Peritoneal Multifocal Calcified Fibrous Tumor Detected Incidentally During Cesarean Delivery: Case Report

Sezaryen Esnasında Rastlantısal Saptanan Peritoneal Multifokal Kalsifiye Fibröz Tümör

Yılmaz BAŞ,[a]
Güven GÜNEY,[a]
Pınar UZBAY,[a]
Hande ERENLER BEHİCE,[a]
Çiğdem ÖZDEMİR[a]

[a]Clinic of Pathology,
Çorum Training and Research Hospital,
Çorum

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Yazışma Adresi/*Correspondence:*
Güven GÜNEY
Çorum Training and Research Hospital,
Clinic of Pathology, Çorum,
TÜRKİYE/TURKEY
guvenguney61@hotmail.com

ABSTRACT Calcified fibrous tumor (CFT) is a rare fibroblastic neoplasia of soft tissue origin. Its pathogenesis and etiology are yet to be established. It has a broad anatomical distribution. Clinical signs and symptoms may be confused with those of other fibroblastic tumors of soft tissue origin and any metastatic neoplasia, as well. Histomorphological characteristics of the tumor may resemble other spindle-cell tumors and therefore, its diagnosis may be difficult. Excision of the tumor may be curative when the surgical boundaries are preserved. In a 28-year-old, 37-week pregnant, multiple nodules were detected incidentally within the peritoneum during emergency cesarean delivery. There were a total of 20 nodules with sizes ranging from 0.2 cm to 0.8 cm in diameter. In histopathological examination, we observed rare calcifications and lymphoid aggregates within a fibrosclerotic and fibroelastic stroma. In immunohistochemistry analysis, the tumor cells were diffuse positive for vimentin and focal and weak positive for SMA and CD34. We aimed to present our observations regarding this patient who diagnosed to have CFT upon histological and immunohistochemistry findings since this rare clinical-pathological entity may be confounded with benign and malignant tumors including metastasis.

Key Words: Peritoneal neoplasms; incidental findings

ÖZET Kalsifiye fibröz tümör (KFT) yumuşak dokunun nadir bir fibroblastik tümörüdür. Patogenezi ve etiyolojisi henüz tanımlanamamıştır. Geniş bir anatomik dağılıma sahiptir. Klinik olarak gerek yumuşak dokunun diğer fibroblastik tümörleriyle gerekse metastatik bir neoplaziyle karıştırılabilir. Histomorfolojik olarak diğer iğsi hücreli tümörlere benzerdir ve bu nedenle tanı güçlüğü doğurabilir. Cerrahi sınırlar korunarak yapılan basit eksizyon küratif olabilmektedir. 28 yaşında 37 haftalık gebe bir kadında acil sezaryenle doğum esnasında, peritonda, tesadüfi olarak çok sayıda nodül saptandı. Makroskobik olarak en büyüğü 0,8 cm ve en küçüğü 0,2 cm çapında 20 adet nodül tespit edildi. Histolojik olarak hiyalinize fibrosklerotik ve fibroblastik bir stromada seyrek kalsifikasyonlar ve lenfoid agregatlar izlendi. İmmünohistokimyasal olarak vimentin yaygın pozitif, SMA ve CD34 fokal ve zayıf pozitif boyandı. Biz, histolojik ve immünohistokimyasal bulguları ile KFT tanısı alan bu vakayı metastaz dahil benign ve malign neoplaziler ile karışması ve nadir bir klinikopatolojik antite olması nedeniyle yayınlamayı amaçladık.

Anahtar Kelimeler: Peritoneal tümörler; tesadüfi bulgular

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Calcified fibrous tumor (CFT) is seen mostly in childhood and early adulthood with its pathogenesis and etiology being uncertain. It has a broad anatomic distribution including subcutaneous and deep soft tissues. Clinical signs and symptoms may be confused with those of other fibroblastic tumors of soft tissue origin and any metastatic neoplasia, as well.[1]

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Histomorphological characteristics of the tumor may resemble other spindle-cell tumors and therefore, its diagnosis may be difficult. Resection of the tumor is adequate for treatment. Local recurrence is rare.

CASE REPORT

CLINICAL PRESENTATION

A 28-year-old, 37-week pregnant was taken to the operating room for an emergency cesarean delivery. During the operation, multiple nodules were found incidentally within the peritoneum, which aroused suspicion for presence of metastatic lymphoid involvement. A sample excised for frozen section was diagnosed as benign fibrotic nodule. The other nodules were determined by palpation, all were excised and referred to histopathological examination in 10% formaldehyde solution. The patient had no additional history of another operation or trauma except for her previous cesarean delivery. Total blood count and other laboratory findings were within normal ranges. She had not undergone any radiographic investigation during pregnancy. No recurrence occurred within a follow-up time of 15 months after the cesarean delivery.

PATHOLOGICAL FINDINGS

Macroscopically, a total of 20 oval-shaped, cream-colored, solid masses with sizes ranging from 0.2 cm to 0.8 cm in diameter were extracted from a 11x9x1 cm sized peritoneal fatty tissue. Sectional surfaces were solid and were of cream-colored fibrotic appearance.

