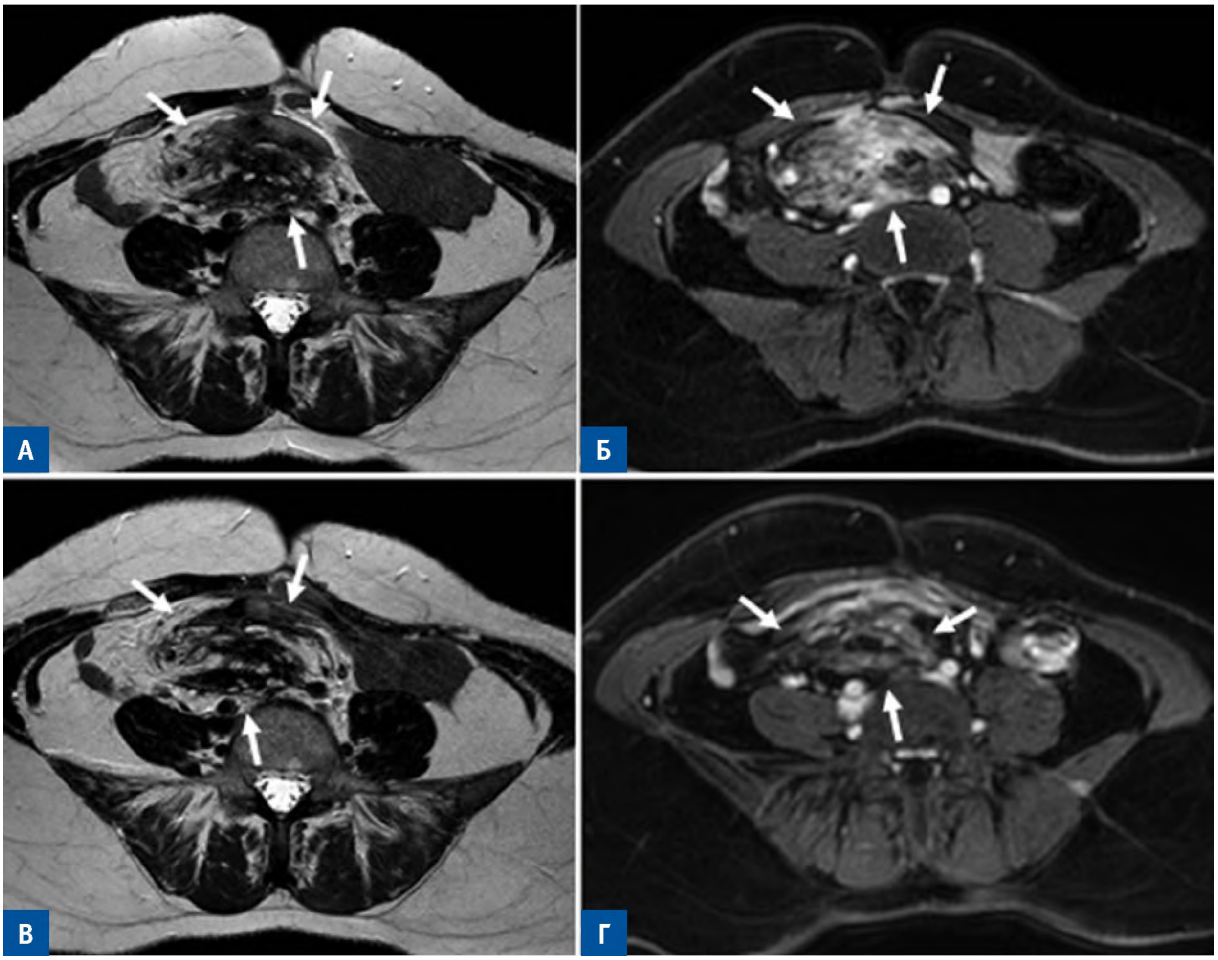

Figure 9. *MRI with intravenous contrasting in patient Y., 39 years old. Axial projections. А, В — T2-WI; Б, Г — post-contrast T1-WI with adipose suppression. Infiltrative DF root of the small intestine mesentery; heterogeneous MR signal (А) with pronounced accumulation of contrast agent (Б) (arrows). In the study 12 months after chemotherapy, the area of the lesion did not change, there was a predominance of a hypointense MR signal (В) with a slight accumulation of a contrast agent (Г) (arrows)*

of the formation was revealed with a predominance of a hyperintensive signal on T2-WI compared to the previous study and a pronounced accumulation of a contrast agent. With further follow-up, two of these three patients showed a positive response to the therapy: the size of the desmoids remained the same, but a decrease in the intensity of the MR signal on T2-WI was determined with the appearance of hypointensive zones, the accumulation of contrast agent was moderate, insignificant in some areas (Fig. 9).

DISCUSSION

The risk of desmoid tumors in patients with FAP may be associated with a number of factors, such as female gender, abdominal surgery, the family history of desmoid fibroids, mutations in the *APC* gene of a certain location [3,5,26–31]. FAP-associated DFs in most cases develop within 5 years after surgery [27]. In this study, two-thirds of the patients were women and, in the majority of cases (30/35, 85.7%) desmoids were revealed in 2–4 years after colectomy. In most cases (33/35, 94.4%) with CT and MRI, desmoid tumors were detected in the mesentery and mesentery root of the small intestine, and in every fourth there was a combination of intra-abdominal site of desmoids with lesion to the anterior abdominal wall.

This coincides with the literature data on predominantly intraabdominal site of desmoid tumors in FAP, as well as their often combination with extraabdominal DF [3,4,8,11].

