

Solitary Fibrous Tumor of Sigmoid Colon as a Differential Diagnosis of Adnexal Mass

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Abstract

Solitary fibrous tumor is a rare neoplasm commonly arising from thoracic pleura. However, we came across an unusual site of solitary fibrous tumor in a 32 year old female with complains of lower abdominal pain of one month duration. On investigation, she was found to have a solitary fibrous tumor in the left iliac fossa. Solitary fibrous tumor should always be considered as a differential diagnosis for patient presenting with abdominal mass.

Keywords: Adnexal mass; Fibrous tumor; Urinary obstruction; Case report

Introduction

Solitary fibrous tumor is a rare spindle cell neoplasm that most commonly arises from the thoracic pleura [1]; however, solitary fibrous tumor is increasingly being reported to arise from any site in the soft tissues [2]. Most extrapleural solitary fibrous tumors occur in adults between 20 and 70 years of age and tend to occur equally in men and women². These are traditionally benign, slow growing tumors that often remain asymptomatic until they compress other structures producing pain, urinary obstruction or retention, bowel obstruction or constipation, a palpable mass, and neurologic or vascular symptoms [2,3].

Radiologically Computerised tomography is the single best modality to diagnose solitary fibrous tumor which is seen as soft tissue attenuation on unenhanced scans and shows relatively homogenous intense background enhancement on contrast-enhanced scans (from rich vascularization). Non-enhancing areas may be present, corresponding to necrosis, myxoid degeneration, or haemorrhage within the tumour. A pedicular attachment may also be seen. These rare mesenchymal derived spindle neoplasms are currently classified as “typical” or “malignant.” Surgical excision is the treatment of choice with a 5-year survival close to 100% if completely resected. However, malignant histology is the best predictor of poor outcome [3-5].

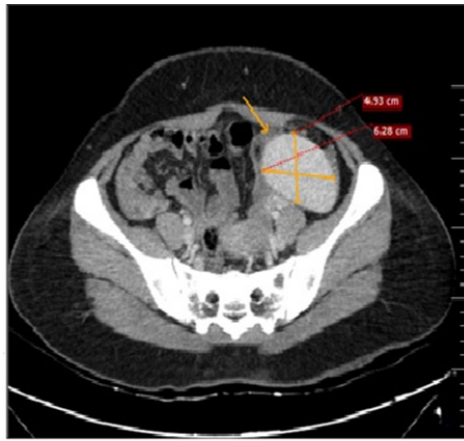
Case Report

Clinical summary : A 32 year old female presented with history of left lower abdominal pain since 1 month , which was dull aching , radiating to left lower back and aggravates on micturition. On examination patient is stable , with soft , non-tender abdomen The clinical differential diagnosis at this point included ovarian mass or cyst with possible rupture, urinary tract infection and inflammatory bowel disease.

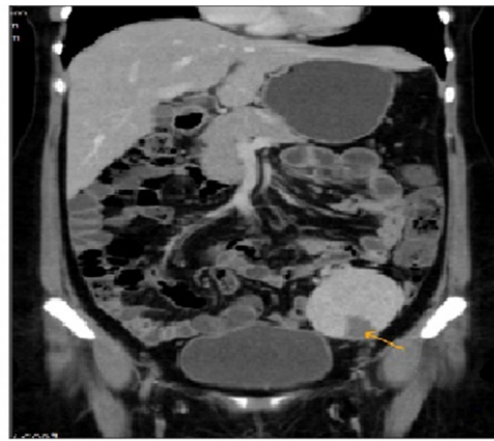
An abdominal computed tomography (CT) scan demonstrated well-defined homogenous lesion in the left iliac fossa showing intense homogenous early arterial & portovenous phase enhancement (**Figure 1**) except for a small non-enhancing peripheral area in the inferior aspect (**Figure 2**). Lesion measures 6.8 x 4.3 x 6.5cm with multiple vessels (arterial feeder- inferior mesenteric artery(IMA) and venous drainage into IMV and left renal vein) along the posterosuperior aspect.

