

Pelvic mass with pulmonary metastasis: A rare differential with newer trends in their management

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A 53-year-old gentleman presented with a painless lump in hypogastrium for 6 months. He also complained of increased frequency of urination, dysuria and constipation. The patient reported to have cough with expectoration containing occasional blood stained sputum. On examination, a 6 × 5 cm well defined, smooth, non-tender, and hard lump with palpable lower margin was found occupying the hypogastrium (Fig. 1). Per-rectal examination revealed a mass arising from the left pelvic wall and compromising the rectal lumen but with retained mucosal integrity.

Ultrasonography of abdomen revealed 9 × 8 cm homogenous, hypochoic, solid mass with significant vascularity arising from the left lateral pelvic wall with left side moderate hydro-ureteronephrosis. Contrast enhanced computer tomography (CECT) of the abdomen demonstrated a homogenous, well demarcated, hypo attenuated mass of 12.6 × 10.6 cm originating from the left lateral pelvic wall with significant contrast enhancement (Fig. 2). The tumor was compressing on the rectum and bladder, with maintained fat planes. Radiograph of chest

showed multiple cannon ball lesions (Fig. 3). CT of head and bone scan was normal.

Tumor resection was not contemplated due to the presence of pulmonary lesions. Multiple chunks retrieved through tru cut biopsy of the lump were found to be unsuitable by the pathologist to formulate a definitive diagnosis. Incision biopsy of tumor was carried out as the next step to aid in tissue diagnosis. Histology suggested spindle cell tumor with mild nuclear atypical, but no mitotic figures. Immunohistochemistry (IHC) was positive for CD-34, CD-99, and bcl-2, Vimentin and KI 67<1%; however, markers for Actin, Desman and S-100 were negative (Fig. 4). High resolution CT thorax revealed multiple pulmonary nodules, and CT guided biopsy confirmed spindle cell nature of the pulmonary nodule.

QUESTIONS

- 1: What is the diagnosis?
- 2: What should be the plan of management in this patient?

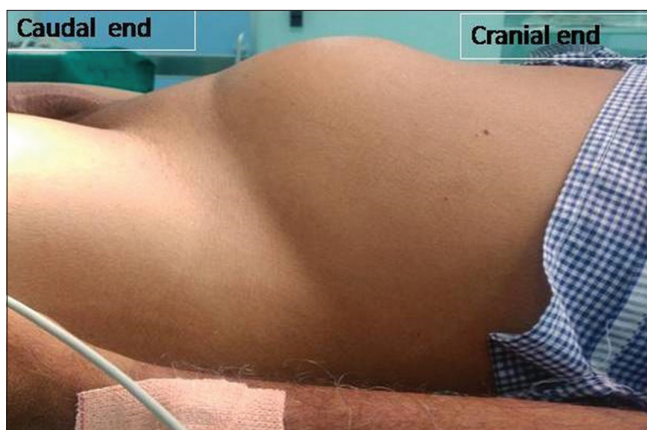


Figure 1: The patient presented with hypogastric lump and compressive symptoms on bladder and rectum

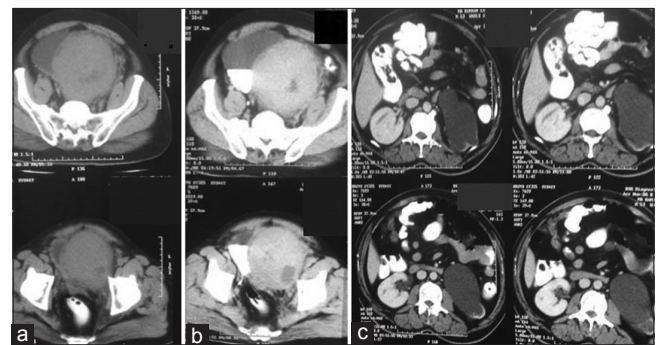


Figure 2: (a) Contrast enhanced computer tomography pelvis: Axial view showing well defined encapsulated heterogeneous pelvic mass 12.6 × 10.6 cm from the left lateral pelvic wall, (b) the mass has few necrotic areas within it and significant uniform post intravenous contrast enhancement. The mass was displacing rectal/bladder wall with maintained fat planes, (c) pelvic mass leads to obstruction of the left kidney drainage (moderate left side hydronephrosis)

