CASE REPORT

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Rare solitary fibrous tumor of the stomach: A case report

Redak solitarni fibrozni tumor želuca

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Abstract

Introduction. Solitary fibrous tumors are rare soft tissue tumors of submesothelial origin and variable malignant potential. The most common localization is pleural, whereas only 0.6% are of extrapleural localization. Solitary fibrous tumor of the peritoneum, especially of gastric serosa is an extremely rare form of this tumor. **Case report.** We presented a 65-year-old female patient with solitary fibrous tumor of the stomach. Histopathological analysis of removed tissue showed the presence of tumor tissue built of spindle cells, elongated nuclei with moderately abundant cytoplasm. Cells were in a noncohesive arrangement, in smaller areas distributed in the form of palisade. There were amounts of hipocellular connective tissue, hyalinised, with small foci of dystrophic calcification. Mitoses were rare (less than 3/10 HPF). Blood vessels surrounded the connective tissue. Reviewed material did not contain elements of the parent organ. Immunohistochemically there were positivity on CD34 and vimentin, and negativity to S100, SMA, CD117, dezmin, and Ki-67 is < 2%. The change was diagnosed as a solitary fibrous tumor. **Conclusion.** Considering that benign solitary fibrous tumors of extrathoracic localizations are extremely rare neoplasms with unpredictable biological behavior and the possibility of recurrence, a long-term clinical and endoscopic follow-up on yearly basis of patients with this disease is recommended.

Key words:
fibroma; peritoneal neoplasms; stomach; diagnosis; immunoenzyme techniques; tumor markers, biological; antigens, cd 34; vimentin; immunohistochemistry.

Apstrakt

Uvod. Solitarni fibrozni tumori su retki mekotkivni tumori submezotelijalnog porekla, sa varijabilnim malignim potencijalom. Najčešće su pleuralne lokalizacije, dok je svega 0,6% smešteno ekstrapleuralno. Solitarni fibrozni tumor peritoneuma, posebno gastrične seroze, veoma je retka forma ove vrste tumora. **Prikaz bolesnika.** Prikazana je bolesnica stara 65 godina, sa solitarnim fibroznim tumorom želuca. Patohistološka analiza otklonjenog tkiva pokazala je postojanje tumorskog tkiva sagrađenog od vretenastih ćelija, izduženih jedara sa srednje obilnom do oskudnom citoplazmom. Tumorske ćelije bile su u nekohezivnom rasporedu, u manjim područjima raspoređene u vidu palisada. Između opisanih tumorskih ćelija bila je prisutna veća količina hipocelularnog vezivnog tkiva koje je mestimično bilo hijalinizovano, sa veoma malim fokusima distrofijskih kalcifikata. Mitoze su bile veoma retke (manje od 3/10 HPF). Bili su prisutni i krvni sudovi koje je okružavalo vezivo. U pregledanom materijalu nije bilo elemenata ishodišnog organa. Imunohistohemijski, dobijena je pozitivnost na CD34 i vimentin, negativnost na S100, SMA, CD117, dezmin, a Ki-67 je < 2%. Promena je dijagnostikovana kao solitarni fibrozni tumor. **Zaključak.** S obzirom na činjenicu da su benigni solitarni fibrozni tumori sa ekstratorakalnom lokalizacijom ekstremno retke neoplazme sa nepredvidivim biološkim ponašanjem i mogućnošću ponavljanja, preporučuje se dugoročno kliničko i endoskopsko praćenje na godišnjem nivou kod bolesnika sa tim oboljenjem.

Ključne reči:
fibrom; peritoneum, neoplazme; želudac; dijagnoza; imunoenzimske tehnike; tumorski markeri, biološki; antigeni, cd 34; vimentin; imunohistohemija.

Introduction

Solitary fibrous tumors are a rare group of spindle cell neoplasm which includes fibrous-fiber cell solitary fibrous tumors and associated lesions such as giant cell lipomatous hemangiopericytoma and angiofibroma [1]. Initially described solitary fibrous tumors were of pleural origin localized in the chest cavity [2]. Today are described extrathoracic localiza-

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tions of this tumor, such as: retroperitoneal, abdominal cavity, the head and neck [3–5]. This type of tumor most frequently occurs in middle-aged people of both sexes [6]. Extrathoracic solitary fibrous tumors, unlike thoracic, are generally followed by symptoms in the form of pain or pressure, depending on the size and location of the tumor mass [4, 7]. Solitary fibrous tumors, of any localization are usually of benign origin and surgical resection is curative therapy for almost the majority of these tumors [8]. Solitary fibrous tumors can recur, and metastasize after surgical resection, because in some cases the biological behavior of the tumor does not correlate with histopathology, and therefore the long-term clinical follow-up is mandatory for all patients [6]. These tumors independent on their localizations, appears to have a common characteristic of immune reactivity to the CD34.

