

Giant Cell Angiofibroma of Pelvis

Abstract

Giant cell angiofibroma (GCA) is a rare, soft-tissue neoplasm that has a tendency to arise from the mesenchymal tissues of the head and neck, especially involving the orbital region. It is a benign neoplasm characterized by the presence of multinucleated stromal giant cells and angiectoid spaces. GCA belongs to the group of solitary fibrous tumor with histomorphological features intermediate between those of solitary fibrous tumor and giant cell fibroblastoma. CD34 immunoreactivity of tumor cells carries potential diagnostic value. Recurrence after the complete surgical excision is rare. We report the case of a 62-year-old male who presented with complaints of pain and heaviness in the lower abdomen. On computed tomography, a well-defined solid cystic mass was observed in the pelvis. Surgical resection was done, and the histopathological and immunohistochemical examination findings rendered the diagnosis of GCA. The patient had an uneventful postoperative period.

Keywords: *Angiofibroma, giant cell, neoplasm, pseudovascular*

Introduction

The WHO defines giant cell angiofibroma (GCA) as a “nonrecurrent, benign neoplasm containing multinucleated giant stromal cells and angiectoid spaces.”^[1] GCA is distinguished by its patternless proliferation of the spindle cell containing numerous pseudovascular spaces, floret-like multinucleated giant cells, and a highly vascularized stroma that may show secondary hyalinization or myxoid changes. GCA belongs to the spectrum of solitary fibrous tumors (SFTs),^[1] with its histological features intermediate between those of SFT and giant cell fibroblastoma (GCF) of soft tissue.^[2,3] The incidence of extraorbital GCA is very rare and can occur in the posterior mediastinum, back, axillary region, retroperitoneal spaces, and inguinal regions.^[4] These tumors are usually well circumscribed, smaller in size, and variably encapsulated; however, the extraorbital tumors tend to be larger in size and may simulate a malignant or aggressive process. Complete surgical resection of the tumor is the treatment of choice in the management of GCA. We describe here a case of GCA arising within the pelvis of a 62-year-old male, to

emphasize the diversity in the occurrence of this rare neoplasm and also to highlight its relevant clinical, histomorphological, immunohistochemical, and molecular characteristics that would help in rendering the correct diagnosis and differentiating this tumor from its close mimickers.

Case Report

A 62-year-old male presented to the surgery outpatient department with complaints of pain and heaviness in the lower abdomen for 6 months. On local examination, a lump was felt in the right iliac fossa measuring 6 cm × 5 cm. His general examinations were unremarkable and the routine laboratory tests were also normal. Ultrasonography (USG) and contrast-enhanced computed tomography (CT) revealed a well-defined solid cystic mass in the pelvic region, involving the recto-vesicle space and extending toward the right iliac fossa. No retroperitoneal lymph node or free fluids was observed. Rest of all the organs were normal except for mild prostatomegaly. On surgical exploration, a soft-tissue mass was found in the recto-vesicle space that was adherent to the bladder on its posterior surface and was extending up to the right inguinal region. The mass was dissected out from the inguinal region with relative ease, however, the portion that

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was densely adherent to the posterior bladder wall posed much difficulty, but was carefully freed and the tumor mass was excised in its entirety with no evidence of residual disease. The specimen was then sent for histopathological examination. On gross examination, the tumor mass appeared tan gray with ill-defined margins, measuring 11 cm × 10 cm × 7 cm. It was soft in consistency. On cut-section, grayish-white vague nodularities with areas of hemorrhage and cystic changes were seen.

The microscopic examination showed the neoplastic proliferation of variable cellularity with pushing margins. The tumor was composed of oval-to-spindle-shaped cells arranged in a patternless manner, surrounding the pseudovascular spaces in a collagenized stroma [Figure 1], intermingled with numerous scattered multinucleated giant cells with nuclei arranged in the center of the cells [Figure 2], and resembling floret giant cells. These cells were also seen lining those pseudovascular spaces of varying caliber with perivascular hyalinization and infrequent mitotic figures. Areas with cystic changes were also noticed. Immunohistochemical staining for Ki-67, CD31, CD34, CD68, CD 99, CD117, BCL 2, smooth muscle actin (SMA), desmin, S100, and pancytokeratin were performed with their adequate controls. The tumor cells and the giant cells were diffusely and strongly positive for CD34 [Figure 3], BCL 2, and focal positivity for CD99; however, CD68, CD31, CD 117, SMA, desmin, S100, and pan-CK were completely negative. Ki 67 was very low ($\leq 1\%$). The combination of histomorphological features and immunohistochemical findings rendered the final diagnosis of GCA. On follow-up, the patient is in good health without any evidence of local recurrence.

Discussion

GCA was considered a giant cell rich form of hemangiopericytomas (HPC)-SFTs by Dei Tos *et al.*^[5] GCA is an entity that is considered to be a histological variant of HPC-SFT spectrum. Although GCA displays all the morphological features of a classic SFT but is still distinguished by its pseudovascular spaces rimmed by multinucleated stromal giant cells. HPC, SFT, and GCF are soft-tissue neoplasm of fibroblastic origin that comes under the differential diagnosis of GCA and show overlapping histomorphologic and immunophenotypic findings.^[6] GCA shows wide age distribution from the 2nd to 8th decade of life with a mean age of 45 years,^[7] also, this tumor shows a diverse anatomic distribution, commonly occurring in orbital location, including the eyelids, nasolacrimal duct, and the lacrimal sac. Handful of cases involving the head-and-neck region outside the orbit, including scalp, retroauricular region, parotid gland, cheek, submandibular region, buccal mucosa, and other areas as posterior mediastinum, back, axillary and inguinal regions, retroperitoneum, and vulva have also been reported.^[3,8,9] GCA clinically occurs as a