Discussion

Solitary Fibrous Tumor (SFT) is a rare mesenchymal tumor (spindle cell neoplasm) originating in body cavity sites, most commonly pleura, peritoneum and meninges or at any site in the soft tissue including seminal vesicle. When arising from the pleura, 13-23% are classified as malignant in contrast to most extrapleural tumors which, with the exception of those of



[Fig.1]



[Fig.2]

Figure 1: A well-defined homogenous lesion in the left iliac fossa showing intense homogenous early arterial & portovenous phase enhancement (indicated with an arrow).
Figure 2: A small non-enhancing peripheral area in the inferior aspect of the homogenous lesion in the left iliac fossa. (indicated with an arrow).

mediastinal origin, have a benign outcome [6]. Most SFTs will present as asymptomatic masses discovered incidentally on chest imaging. Those that do present with symptoms present with nonspecific chest complaints such as chest pain, dyspnea or cough. Intra-abdominal SFT are generally asymptomatic until they reach large enough size to cause mass effect on other organs and localized symptoms like pain, urinary obstruction and constipation. Tumor size at presentation is variable and is typically within a range from 0.8-30 cm (median-5 cm) [7]. Although most SFTs are characterized by a non-aggressive clinical course, malignant transformation and a large size have been associated with a poor outcome, thus making long-term follow-up in all cases strongly advisable. Lesions located in the mediastinum, abdomen, pelvis or retroperitoneum tend to behave more aggressively than those in the limbs. Metastases are most often reported in the lungs, bone and liver. On rare occasions SFT can present with paraneoplastic syndromes, the most commonly described being non-islet cell hypoglycaemia. This is due to tumor production of high molecular weight insulin-like growth factor (IGF), specifically IGF-II [8].

The morphological criterion of all these lesions is represented by a well-circumscribed, tan-coloured, rubbery mass, which is tethered by a pedicle and partially encapsulated. It is described as a 'patternless' proliferation of bland-looking spindly to oval epithelioid cells that form short fascicles and/or clusters, admixed with thick or thin collagen bands, and a prominent branching vasculature. Mature adipocytes and giant multinucleated stromal cells may be present [9]. The tumour is difficult to detect because it has no typical radiologic features. Contrast-enhanced CT scan is usually performed: it reveals a well-defined capsule surrounding a nearly homogeneous mass with progressive enhancement from the arterial to the venous phase and occasionally multiple small non-enhancing portions.

In our case study, the general features present were left lower dull aching abdominal pain since 1 month which aggravated on micturition and an abdominal mass. Hence, an adnexal mass was considered. An adnexal mass is a growth that occurs in or near the uterus, ovaries, fallopian tubes, and the connecting tissues. They can be solid or filled with fluid and can be benign or malignant. The differential diagnosis at this point included ovarian mass or cyst with possible rupture, urinary tract infection and inflammatory bowel disease. CT showed a well

defined homogenous lesion measuring 6.8 x 4.3 x 6.5cm with multiple vessels.

A pre-treatment biopsy to diagnose and grade the mass is ideal. Because of the unpredictable behaviour of SFTs, surgery is considered the treatment of choice. The use of chemotherapy is reserved to metastatic or symptomatic non-resectable SFTs, but there are no standard chemotherapeutic indications or regimens. However, the most effective drugs seem to be anthracyclines and ifosfamide, followed by gemcitabine and dacarbazine, which are commonly used in soft tissue sarcomas. Radiation therapy is of some benefit, when used in combination with chemotherapy [10].

Conclusion

Even though SFT is a rare phenomenon, it should be considered as a differential when dealing with an adnexal mass. Given the broad differential and genetic overlap in many of these tumors, work-up and management should be determined in a multidisciplinary setting, where personalized aspects of each individual case can be discussed and taken into account. Early diagnosis and treatment can help in apt treatment and prevention of complications.

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