So far, in the available literature there are only a three cases of solitary fibrous tumor with extrathoracic localization, in the area of gastric serosa [9–11].

Case report

After preoperative examination a 65-year-old female patient was operated at the Clinic for Abdominal, Endocrine and Transplantation Surgery, in the Clinical Center of Vojvodina, Novi Sad, Serbia. Specimen removed was a spherical formation with referral clinical diagnosis- "Gastrointestinal stromal tumor" and sent to histopathological examination. By macroscopic examination received material was described in the form of spherical fragment size 2.5 × 2.3 × 1 cm, smooth, pale yellowish surface, on serial sections swirling material, white grayish color. The material was processed in its entirety.

After routine histological processing of materials: fixed in 10% formalin, dehydrated, embedded in paraffin, cut to a thickness of 4 micrometers, hematoxylin and eosin staining method, histological analysis were performed. Tumor tissue built of spindle cells with elongated nuclei and with moderate abundant to scant cytoplasm was observed by histological analysis of the samples. Tumor cells were scheduled in a non-cohesive arrangement in a small areas and in the form of the palisade. Between described tumor cells there was a large amount of hipocellular connective tissue, hyalinised, with very small foci of dystrophic calcification. Mitoses were very rare (less than 3/10 HPF). There were also blood vessels surrounding the connective tissue. Reviewed material did not contain elements of the parent organ, in one of the clips the tumor was surrounded by a muscle layer which might originate from the wall of the parent organ (Figure 1).

After standard staining methods, special immunohistochemical staining methods to S100, SMA, CD117, Desmin, CD34, Vimentin, DOG1, PDGFRA and Ki-67 were done (Table 1). The following immunoprofile was obtained: S100-, SMA-, CD117 – (Figure 2), desmin-, DOG1- (Figure 3), PDGFRA- (Figure 4), CD34 + (Figure 5), vimentin + (Figure 6), Ki-67+ < 2% (Figure 7).

Described histological structure of the tumor tissue was diagnosed as a benign mesenchymal tumor, with respect to the histological description and the obtained immunohistochemical profile it was in favor of solitary fibrous tumor.

Table 1

Applied antibodies for immunohistochemistry

Antibody	(Clone)	Source	Dilution	Result
CD34	(QBEnd 10)	DAKO, Carpinteria,CA,USA	1:40	+
Vimentin	(V9)	Fremont, CA94538-6406, USA	1:50	+
Desmin	(D33)	Fremont, CA94538-6406, USA	1:50	-
DOG1	(rabbit monoclonal)	Termo Fisher Scientific Inc.Waltham, USA	1:25	-
PDGFRA	(rabbit polyclonal)	Termo Fisher Scientific Inc.Waltham, USA	1:100	-
S-100	(rabbit polyclonal)	Fremont, CA94538-6406, USA	1:300	-
CD-117	(rabbit polyclonal)	Fremont, CA94538-6406, USA	1:100	-
SMA	(alfa-sm-1)	Novocastra,Newcastle, UK	1:50	-
Ki-67	(rabbit polyclonal)	Fremont, CA94538-6406, USA	ready-to-use	+ < 2%

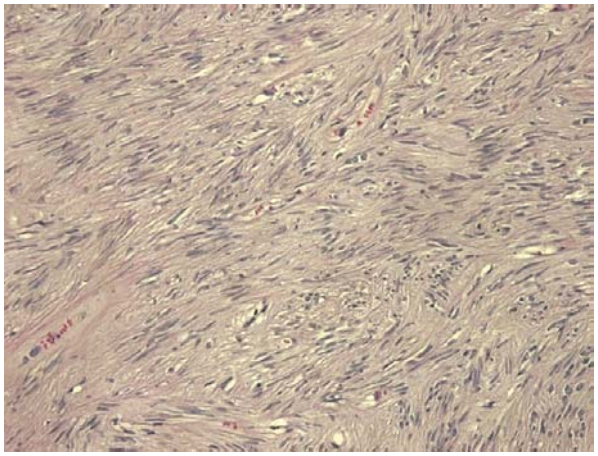

Fig. 1 – Tumor is surrounded by muscle layer that may originate from the wall of the parent organ (HE, ×100).

