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Case report

Giant Perianal Angiomyofibroblastoma – A Case Report

Zdravko Perko[1], Merica Glavina Durdov[2], Nikica Družijanić[1], Damir Kraljević[1] and Joško Juričić[1]

[1] University Department of Surgery, University Hospital Split, Split, Croatia
[2] Department of Pathology, University Hospital Split, Split, Croatia

A B S T R A C T

A 45-year old female had a long history of slow growing perianal tumor at the right side of her anus. Encapsulated tumour was found intraoperatively and completely excised using the Harmonic Scalpel. Tumour was well-circumscribed and relatively firm; measuring 12x6x4 cm. Histologically it was composed of oval to spindle cells with minimal nuclear atypia, set in mucous matrix with numerous thin-walled blood vessels. Immunohistochemically, expression of smooth--muscle actin and desmin, as well as estrogen and progesterone receptor were found in the tumour cells. The diagnosis of angiomyofibroblastoma was established. This rare benign tumour typically involves vulvovaginal, pelvic and perinal region. It is important to separate this neoplasm from locally invasive aggressive angiomyxoma and low grade fibromyxoid sarcoma, which can arise in the the same localisation. The patient was discharged on the third postoperative day and no recurrence was noted in 18 months follow-up.

***Key words**: female genital neoplasm, rare benign tumors, harmonic scalpel*

Introduction

Angiomyofibroblastoma (AMFB) is recently described rare neoplasm that usually involves vulvar region, pelvis and perineum of women in the generative age[1]. The tumor appears as a slow growing mass that usually ranges 6 to 13 cm in its maximum span and have pushing peripheral margins[2]. This tumour has to be separated from infiltrative and prognostically less favorable lesions namely aggressive angiomyxoma and low grade fibromyxoid sarcoma[3,4].

Patient and Methods

Our patient is an 45-year old female with several years history of slow growing perianal tumour on the right side of her anus. The tumour was mobile, painless and covered with normal skin. Two-dimensional computerized tomography showed the tumor to be 5.8 by 12.5 cm in diameter and without infiltration of the adjacent organs (Figure 1). A barium enema study was normal and all laboratory tests were within normal ranges. A punch-biopsy specimen of the mass showed lipocytes only.

The patient was operated upon in the lithotomy position. Intraoperatively the tumor was found to be encapsulated, without infiltration of surrounding tissue. A complete excision was performed using high-powered ultrasonic dissection by Harmonic Scalpel (Ethicon Endo--Surgery, Cincinnati, USA, Figure 2 and 3). This dissection technique is widely used in open and laparoscopic operations and is generally perceived to carry a lower risk of collateral damage and to provide better quality pathology specimens than more traditional methods[5].

Results

A vacum drain was placed intraoperatively and removed day after. The patient was discharged on the third postoperative day, but re-admitted four days after due to perianal infection. Incision and drainage was performed. The postoperative course was uneventful and no tumor recurrence was noted after 18 months of follow-up.

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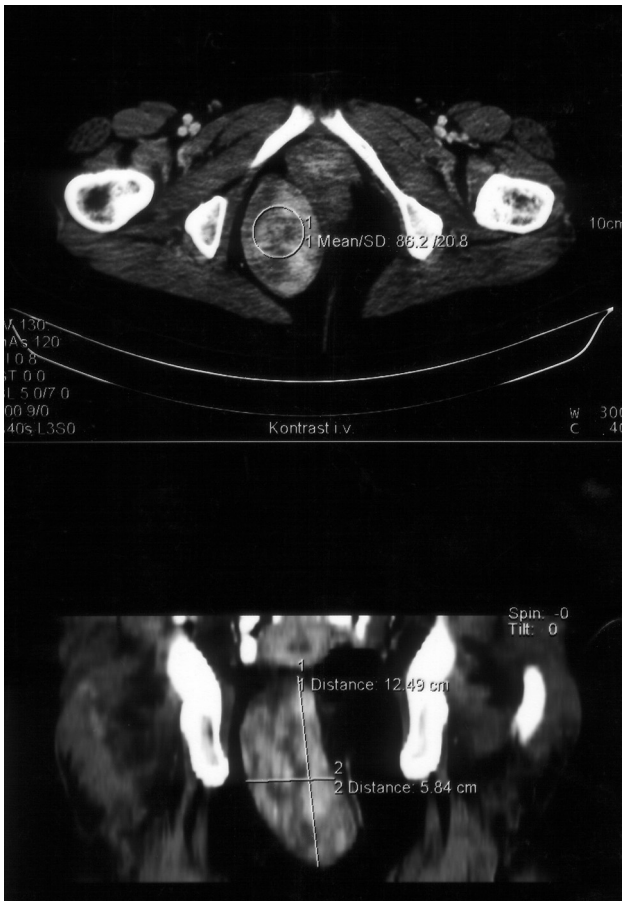

Fig. 1. CT scan shows a perianal tumor.