When the mesentery of the small intestine was affected, infiltrative-nodal (24/35) and infiltrative (6/35) forms of DF prevailed, with infiltrative growth to adjacent organs and structures, primarily to the small intestine and mesenteric vessels. A number of papers inform that such changes can lead to intestinal obstruction and compression of vascular structures [11,16,21]. When the ureters are involved in the tumor, hydronephrosis may develop [14,15,23]. In the study, the spread of DF from the mesentery root to the ureter in two cases required ureter stenting due to the hydronephrotic transformation of the kidney. The nodal form of desmoids was revealed in 13 (37.1%) patients, including 8 cases in combination with infiltrative-nodal DF of the mesentery of the small intestine. In most cases, nodal formations were located in the anterior abdominal wall. The presence of a large nodal DF in the mesentery of the small intestine developed displacement and compression of the mesentery vessels, which required surgical removal of the tumor.As for the diagnostic capabilities of CT and MRI in detecting DF in FAP, some authors believe that both methods can be used to determine the location and extent of the tumor [3, 11], others prefer MRI, especially in young patients to exclude radiation, and consider it appropriate to use CT only to detect complications [16,24]. According to the data obtained, both methods made it possible to visualize desmoid formations both in the mesentery of the small intestine and in the anterior abdominal wall, enabled to assess the extent of the process and the nature of the involvement of adjacent organs and structures. When the infiltrative form of DF was located in the pelvis, MRI had an advantage in differential diagnosis between the tumor and postoperative fibrous changes. Given the predominantly intra-abdominal location of DF in FAP, from our point of view, it is advisable to use CT for the primary detection of desmoids in these patients. This is due to the short scanning time (30–60 seconds), the ability to simultaneously visualize the organs of the peritoneal cavity and pelvis, vessels, as well as intestinal obstruction and excretory function of the kidneys when the ureter is involved in the process. MRI requires a longer time (20–40 min.) and, as a rule, separate scanning of the abdominal cavity and pelvic organs.

In addition to detecting DF and determining the extent of changes, imaging methods are used for dynamic control and evaluation of the tumor response to systemic therapy. It is believed that in CT to determine the changes of the tumor process, two parameters are important — the size of the DF and its density (densitometric indicators in the study without intravenous contrast) [11]. In a follow-up of 13 patients using CT, only in two patients an increase in DF was accompanied by a change in the density of formation and a moderate increase in the accumulation of contrast agent. In all other cases, the density indicators did not change when increasing, decreasing or maintaining the same dimensions of the DF. As for the accumulation of contrast agent in the tumor during intravenous contrast, it was insignificant in most patients and did not change significantly during repeated imaging, with the exception of two cases.

Thus, the main parameter for assessing the changes of the tumor in CT in the study was the size of the tumor.

MRI has great capabilities in determining the DF structure due to the high soft-tissue contrast. The signal intensity on T2-WI and post-contrast T1-WI reflect the ratio of cellular and fibrous components of the tumor [11,22]. Most often, in MRI images, desmoids have an inhomogeneous structure with an isointensive or slightly hyperintensive relative to muscle tissue signal on T2-WI. A decrease in the signal intensity on T2-WI is associated with a decrease in the number of spindle cells and an increase in the number of collagen fibers [11]. Thus, the intensity of the MR signal on T2-WI is an important characteristic of DF [11,21,22,24,32,33]. A tumor downsizing and the hyperintensivity of the signal on T2-WI indicate a positive response to systemic therapy [14,20]. It is also believed that desmoids with a high content of spindle-shaped cells actively accumulate a contrast agent with intravenous gadolinium-containing contrast agents [11,16]. A correlation was revealed between the hyperintensivity of the T2-WI signal and the degree of accumulation of contrast agent by a desmoid tumor [24]. These parameters, along with the size of the tumor, allow a more detailed assessment of the changes of the tumor process during repeated imaging, including during systemic therapy. In the study, dynamic control using MRI was performed in 12 patients. At the same time, in a number of patients, during the first and subsequent imaging, a homogeneous hypointensive signal was detected on T2-WI, which was accompanied by a lack of contrast or a slight accumulation of contrast agent on post-contrast T1-WI with adipose suppression. The revealed MR indicators, in our opinion, show a clear predominance of the fibrous component of the tumor and clinically corresponded to the stabilization phase. In two patients during treatment, a decrease in the size of the DF was noted in combination with a decrease in the intensity of the MR signal on T2-WI and on post-contrast T1-WI with adipose suppression, which was regarded as a positive response to the therapy. The progression in three cases was accompanied by an increase in the size of the DF, with a predominance in comparison with the previous study of the hyperintensive signal on T2-WI and a clear accumulation of the contrast agent. It should be said that MRI, unlike CT, allows us to judge the changes of the tumor process even in the absence of size changes in DF [11]. Thus, during further follow-up, two out of three patients with DF progression showed a positive response to the therapy: the size of desmoids remained the same, but there was a decrease in the intensity of the MR signal on T2-WI with the appearance of hypointensive zones, and the accumulation of contrast agent became moderate, insignificant in some areas.

CONCLUSION

CT and MRI are the basic imaging methods for detecting desmoid fibroids in familial adenomatous polyposis, allowing to determine the nature of tumor growth, assess the extent of the tumor process and the involvement of adjacent organs and structures. In dynamic control and evaluation of the response of a desmoid tumor to systemic therapy, MRI has greater diagnostic capabilities compared to CT, since it takes into account not only the size of the desmoid, but also the intensity of the MR signal on T2-WI and the nature of the accumulation of contrast agent on post-contrast T1-WI with adipose suppression.

AUTHORS CONTRIBUTION

Concept and design of the study: *Irina V. Zarodnyuk, Alexander M. Kuzminov, Dmitry Yu. Pikunov*
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