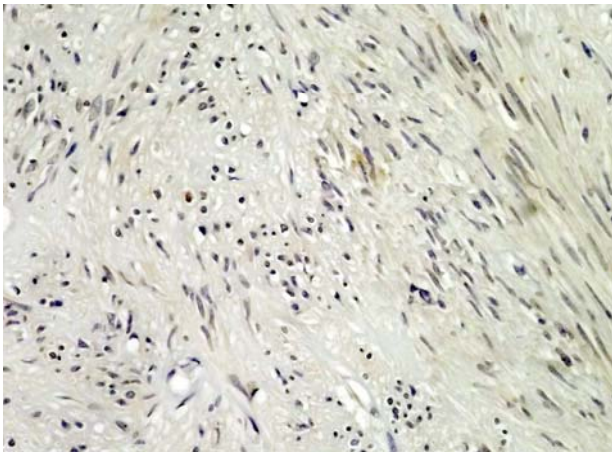

Fig. 2 – Tumor tissue (immunohistochemical stain for CD 117, ×400).

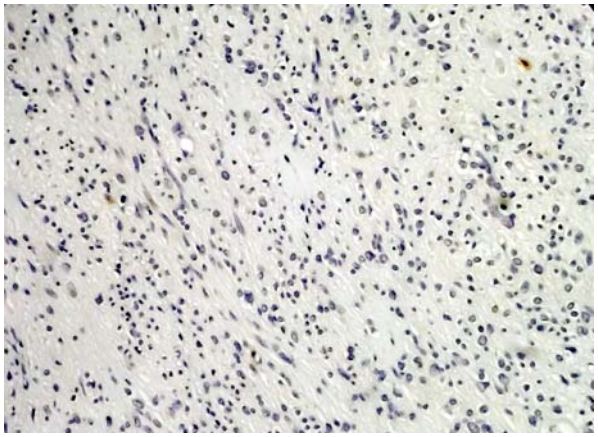

Fig. 3 – Tumor tissue (immunohistochemical stain for DOG1, ×400).

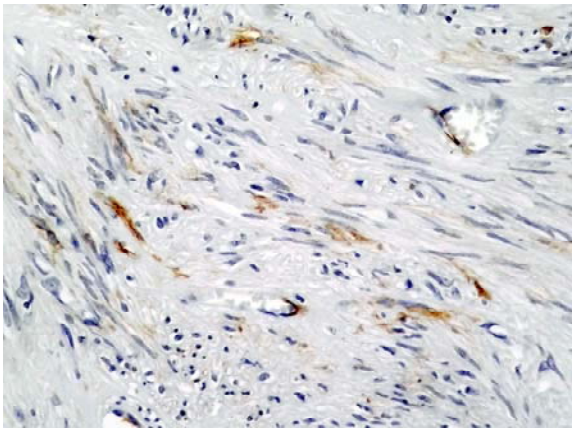

Fig. 4 – PDGFRA immunoreactivity of resected tumor (×400).

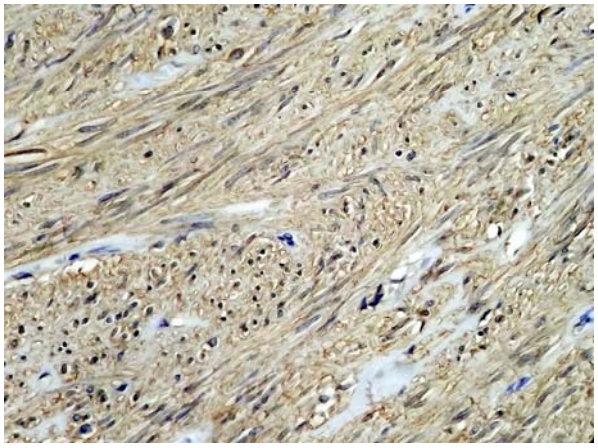

Fig. 5 – Immunohistochemical feature of the tumor: CD34 positivity in tumor cells – nuclear positivity 20% (×400).

