CASE REPORTS
PRIKAZI SLUČAJEVA

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CALCIFYING FIBROUS TUMOR IN THE ABDOMEN – A CASE REPORT

FIBROZNI KALCIFIKUJUĆI TUMOR U ABDOMENU – PRIKAZ SLUČAJA

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Summary

Introduction. Calcifying fibrous tumor is a rare benign neoplasm of soft tissue origin. The tumor is commonly found in young adults. In most cases it is an incidental finding, because patients do not have obvious symptoms. This tumor may appear in different anatomical locations mimicking other stromal lesions. The diagnosis is made based on pathohistological characteristics and an appropriate immunohistochemical profile. The treatment is surgical, and the prognosis is good. **Case Report.** A 19-year-old female patient was admitted for abdominal surgery presenting with abdominal pain and pressure. Abdominal ultrasonography and multislice computed tomography of the abdomen showed a tumor mass in the right hemiabdomen. The patient underwent surgical treatment and the tumor was completely removed. Macroscopic analysis showed that the tumor was encapsulated and had a smooth surface. Microscopically, the tumor consisted of bundles of partially hyalinized collagen fibers with calcifications in the form of psammoma bodies that were permeated with mononuclear inflammatory infiltrates. **Conclusion.** Given the higher incidence of other mesenchymal tumors in the abdomen, due to its rare occurrence, calcifying fibrous tumor presents a diagnostic challenge.

Key words: Neoplasms, Fibrous Tissue; Abdominal Neoplasms; Calcinosis; Morphological and Microscopic Findings; Immunohistochemistry; Diagnosis, Differential

Sažetak

Uvod. Fibrozni kalcifikujući tumor je retka benigna neoplazma porekla mekih tkiva. Tumor je češći kod mladih odraslih osoba. U većini slučajeva se otkriva slučajno jer pacijenti nemaju očigledne simptome. Ovaj tumor se može javiti na različitim anatomskim lokalizacijama imitirajući druge stromalne lezije. Dijagnoza se postavlja na osnovu patohistoloških karakteristika i određenog imunohistohemijskog profila. Tretman je hirurški, a prognoza je dobra. **Prikaz slučaja.** Pacijentkinja, 19 godina, primljena je na abdominalnu hirurgiju sa simptomima u vidu bola i pritiska u abdomenu. Ultrazvučnom sonografijom i kompjuterizovanom tomografijom abdomena verifikovano je prisustvo tumorske mase u desnom hemiabdomenu. Pacijentkinja je podvrgnuta hirurškom tretmanu i tumorska promena je otklonjena u celosti. Makroskopskom analizom tumor je inkapsulisan i glatke površine. Mikroskopski, tumor je sačinjen od snopova kolagenih vlakana delom hijalinizovanih sa kalcifikacijama u vidu psamoznih telašaca koji su prožeti mononuklearnim inflamatornim infiltratom. **Zaključak.** S obzirom na veću incidenciju drugih mezenhimalnih tumora u abdomenu, postavljanje dijagnoze fibroznog kalcifikujućeg tumora je delikatno, imajući u vidu njegovu retku pojavu.

Ključne reči: fibrozne neoplazme; abdominalne neoplazme; kalcifikacija; morfološki i mikroskopski nalazi; imunohistohemija; diferencijalna dijagnoza

Introduction

Calcifying fibrous tumor (CFT) is a benign mesenchymal tumor with a low rate of recurrence after removal [1]. Histologically, it should be distinguished from other, more aggressive, rare mesenchymal lesions [2], especially because it can affect soft tissues at different anatomical sites (intra-abdominal and intrathoracic) [1]. We present a case of a 19-year-old woman with a diagnosed CFT in the abdominal cavity to provide more information about this extremely rare entity.

Case Report

A 19-year-old female patient was admitted for abdominal surgery presenting with abdominal pain and pressure**.** On physical examination, the abdomen was at chest level and palpably soft. Abdominal ultrasonography and multislice computed tomography of the abdomen revealed a tumor, 14 cm in maximum diameter in the right hemiabdomen. The patient underwent surgical treatment. Intraoperatively, it was found that the tumor was in contact with the duodenum, head of the pancreas, and the portal vein. It did

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Abbreviations
CFT – calcifying fibrous tumor
IgG – imunoglobulin G

