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Research Article

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Sarcoma in the pelvis of a young adult male: A rare case report with review of literature

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Abstract Synovial sarcoma is uncommonly documented in the pelvis. Rarely, such cases have dealt with molecular analysis. A 32-year-old boy presented with pain and swelling in his left lower limb of two three duration. He developed acute urinary retention four days prior to his hospital admission, wherein radiological examination unraveled a large soft tissue mass displacing his pelvic muscles, along with a lesion involving his left public bone. Biopsy showed a cellular spindle cell sarcoma, exhibiting hemangiopericytoma-like vascular patter with focal necrosis. Diagnosis of a differentiated synovial sarcoma was offered and confirmed.

Keywords pelvic synovial sarcoma , pelvic tumors, soft tissue sarcomas

Introduction

Synovial sarcoma (SS) is defined as a mesenchymal spindle cell tumor, displaying variable epithelial differentiation and is characterized by a specific chromosomal translocation t (X ;18) (p11 ; q 11) [1-2].

It is uncommon, account for 5-10% of soft tissue sarcomas and is unrelated to the synovium. Traditionally, extremities form the commonest sites of its occurrence in 80-90% cases. However, with the advent of ancillary techniques, it has been identified at unusual locations like head and neck region, lung, prostate [3]. Synovial sarcoma is rare within the abdomen. In this location, it can be confused with other biphasic tumours and with other spindle and round cell sarcomas [1]. Few cases of SS have been documented in the pelvis, especially involving the bone. Still rare is its objective identification with molecular results in this location. Herein, we report a case, differentiated synovial sarcoma in the pelvis of a young adult male, describing the value of MRI and CT.

Case Report

A 32-year-old boy presented with selling and pain in his left lower limb of three months duration. Four days prior to his hospital admission, he developed acute urine retention. Subsequently, he underwent radiological evaluation. X-ray pelvis showed a soft tissue mass displacing the obturator and psoas fat planes. Ultra fast slice plain and contrast enhanced computed tomography (CT) scan [Fig 1,2] , showed a highly vascular, large , lobulated , mixed density mass measuring 13.3 x 12.4x 9.9 cm with speck of calcification , in the left inferolateral aspect of urinary bladder, extending into the upper thigh.

MRI showed heterogeneous tissue processes hyperintense T1 and T2, [Fig 3a, 3b, 3c, 3d] seat empty areas related to stigma hémorragiques. The process was intensely enhanced by the PDC [Fig 3d] at its fleshy portion .He pushes forward and right bladder, seminal vesicle and prostate with loss of separation border.

Anteriorly, the lesion extended till the public symphisis and posteriorly, up to the anterior surface of sacrum without identifiable lysis. All the visceral organs were normal.

Subsequently, a core needle biopsy was performed and the tissue was submitted for histopathological diagnosis.the diagnosis was, malignant tumor proliferation in favor of a Synovial sarcoma.