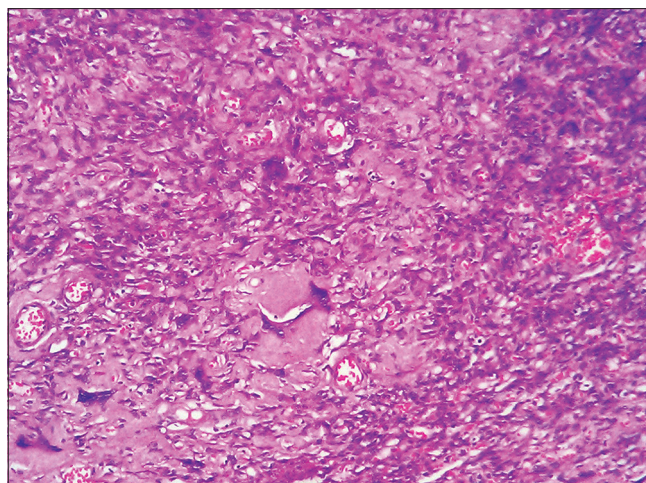


Figure 1: Section showing, tumor composed of oval-to-spindle-shaped cells arranged in a patternless manner surrounding the pseudovascular spaces in a collagenous stroma (H and E, ×100)

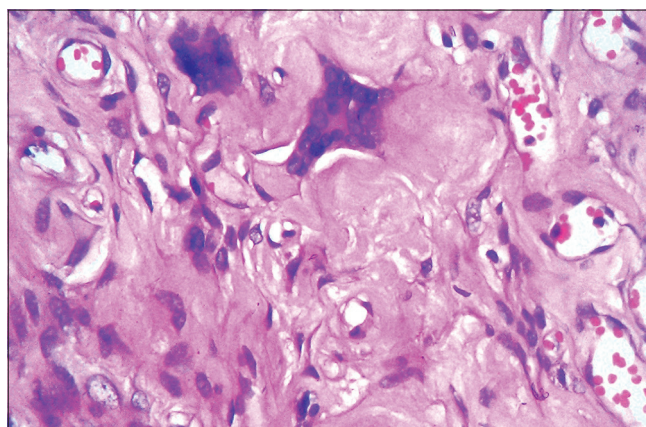


Figure 2: Section shows numerous scattered multinucleated giant cells with nuclei arranged in the center of the cells resembling floret giant cells, lining the pseudovascular spaces (H and E, ×400)

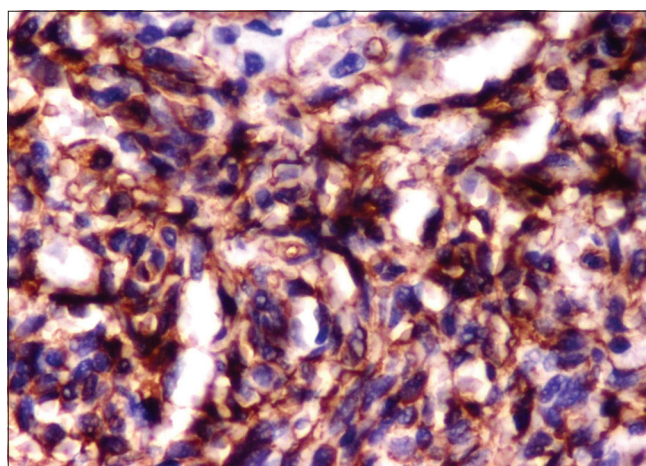


Figure 3: Strong and diffuse positivity for CD34 in the tumor cells and stromal giant cells (immunohistochemistry, ×400)

slow-growing mass that can be painful. Imaging techniques such as USG, CT scan, and magnetic resonance imaging

are helpful not only in locating the tumor but also play a crucial role in assessing its extent of involvement and tumor resectability. Macroscopically, the GCA is small, well-circumscribed lesion that may be variably encapsulated. The average size of this neoplasm is approximately 3 cm, however, soft-tissue lesions, especially in mediastinum and retroperitoneal locations, tend to be larger and can reach up to 10 cm.^[10] Features such as hemorrhage or cystic changes can also be seen. Microscopically, these tumors are well circumscribed, comprising of benign appearing round to ovoid cells with pale, indistinct cytoplasm set in a variably collagenized stroma with irregularly shaped angiectoid spaces lined by mononuclear and multinucleate tumor cells; multinucleated floret giant cells along with many small blood vessels with hyalinized walls. Mononuclear tumor cells and multinucleated stromal giant cells are characteristically positive for immunomarkers such as Vimentin CD 34 and CD99 and less frequently, for BCL2^[10] but consistently negative for muscle-specific actin, desmin, CD31, CD117, and S100.

Surgical resection is the mainstay of treatment. Almost all GCA s exhibit benign behavior. Recurrences after the complete excision is an exception.^[11]

Besides being a rare entity, GCA should be kept as a differential diagnosis when the histopathological examination reveals a well-defined fibroblastic patternless, CD34 (+) tumor mass. GCA is a highly vascular, benign, nonrecurring soft-tissue tumor that has predilection for the orbit and head-and-neck region, still lesions at extraorbital sites have been reported in diverse locations. Histopathologic examination and immunohistochemical analysis help in reaching the conclusive diagnosis of this neoplasm.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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