A Dedifferentiated Solitary Fibrous Tumor in Mesentry with Osteosarcomatous Differentiation

Aparna Gangoli[1], Triparna Ghosh[2], Komal D Chippalkatti[3], Sanjeev Kulgod[4]

ABSTRACT

Introduction: Solitary fibrous tumor, a mesenchymal neoplasm, was initially observed in pleura. Though they were later found in many extra pleural sites. It rarely occurs in mesentery. The biology of solitary fibrous tumor is unpredictable, it ranges from benign to overtly malignant. Dedifferentiation, a phenomenon which is morphologically characterized by abrupt transition between the well differentiated component and high-grade areas of tumor, is well described in soft tissue and bone tumors. However, dedifferentiation within solitary fibrous tumor is a rare phenomenon.

Case report: We report a case of dedifferentiated solitary fibrous tumor arising in mesentery. Computed tomography revealed a large well-lobulated heterogeneously enhanced solid necrotic mass lesion involving the pelvic abdomen. Macroscopically, the resected tumor was 24 x 20 x 10 cm. microscopic examination revealed a malignant spindle cell tumor juxtaposed with osteosarcomatous component. The spindle cell component showed diffuse expression of SMA, CD34 & STAT6. The osteosarcomatous component showed diffuse expression of SATB2.

Conclusion: This is a rare case of mesenteric dedifferentiated solitary fibrous tumor with osteosarcomatous differentiation.

Keywords: Dedifferentiated Solitary Fibrous Tumor, Mesentry

INTRODUCTION

Solitary fibrous tumors, though initially observed in the pleura, can occur at any site of the body.[1] But there are very few reports on retroperitoneal solitary fibrous tumors.[2]

The biology of SFT is very unpredictable. Although most of the SFTs are benign in nature, approximately 10% of cases show aggressive behavior in the form of local recurrence or metastasis.[3] The distinction between benign and malignant SFT (MFST) is difficult. The WHO classification of soft tissue tumors (2013) defines MFST as a tumor with hypercellularity, at least focal moderate to marked cellular atypia, tumor necrosis, 4 mitoses/10 high-power fields (HPFs) and infiltrative margins.[4]

Dedifferentiation within SFTs is a rare phenomenon.[5] It is different from malignant SFTs because in dedifferentiated SFTs, there is an abrupt transition between conventional SFT and dedifferentiated component. Mostly dedifferentiated component is a high-grade sarcomatous component such as rhabdomyosarcomatous or osteosarcomatous component.[6]

Here, we report a rare case of dedifferentiated SFT, with a heterologous osteosarcomatous component arising in mesentery.

CASE REPORT

A 64-year-old lady presented with pain in the right abdomen with decreased appetite. Computed tomography of the abdomen and pelvis revealed an approximately 12 x 12 cm large lobulated heterogeneously enhanced solid necrotic mass lesion involving the pelvic abdomen.

After multidisciplinary team discussion, surgical exploration was performed. Macroscopically, the resected mass showed a multinodular, grey white, fish-flesh appearance with necrotic foci measuring 14 x 14 x 10 cm, surrounded by a fibrous capsule, separating it from surrounding structures like ileum, cecum and ascending colon. Histological sections show predominantly bone forming neoplasm with extensive immature osteoid and osteoblastic component [Fig1]. High grade areas show pleomorphic cells with variable mitosis including abnormal ones. [Fig 2,3]. Also seen is adjacent spindle cell neoplasm comprising thin-walled branching vessels [Fig 4].

The spindle cell neoplasm showed diffuse expression of SMA, CD34 & STAT6 [Fig 5]. Negative to S-100, SOX10, CD117, SATB2, Calretinin, D2-40, Caldesmon and beta catenin. The ostosarcomatous component showed diffuse expression of SATB2 [Fig 6] & vimentin. Negative to CK, DOG 1 and CD117. The final diagnosis was dedifferentiated SFT comprised of conventional SFT and undifferentiated high-grade sarcoma with an osteosarcomatous component.