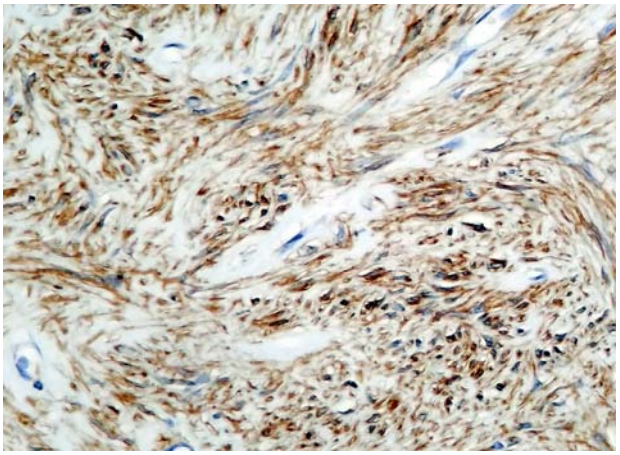

Fig. 6 – Tumor cells showing immunohistochemical positivity for vimentin: cytoplasmic positivity 60% (×400).

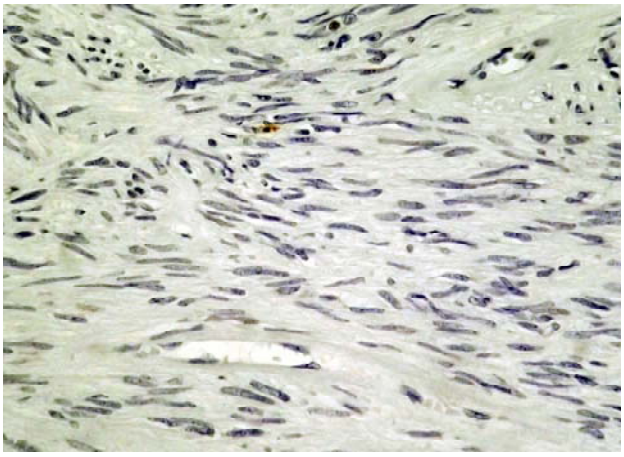

Fig. 7 – Immunohistochemical feature of the tumor: KI-67 positivity in tumor cells – positivity less than 2% (×400).

Discussion

Solitary fibrous tumors are rare forms of the spindle-shaped cell neoplasm, typical pleural origin but also published were the forms of these tumor with extrathoracic localization such as the stomach, retroperitoneum and head and back region [12]. Most solitary fibrous tumors are benign and have a favorable prognosis, although also described in 13–23% cases of pleural localization malignant tumor forms [8].

Solitary fibrous tumors of extrathoracic localization show pronouncedly histological variability [13, 14] as a result of which the differential diagnosis takes into account numerous neoplasms, such as mesenteric tumors, gastrointestinal stromal tumors, tumors of smooth muscles, an in-

flammatory myofibroblastic tumors, inflammatory fibroid polyps, neurofibromas, and many sarcomatoid mesotheliomas and tumors of mesenchymal origin. Immunohistochemical staining in these circumstances are included in the histological diagnosis. Solitary fibrous tumors showed a marked positivity to CD34 and vimentin and negativity for cytokeratin, SMA, desmin, S100, c-kit, DOG1 and PDGFRA [12, 15]. Although the clinical features and biological behavior of these cancers have been described in several publications, precise clinical behavior of the tumor is still unpredictable. The rarely reappears in the form of local recurrence, tissue destruction in the source place spot, even in the form of remote metastasis. Recurrences can occur after several decades of surgical resection, so the long-term clinical follow-up of these patients is necessary [16].

Benign solitary fibrous tumors extrathoracic localizations are extremely rare neoplasms of mesenchymal origin, however, this does not diminish the importance of diagnosis of these neoplasms, especially as recorded their unpredictable biological behavior in the form of recurrence, local destruction at the site of the original impressions, or even in the form of distant metastases [9].

Conclusion

Considering that benign solitary fibrous tumors of extrathoracic localizations are extremely rare neoplasms with unpredictable biological behavior and the possibility of recurrence, a long-term clinical and endoscopic follow-up on the yearly basis of patients with this disease is recommended.

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