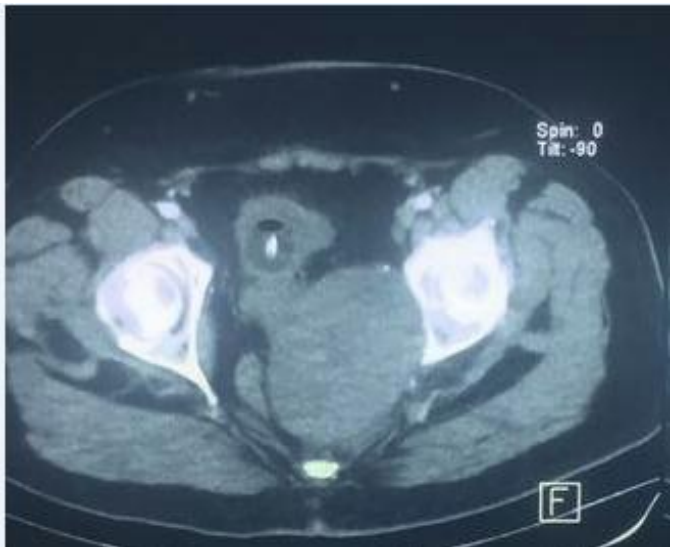

Figure 1a: pelvic CT, axial section, with injection of contrast

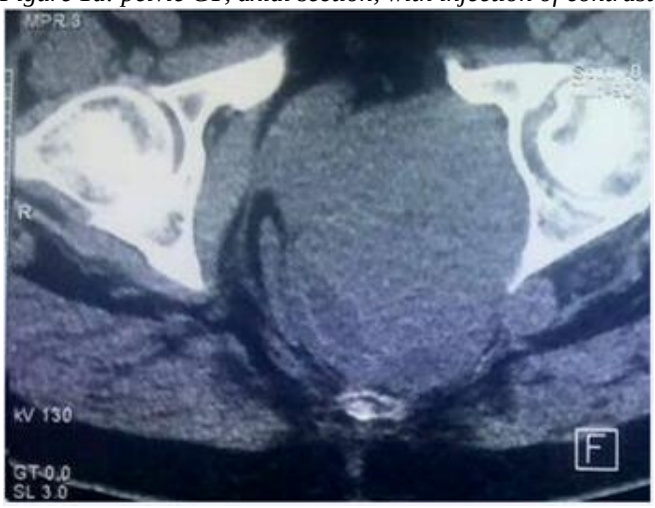

Figure 1b: pelvic CT, axial section, with injection of contrast

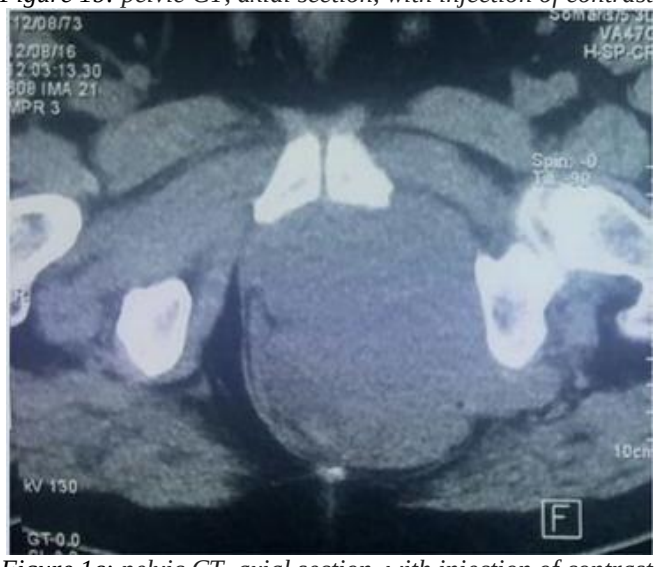

Figure 1c: pelvic CT, axial section, with injection of contrast

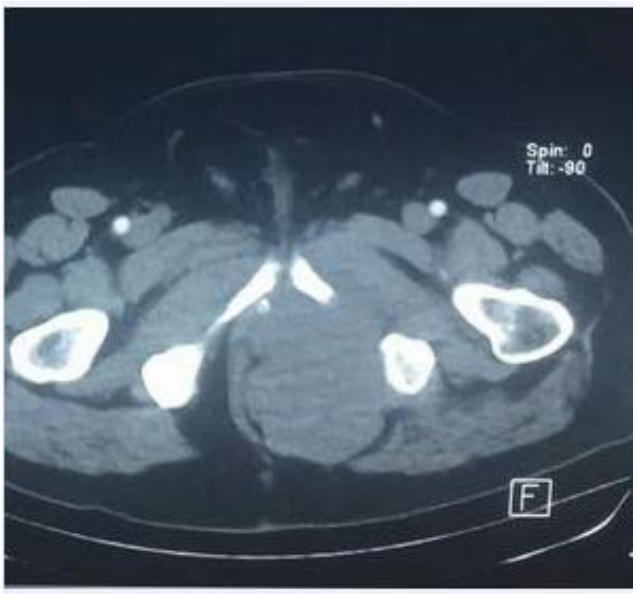

Figure 1d: pelvic CT, axial section, with injection of contrast

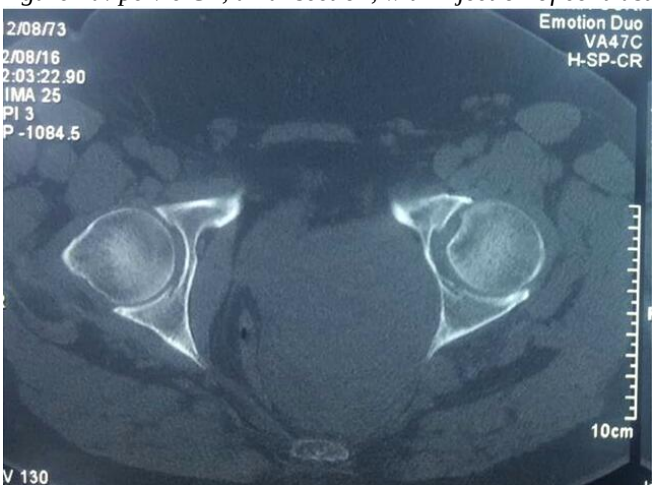

Figure 2: pelvic CT, axial section, with injection of contrast

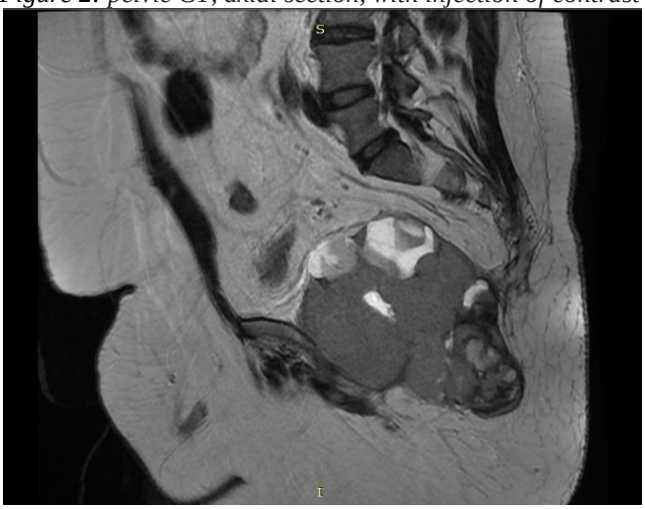

Figure 3a: pelvic MRI sagital T2 sequence without gadolinium

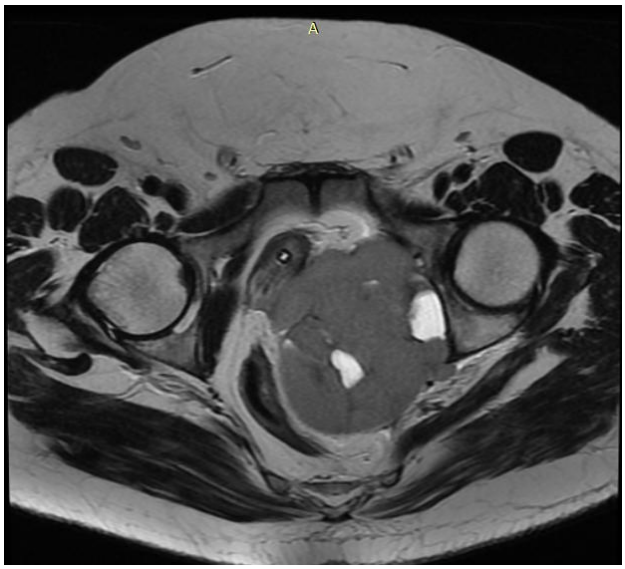

Figure 3b: pelvic MRI axial T2 sequence without gadolinium

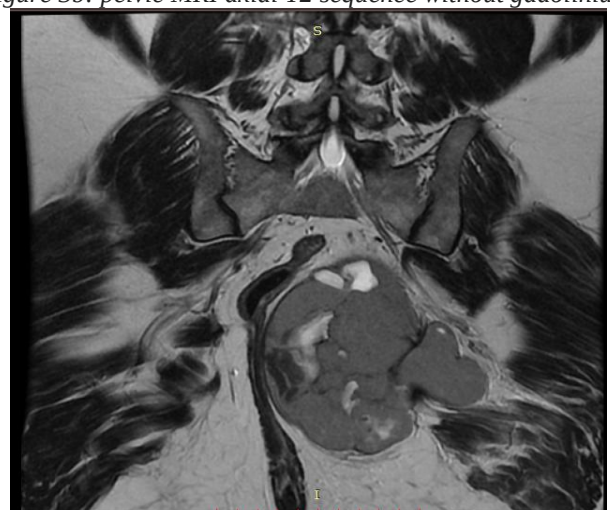

Figure 3c: pelvic MRI coronal T2 sequence without gadolinium

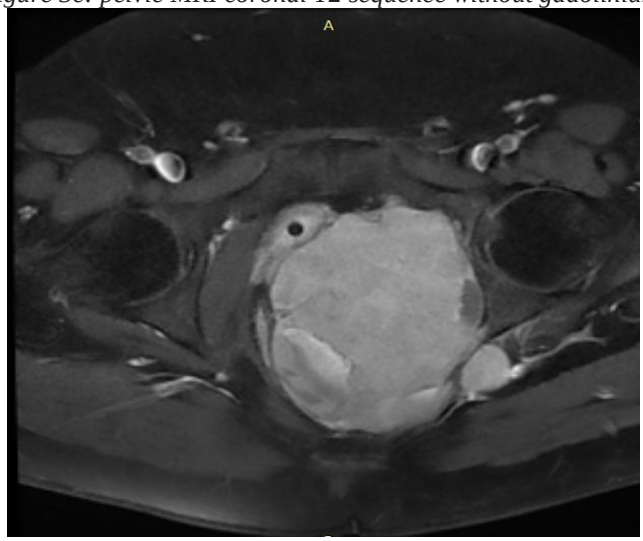