DISCUSSION

Klemperer and Rabin originally described SFT in the pleura.[7] But later it has been seen that SFT can arise from many extrathoracic sites including retroperitoneum, head and neck, peritoneum, abdomen, meninges, orbit, upper respiratory tract, salivary glands, thyroid, liver, adrenal gland, kidney,

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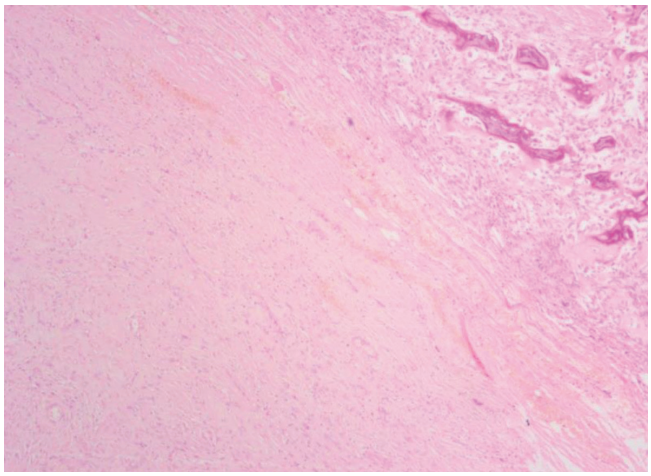