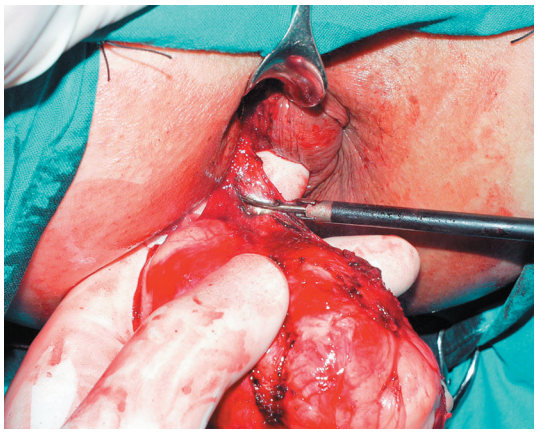

Fig. 2. Illustrating the final stages of tumor excision using the harmonic scalpel.

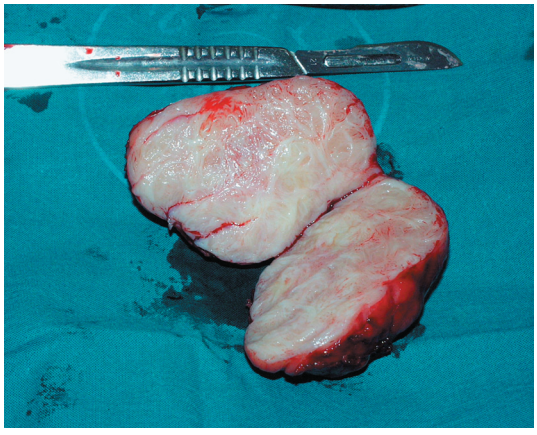

Fig. 3. The excised tumor cut in its long axis to illustrate dense, grey stroma.

Morphological findings

The excised tumor was a well-circumscribed, firm grey mass measuring 12 x 6 x 4 cm. Histologically it was comprised of plump oval, ovoid to spindle-shaped cells with minimal nuclear atypia set in copious edematous matrix in which there are numerous capillary-sized blood vessels (Figure 4). Occasional lymphoid aggregates and mastocytes were found within the tumor tissue. Immunohistochemical analysis confirmed expression of smooth-muscle actin and desmin in the cyto- plasm of tumour cells, as well as expression of estrogen and progesteron receptors in their nuclei (Figure 5, plate a–d).

Discussion

Mesenchymal neoplasms of modified genital skin and mucosa are uncommon. Most of these lesions are seen in females and comprise a family of vulvovaginal soft tissue tumors which includes the fibroepithelial stromal polyps, angiomyofibroblastoma, cellular angiofibroma, aggressive angiomyxoma, vaginocervical myofibroblastoma, vulvar leiomyomatosis and other smooth muscle tumors. Angiomyofibroblastoma is especially rare[1–4]. Most cases of AMFB occur in female genital area as a mass in the vulva, although recent reports described occurrence in vagina, perineum and inguinal areas[2,7,8].

Angiomyofibroblastoma is a tumor described by Fletcher et al in 1992[9]. The designation »angiomyofibroblastoma« is based on the two integral components of the tumor: blood vessels and stromal cells. The vascular component is prominent and intimately associated with stromal cells. Histogenetically it is believed that this tumor is derived from a perivascular stem cell with a capacity for adipose and myofibroblastic differentiation probably governed by hormonal, local microenvironment and growth factor/cytokine – related influences[6].

These tumors have ranged from 0.5 to 14 cm in their greatest dimension[1,3,9,10]. The lesions are mostly well-circumscribed. Margins are well delineated and non-infiltrative. Fibrous pseudocapsule of varying thickness may be present. Typically, tumour is composed of blood vessels in background of colagenous to oedematous stroma with alternating hyper and hypocellular regions.

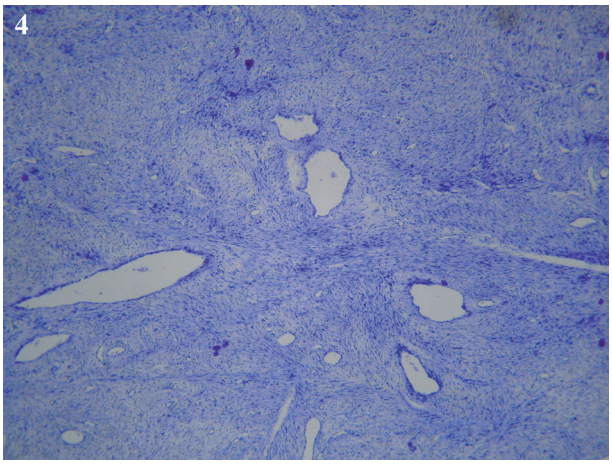

Fig. 4. The tumor tissue is composed of spindle cells and numerous thin-walled blood vessels, hematoxylin - eosin staining (x100).