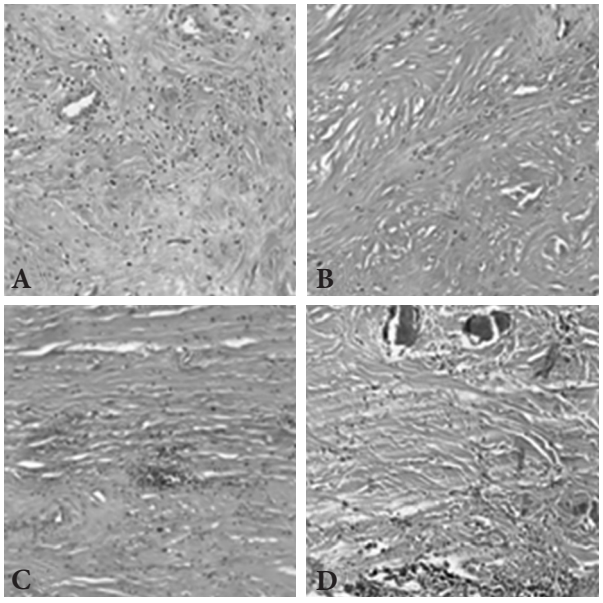

Figure 1. Microscopic appearance of CFT: (a) HE, x 10; (b); HE, x 10; (c) HE, x 10; (d) HE, x10
Slika 1. *Histološki izgled fibroznog kalcificirajućeg tumora: (a) HE, x 10; (b); HE, x 10; (c) HE, x 10; (d) HE, x 10*

not infiltrate the surrounding structures and was removed completely. On gross examination, the tumor was oval, encapsulated, and hard in consistency, with a smooth and shiny surface, measuring 11 x 9 x 7 cm. On cross-section, the tumor had a whirling pattern and it was pale with a darkly colored central area. Microscopic examination with hematoxylin and eosin staining showed circumscribed bundles of collagen fibers partially hyalinized and with sparse areas of inflammatory infiltrates composed of lymphocytes and plasma cells with folliculoid appendages **(Figures 1a, 1b and 1c)**. The bundles of collagen contained rare spindle cells, sparse cytoplasm and elongated nuclei and foci of dystrophic calcifications in the form of psammoma bodies **(Figure 1d).** The immunohistochemical staining showed that the spindle cells were partially positive for bCl2 and CD34, and negative for beta-catenin, STAT6, anaplastic lymphoma kinase, CD117, and DOG1. The plasma cells showed high ratio of IgG4-positive to IgG-positive plasma cells **(Figure 2)**. The proliferation index (% Ki-67 positive cells) was 1%.

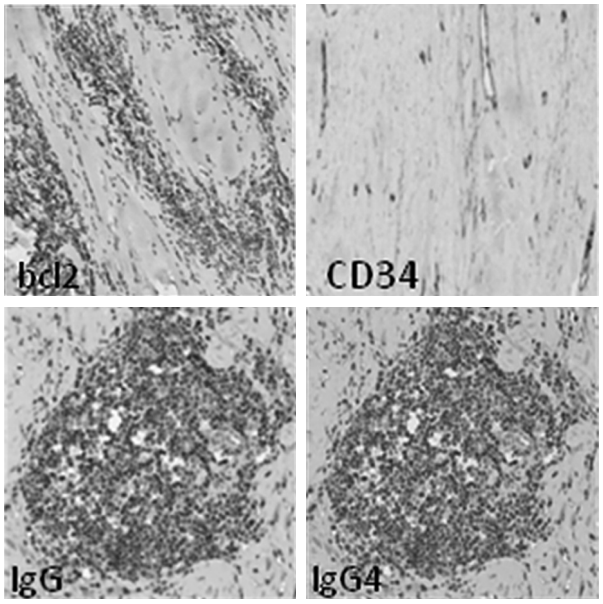

Figure 2. Immunohistochemical analysis showing: (a) bcl2 positivity, HE, x 10; (b) CD34 positivity, HE, x 10; (c) IgG positivity, HE, x 20; (d) IgG4 positivity HE, x 20
Slika 2. *Imunohistohemijska analiza pokazuje: (a) bcl2-pozitivnost, x 10; (b) CD34 pozitivnost, x 10; (c) IgG pozitivnost, x 20; (d) IgG4 pozitivnost, x 20*

According to the histomorphological features and immunophenotype of the tumor cells, the diagnosis of CFT was made. The postoperative course was uneventful and the patient was discharged in good general condition. The patient was followed up for 3 months after surgery and recovered well.