Figure-1: 4x-scanner view

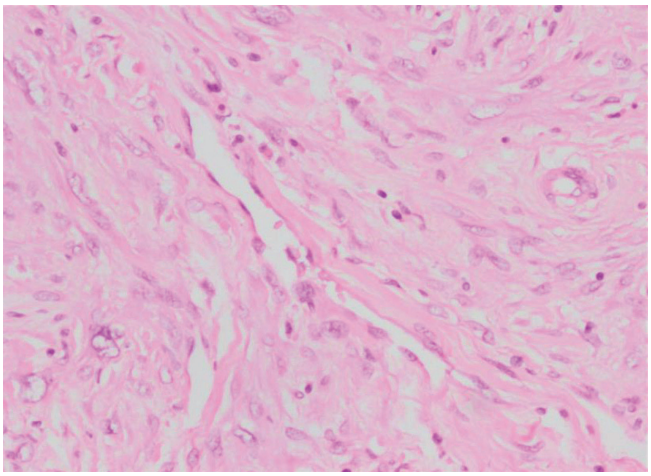

Figure-4: showing branching blood vessels

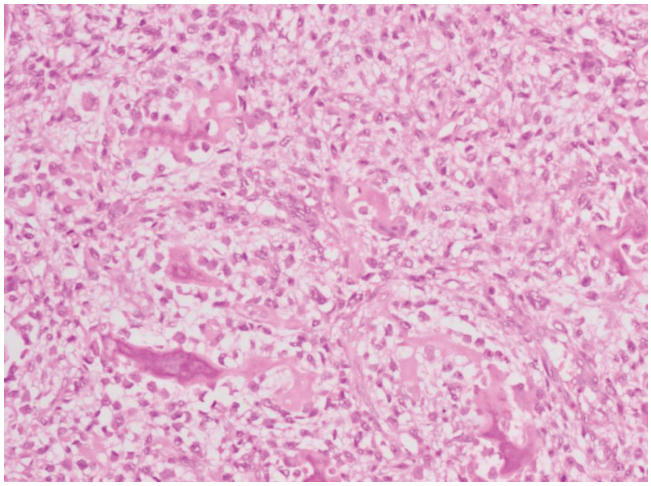

Figure-2: low power view

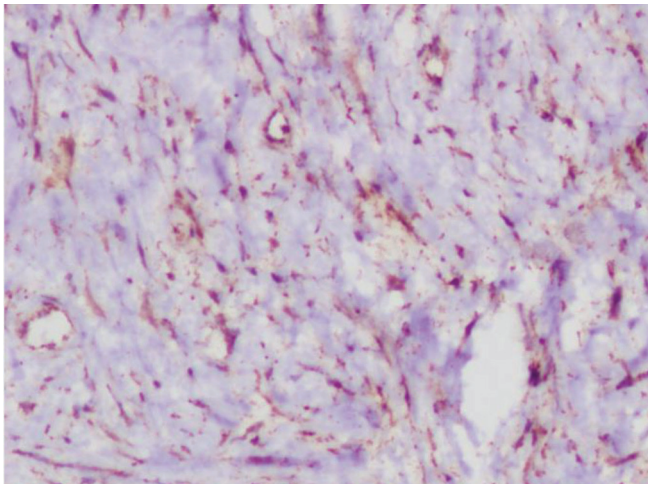

Figure-5: STAT6 nuclear positivity in SFT component

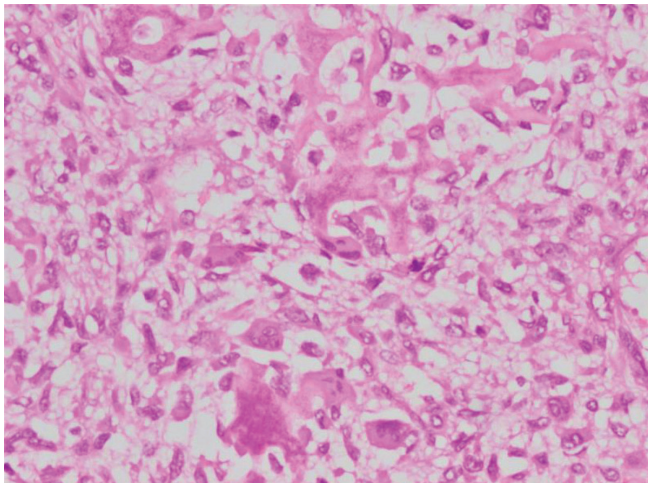

Figure-3: high power view

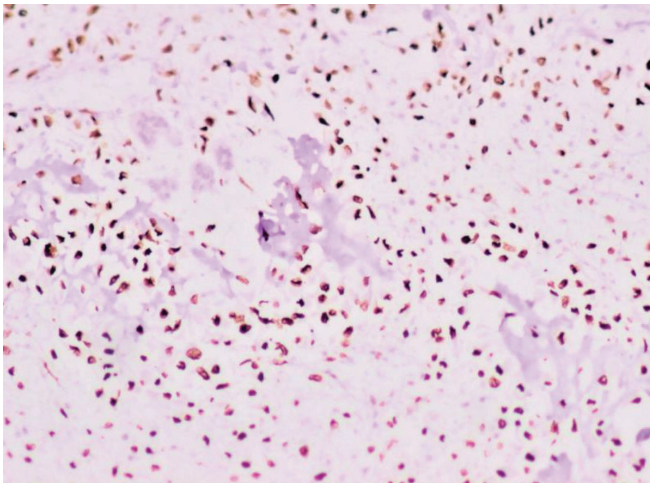

Figure-6: SATB2 positive in osteosarcomatous component

spermatic cord, urinary bladder, prostate, uterine cervix, spinal cord and periosteum.[7,8] Here in this report, we present a rare case of SFT arising from mesentry. To the best of our knowledge only 15 cases reported in the literature.[9]

Histologically, SFT is composed of spindle to ovoid cells with pale eosinophilic cytoplasm within a variable collagenous stroma and staghorn -shaped blood vessels.[10] Many subgroups of SFT have been found like fibrous form, lipomatous (fat- forming), giant cell rich and dedifferentiated.[11] The biologic behaviour of SFT is difficult to predict. Though malignant SFT shows malignant features throughout the tumor, dedifferentiated SFT shows an abrupt transition between the area of classical SFT and dedifferentiated component.[12] The dedifferentiated component is mainly high grade sarcoma with or without heterologus element such as rhabdomyosarcoma or osteosarcoma.