Microscopically, the lesion is non-encapsulated, well-circumscribed and surrounded by single-row coelomic epithelium in parts. They have a hypo-cellular stroma comprised of fibroblasts and hyalinized fibrosclerotic collagen fibers. The stroma contains lymphoplasmocytic cell reaction, lymphoid aggregates, rare psammomatous and dystrophic calcifications condensing mostly in perivascular area (Figure 1-4). Mitosis, necrosis and atypical cells were not observed. Tumor cells were positive for vimentin in immunohistochemistry and focal weak positive for smooth-muscle actin. Coelomic epithelium was positive for pan cytokeratin and lymphoid elements were positive for LCA. Tumor cells were negative for FVIIIa, CD 34, CD 68, Desmin, S100, CD117, CD31, NSE and ALK-1.

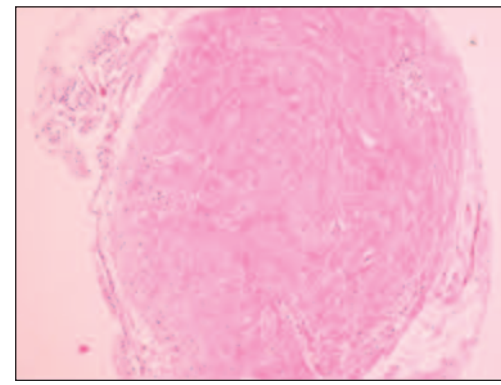

FIGURE 1: Well-circumscribed nodule having a hypocellular stroma (HE, x40).

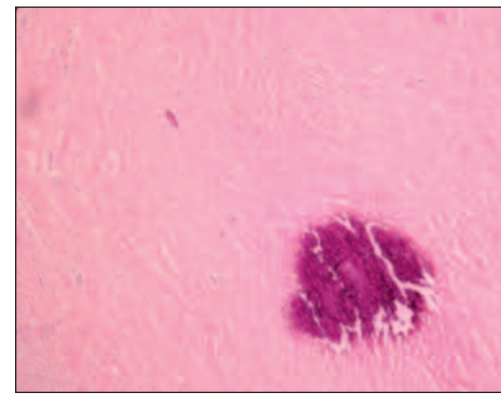

FIGURE 2: Dystrophic calcification within fibrosclerotic collagenous stroma (HE, x200).

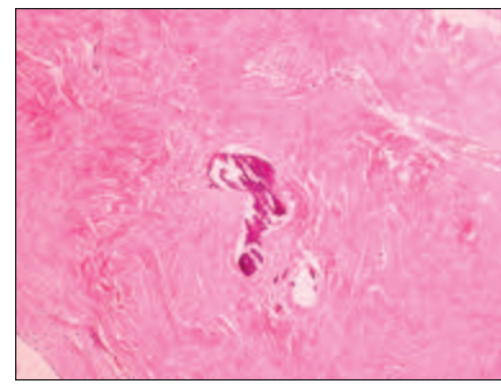

FIGURE 3: Psammomatous calcification within fibrosclerotic collagenized stroma (HE, x400).

DISCUSSION

CFT, which is a rare benign tumor of soft tissue origin was first described in 1988 by Rosenthal and

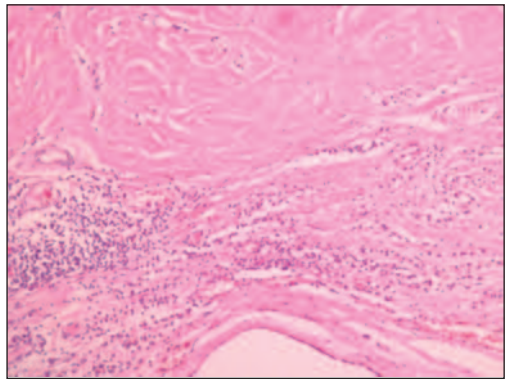

FIGURE 4: Inflammatory cells forming lymphoid aggregate in stroma (HE, x200).

Abdul-Karim in a report of two cases that was entitled "Childhood fibrous tumor with psammoma bodies".[2] The term "Calcified fibrous pseudotumor" was first used by Fetsch et al. in a series of 10 patients aged from 1 to 33 years old.[3] Fetsch et al. suggested that the lesion which develops following a reactive pro-inflammatory process may be seen in advanced age and its calcifications may not invariably be psammomatous. In Ogasawara et al.'s article, Van Dorpe et al. reported in 1999 that calcified fibrous pseudo-tumor may be the late sclerosed stage of an inflammatory myofibroblastic tumor, based on their observations in a 17-year-old patient.[4] In a study of 15 cases aged between 1 and 65 years old by Nascimento et al., calcified fibrous pseudo-tumor was defined as a different benign mesencyhmal neoplasia with a low recurrence risk and omitting the prefix "pseudo", the authors named the tumor as "calcified fibrous tumor".[5] According to the report of World Health Organization, the tumor was defined as a benign tumor which recurs in occasion and has several prognostic factors.[6]

According to reports from large series, distribution of CFT cases is equal among males and females.[1-7] It may be seen between 1 and 65 years of age with children and young adults being the most commonly affected.[1] Our patient was 28-year-old woman. Tumor size ranged from 0.2 to 20 cm in previous reports.[3,8,9] In our study, the smallest nodule was 0.2 cm in diameter and was not containing calcification whereas the largest nodule was 0.8 cm in diameter and was containing calcification.