Discussion

The CFT was first described by Rosenthal and Abdul-Karim as a childhood fibrous tumor with psammoma bodies [3], but it was renamed by Fetsch et al., as a calcifying fibrous pseudotumor [4]. In 2002, the World Health Organization established the name for this lesion as CFT, in the new classification of soft tissue tumors that originated from fibroblasts/myofibroblasts [1, 5]. Meta-analysis by Chorti et al. shows a trimodal pattern of age distribution with the first peak from birth to 4 years, the second in the mid-20s, and the third in the mid-30s [1, 2]. It mainly affects women and the average age at the time of diagnosis is 33.58 years [2]. However, our patient was an exception. The etiopathogeny of CFTs is unclear; several theories have been developed about posttraumatic or genetic pathogenesis, as well as a relationship between IgG4-related disease and inflammatory myofibroblastic tumors [2]. Histopathological characteristics of CFT are abundant hyalinized paucicellular collagen, dystrophic calcifications and mononuclear inflammatory infiltrates [2, 6]. Spindle cell mesenchymal lesions and inflammatory conditions are included in the differential diagnosis of CFT [6]. Immunohistochemical analysis of CFT shows uniformly negative staining for anaplastic lymphoma kinase, which is typically positive in inflammatory myofibroblastic tumors [6–8]. Considering the location of the tumor in our case, the use of CD117 immunohistochemistry is important to delineate the gastrointestinal stromal tumor from CFT, because the spindle cells are negative for CD117 in CFTs [6]. Solitary fibrous tumor is immutable CD34 positive as in our case, and reactivity for BCL-2 is not described in CFT [6], but in our case it was positive. In some cases of CFT, the plasma cell population may stain positively for IgG and IgG4

and show increased IgG/IgG4 ratio [8, 9]. High IgG4-to-IgG ratio (41%) supports the view that CFT may be a manifestation of IgG4-related disease (IgG4-RD) [8, 10]. In our case, CFT showed a significant IgG4-postive plasma cell infiltrate and IgG/IgG4 ratio higher than the 40%. However, the major criterion of IgG4-related disease is obliterative phlebitis, but it is not a feature of CFT [6, 8]. The IgG4-related disease usually responds to corticosteroid therapy, but its efficacy in CFT has never been established [6]. Surgical excision is the appropriate treatment for abdominal CFT [11].

Conclusion

Calcifying fibrous tumor is a rare neoplasm of soft tissue origin with benign biological behavior. The diagnosis is based on histological features, because it has no specific clinical and radiological characteristics. We should keep in mind the rare occurrence of this entity and a difficult differential diagnosis, especially when it is localized in the abdomen.

References

1. Prucker J, Salaheddin-Nassr Y, Leidl S. Calcifying fibrous tumor of the terminal ileum mesentery: case report. Medicine (Baltimore). 2018;97(51):e13351.

2. Sabrine D, Hafsa E, Amine R, Zakia B, Fouad Z. Calcifying fibrous tumor of the mesentery: a case report and a review of the literature. Clin Pathol. 2020;13:2632010X20930689.

3. Rosenthal NS, Abdul-Karim FW. Childhood fibrous tumor with psammoma bodies. Clinicopathologic features in two cases. Arch Pathol Lab Med. 1988;112(8):798-800.

4. Fetsch JF, Montgomery EA, Meis JM. Calcifying fibrous pseudotumor. Am J Surg Pathol. 1993;17(5):502-8.

5. Li BJ, Yang XD, Chen WX, Shi YH, Nie ZH, Wu J. Calcifying fibrous tumor of stomach: a case report. Medicine (Baltimore). 2017;96(47):e8882.

6. Larson BK, Dhall D. Calcifying fibrous tumor of the gastrointestinal tract. Arch Pathol Lab Med. 2015;139(7):943-7.

7. Miyashita S, Ryu Y, Takata H, Asaumi Y, Sakatoku M, Seike T, et al. Imaging findings of gastric calcifying fibrous tumour. BJR Case Rep. 2016;2(4):20160064.

8. Turbiville D, Zhang X. Calcifying fibrous tumor of the gastrointestinal tract: a clinicopathologic review and update. World J Gastroenterol. 2020;26(37):5597-605.

9. Baumann KB, Orestes MI, Heaton SM, Whiting RE, Wendzel NC, Foss RD. Calcifying fibrous tumor of the neck. Head Neck Pathol. 2020;14(2):507-11.

10. Zhang H, Jin Z, Ding S. Gastric calcifying fibrous tumor: a case of suspected immunoglobulin G4-related gastric disease. Saudi J Gastroenterol. 2015;21(6):423-6.

11. Hort A, Chen AZL, Moghadam A, Pang T. Calcifying fibrous tumour torsion: a rare cause of abdominal pain. BMJ Case Rep. 2020;13(10):e238220.

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