Morphologically, this was a typical case of dedifferentiated SFT, where we found both conventional SFT area and high grade osteosarcomatous area. These both components also showed distinctive immunohistochemical differences. In this case, diffuse immunohistochemical expression of SMA , CD34 and strong nuclear STAT6 expression was seen only in conventional SFT component and was absent in osteosarcomatous component. Molecular analyses have discovered that almost all SFTs harbor an NAB2-STAT6 fusion gene . This gene is considered specific for this tumor type and according to the recent studies, STAT6 immunohistochemistry is considered as a reliable surrogate for detection of the fusion gene.[13] Thus the strong nuclear immunohistochemical expression of STAT6 in spindle cell compenent confirms the diagnosis of conventinonal SFT component. This spindle cell component was negative to S100, SOX10, CD117, DOG1, Ccalretinin, D2-40, caldesmon and betacatenin and therefore ruling out the possibility of neurofibroma, GIST, mesothelioma, smooth muscle neoplasm and fibromatosis. On the other hand, the dedifferentiated element showed sarcomatous tumor cells that produces estensive immature osteoid. These sarcomatous tumor cells showed diffuse expression of SATB2 and vimentin, confirming the diagnosis of osteosarcoma.

The differential diagnosis of MSFT includes benign and malignant lesions, such as malignant peripheral nerve sheath tumor (MPNST), leiomyosarcoma, GIST, mesothelioma and fibromatosis.

MPNST has features of heterogeneous spindle cells with variable growth pattern and arrangement and has bizarre giant cells, high mitotic activities, and distinguishing patterns of necrosis.[14] But in our case, immunonegativity for S100 and SOX10 was helpful to exclude the diagnosis.

Leiomyosarcoma mainly is composed of spindle shaped cells with plump, blunt ended nuclei and moderate to abundant, pale to brightly eosinophilic fibrillary cytoplasm along with necrosis and brisk mitosis including atypical ones. Dedifferentiated leiomyosarcoma shows features of low grade leiomyosarcoma with a discrete heterologous component.[15] In our case, immunonegativity for SMA and Caldesmon was helpful to exclude the diagnosis.

GIST has three morphologic types- 1. Spindle 2. Epithelioid and 3. Mixed. Spindle type has many subtypes like sclerosing, palisaded, vacuolated, diffuse hypercellular, sarcomatoid features with significant nuclear atypia and mitotic activity.[16] But in our case, immunonegativity for CD117 and DOG1 excluded the diagnosis of GIST.

Sarcomatoid variant of mesothelioma has features of spindle cells growing in a storiform pattern in a collagenized background with stromal invasion and bland necrosis.[17] But in our case, tumor cells were immunonegative for calretinin and D2 40, thus excluding the diagnosis of mesothelioma.

Fibromatosis is mainly composed of uniform spindle cells with pale cytoplasm in a collagenized stroma along with minimal nuclear atypia and variable mitotic count. It has also different patterns like hypocellular, hypercellular, keilodal, staghorn vessel and myxoid pattern.[18] However in our case, immunonegativity for beta-catenin was helpful to exclude the diagnosis of fibromatosis.

The cases which show gradual transition from classic SFT/MFST to a high grade sarcomatous component or the tumor having high grade sarcomatous component without any trace of SFT/MSFT at a site of local relapse or metastasis, were described as “evoluted SFT” by Collini et al.[19] They used the term Dedifferentiated SFT only to describe cases with evidence of an abrupt transition from the classic SFT/MSFT to the high-grade sarcomatous component. According to this criteria, our case corresponds to dedifferentiated SFT as the tumor was composed of two separate components, one component showing classic SFT component and other component showing high grade osteosarcomatous component.

CONCLUSION

Primary SFT originating in retroperitoneum is extremely rare. In summary, here we present such a rare case of dedifferentiated SFT in retroperitoneum. The immunohistrochemistry for STAT6 and SATB2 was very helpful to establish the diagnosis. Therefore, though it is very rare, the differntial diagnosis of SFT for the lesions located in retroperitoneum, should be kept in mind. Malignant histopathological features and/or dedifferentiated areas in tumors with hemangiopericytomatous pattern should not be omitted by careful histopathological examination. An updated immunohistochemical panel including more specific antibodies for these cases should be applied. In order to increase our knowledge about the clinicopathological features and biological nature of these tumors, there is a need for more cases of extrathoracic/extrapleural SFT.

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