ANSWER 1

Histopathology was suggestive of solitary fibrous tumor (SFT). This rare soft tissue sarcoma of mesenchyme cell origin closely resembles hemangiopericytoma (an extremely vascular tumor). In 2006, WHO identifies SFT as a distinctive entity from hemangiopericytoma. SFT can arise in mesothelium tissues (pleura, peritoneum, and pericardium), soft tissues and visceral organs (lung, meninges, thigh, thyroid, breast, nose, pharynx etc.) [1]. The most common site of SFT is pleura. Calcification (26%), hypoglycemia, osteodystrophy, arthralgia, and clubbing

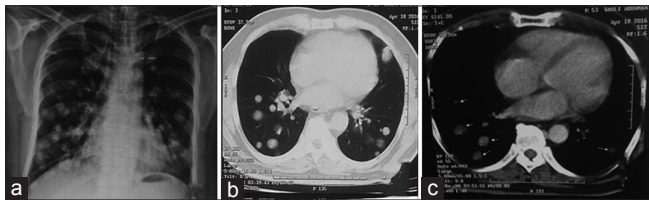


Figure 3: (a) Chest radiograph showing multiple nodules of various sizes at basal and sub-pleural location, (b and c) lung and mediastina window of high-resolution computed tomography thorax (axial view) showing multiple sharply defined rounded pulmonary nodule of various size

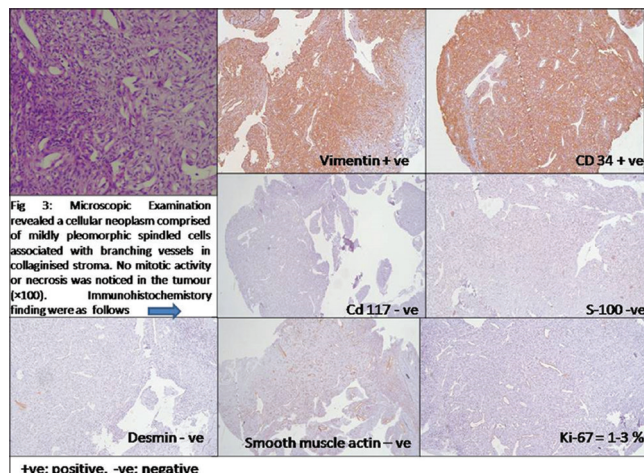


Figure 4: Histopathology and immuno-histochemistry of pelvic mass



Figure 5: Contrast enhanced computer tomography pelvis axial view, one month post embolization tumor have increased area of necrosis, however no significant reduction in size of tumor

are the common manifestations in pleural SFT. It usually has a benign clinical course, but prone to recur either locally or at distant sites despite clear margin of resection. In the absence of specific imaging characteristics that can aid in the diagnosis histopathological examination complemented by IHC study is considered the cornerstone for identification of SFT. Expression of CD-34 positive markers with negative S-100 markers is highly suggestive of a diagnosis of SFT. The malignant lesions are characterized by large tumor size, sessile growths, high mitotic index (>4 mitotic figures/10 HPF), nuclear pleomorphic, high cellularity and the presence of necrosis and/or hemorrhage. The relationship between histological features and clinical behavior of SFT is not so clear and the tumor has an unpredictable course [2].

Answer 2

Complete surgical excision is the primary modality of treatment. The quality of initial excision in achieving tumor free margins is considered the most important prognostic factor. Adequate surgical resection has been associated with a 10 years survival rate of 54-89% in patients with localized SFT. Radiotherapy had been attempted in a number of patients for local tumor control when resection margins were found to be involved on histopathological examination. In patients having unresectable and metastatic SFT anticancer drugs (anthracyclines with or without ifosfamide or trabectedin, and gemcitabine with docetaxel), and anti-angiogenic drugs (bevacizumab combined with temozolomide) are tried with a partial/no response. Few authors have achieved long term tumor control with sunitinib, sorafenib, or imatinib [3]. The goal of treatment in our patient was to improve on the local compressive effects of the tumor as well as to prolong the progression free survival. Two sittings of tumor embolization combined with oral sorafenib (200 mg/day) managed to arrest the tumor progression (assessed by high-resolution computed tomography thorax and CECT scan of pelvis) over a period of 1 year (Fig. 5). Subsequently the patient opted out of the treatment due to financial constraints and succumbed to fulminant pneumonia and respiratory failure within 6 months of discontinuation of palliative therapy.

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