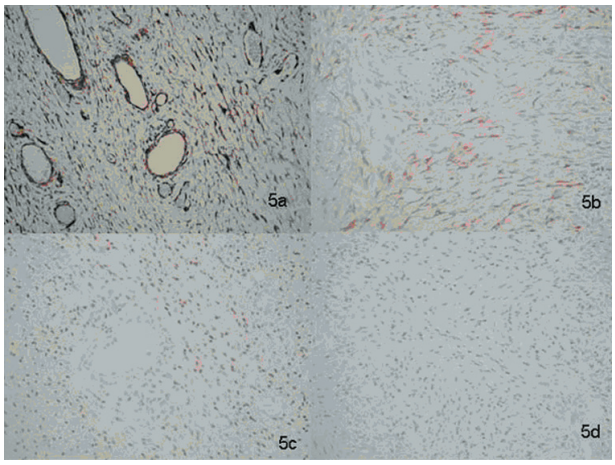

Fig. 5. Tumour cells express smooth-muscle actin (a) and desmin (b) in the cytoplasm and estrogen receptors (c) and progesteron receptors (d) in the nuclei of tumour cells. Immunohistochemical staining (x200) (antibodies and Envision/HRP visualisation system are products of DAKO Glostrup, Denmark).

The immunophenotype of angiomyofibroblastoma is not distinct but most cases are desmin positive and smooth muscle actin negative. However, some are negative for desmin or positive for smooth muscle actin. Diffuse positivity for ER and PR was present. This raises the possibility that angiomyofibroblastoma is a hormone responsive neoplasm.

The most important differential diagnosis of AMFB includes aggressive angiomyxoma, low grade fibromyoid sarcoma and cellular angiofibroma[2,11,12].

Aggressive angiomyxoma is locally invasive neoplasm first described by Steeper and Rosai[1,6]. Angiomyofibroblastoma can be distinguished from aggressive angiomyxoma by its circumscribed, non-infiltrative borders, much higher cellularity, more numerous blood vessels, minimal stromal mucin and rarity of erythrocyte extravasation.

Low grade fibromyxoid sarcoma is localised in the lower limp girdle of both sexes in the third to fifth decade. This locally invasive neoplasm has potential to metastasis. Tumour tissue is composed of whorls of bland looking fibroblasts which are vimentin positive, embedded in dense collagenous stroma..

Cellular angiofibroma shares similarities with AMFB in terms of age, sex and location. This lesion typically presents as a small well circumscribed mass. In contrast to AMFB can be locally aggressive. The cellular component is composed of spindle cells arranged in short intersecting fascicles admixed with thick walled hyalinised blood vessels.

In this case report we have described a perianal tumor without invasion of adjacent organs, diagnosed as angiomyofibroblastoma. Simple surgical excision was complicated by infection, which is not uncommon following surgery in this anatomical region. There was no tumor recurrence after 18 months follow-up.

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Z. Perko

Department of Surgery, University Hospital Split, Spinčićeva 1, 21000 Split, Croatia
e-mail: zperko@kbsplit.hr

VELIKI PERIANALNI ANGIOMIOFIBROBLASTOM – PRIKAZ SLUČAJA

S A Ž E T A K

Četrdeset i pet godišnja žena je bolovala od dugotrajnog sporastućeg perianalnog tumora na desnoj strani čmara. Tumor je bio inkapsuliran i odstranjen u cijelosti primjenom titrajućeg rezača (harmoničnog noža). Tumor je bio dobro ograničen i relativno čvrst, dimenzija 12x6x4 cm. Histološki je bio građen od ovalnih i vretenastih stanica s minimalnom atipijom jezgri, u mukoznom matriksu s brojnim krvnim žilama tankoga zida. Imunohistokemijski se u tumorskim stanicama našla ekspresija aktina i dezmina u glatkim mišićima, kao i estrogenski i progesteronski receptori. Postavljena je dijagnoza angiomiofibroblastoma. Ovi rijetki benigni tumori tipično invadiraju vulvovaginalnu, pelvičnu i perianalnu regiju. Važno je ove tumore razlikovati od lokalno invazivnih agresivnih angiomiksoma i fibromiksoidnih sarkoma niskoga stupnja, koji se mogu nalaziti u istoj regiji. Bolesnica je bila otpuštena trećeg poslijeoperacijskog dana i bez recidiva je 18 mjeseci nakon operacije.