CFT has a broad anatomic distribution and may be encountered in a variety of organ and tissues including oral cavity, subcutaneous tissue, muscle and deep soft tissue, extremities, thoracic wall, axillary area, pleura, mediastinum, neck, breast, myocardium, adrenal gland, peritoneum-omentum, gastrointestinal tract, lungs, mesentery organs, scrotum and peritesticular area.[1-8,10-12] Clinical signs and symptoms may be confused with those of other fibroblastic tumors of soft tissue origin and with any metastatic neoplasia, as well.

In our case, we incidentally found a total of 20 adjacent nodules in peritoneum, all of which were excised for histopathological investigation. In histopathological examination, we found a hypocellular stroma comprised of fibroblasts and hyalinized fibrosclerotic collagen fibers. The stroma was containing lymphoplasmocytic cell reaction, lymphoid aggregates and rare psammomatous and dystrophic calcifications which condensed mostly in perivascular area (Figure 1-4). There were no mitosis, necrosis or atypical cells.

CFT, as in our case, may be encountered as multiple lesions. Karl et al. reported two cases with peritoneal multifocal CFT.[6] Shibata et al. reported multiple CFT within the pleura.[13] Malignant transformation has not yet been reported.[1] In our case, recurrence did not occur within a follow-up of 14 months after the cesarean delivery.

Differential diagnosis of CFT includes inflammatory myofibroblastic tumor, desmoid fibromatosis, nodular fasciitis, solitary fibrous tumor, desmoplastic fibroblastoma, true fibroma of the oral mucosa, giant-cell fibroma, irritation fibroma, amyloidoma, soft tissue ectopic calcification, schwannoma, inflammatory fibroid polyp, leiomyom, gastrointestinal stromal tumor (GIST) and osteochondroma.[1,14]

Differentiating the CFT from inflammatory myofibroblastic tumor (IMT) may be challenging since fibroblasts, inflammatory cells and calcification may be found in both.[1,12] Histologically, calcification is always present in CFT. In IMT, inflammatory cells are polymorphic whereas they are lymphoplasmocytic in CFT. CFT has a uniform

feature whereas IMT has a multi-pattern structure.[1] According to immunohistochemistry analysis, ALK-1 expression is diffuse, Factor VIIIa and SMA are strong positive in IMT whereas ALK-1 expression may be rare and focal, Factor VIIIa may be focal positive and SMA may be very rare and focal positive in CFT.[1] In our case, vimentin was positive, ALK-1 and Factor VIIIa were negative and SMA was weak focal positive within the spindle cells of the lesion. Similar to us, Benizhak et al. reported that vimentin was positive and SMA and CD 34 were focal positive in spindle cells of the lesion.[11] Similarly, in study of Ogasawara et al., vimentin was positive.[4] Similar to ours, in studies of Agaimy et al. and Ogasawara et al., CD117, S100, SMA, Desmin, ALK1 and also h-caldesmon and PDGFRA were negative.[4,8] Weynand et al. reported a case with CFT in peritoneum showing 34 positivity.[14] Desmoid fibromatosis contains marked myofibroblastic elements arranged in long bundles.[1] In solitary fibrous tumor, spindle cells are seen arranged within a collagenous stroma without creating a pattern. CD34 is diffuse positive.[1]

Etiology and pathogenetic basis of CFT is yet to be established. In a case series of 11 patients, Mao et al. suggested that having been exposed to a previous inflammation or trauma may trigger development of CFT.[9] Similarly, Benizhak et al. suggested that intraabdominal soft tissue lesions may be precursor to the development of CFT.[11] Fukunaga et al. reported in a 20-year-old woman that the lesion had diploid DNA in flow cytometry.[15] Further cytogenetic studies are warranted to establish whether CFT is a true neoplasm or it is a reactive process and also to construct the most appropriate classification.

Excision of the tumor may be curative when the surgical boundaries are preserved. Recurrence was reported only in four cases in which it was attributed to incomplete excision.[1]

As a conclusion, non-metastating multiple peritoneal CFT is found rarely. It is benign neoplasm. CFT should be bear in mind when confronted with any nodular neoplasms clinically. Multifocal tumors may be confounded with benign and malignant neoplasms, especially with metastatic tumors. During histopathological examination, it may have macroscopic or microscopic similarities with other spindle cell tumors. Therefore, misdiagnosis should be avoided since its treatment is possible by total excision. The disease may recur rarely.

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