Figure 3d: pelvic MRI axial T1 FAT SAT sequence with gadolinium

Discussion

Synovial sarcoma (SS) forms a distinct clinical, morphological and a genetic type of a soft tissue sarcoma, which has been described in various body sites. Intraabdominal primary SS is unusual [1]. Nearly 50 cases have been documented so far. Still rare, is a pelvis location, wherein only five cases have been identified to the best of our knowledge. Only one such case has dealt with molecular analysis.

The present case of a SS was seen in a young boy, whose radiological evaluation unraveled a large, pelvic mass, a feature that has not been seen with the similar documented cases. The long duration of symptoms and initial slow growth of synovial sarcomas may simulate those of or give a false impression of a benign process [3].

Even though , the present case of pelvic SS of spindle cell type, like the one described by Cole et al exhibited similar SYT-SSX2 transcript , the other parameters forecast an unfavorable prognosis. Apart from site, larger tumor size, poor differentiation, necrosis, apoptosis along with high mitoses are indicators of a grim prognosis, as noted earlier. As per literature, it has been seen that invariably, pelvic SSs are associated with a dismal outcome as a result of local recurrences ans metastasis, latter that has been noted in 4/6 similar documented cases. [4-6]

Surgery is the treatment mainstay. However, marginal clearance is difficult to achieve in this location. Adjuvant chemotherapy (CT) ans radiotherapy (RT) have been gien. Despite that, recurrences ans metastasis have been noted. Our case is a candidate for surgery with adjuvant CT. In a nutshell, this case reinforces value of MRI and CT in SS at uncommon sites.

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