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Case Report

Desmoplastic Fibromas

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Introduction

Desmoplastic fibroma (DF) is an extremely rare tumor with less than 200 cases in published literature. It is a slowly progressive tumor with well differentiated cells that produce collagen. This is a benign tumor characterized by aggressive local infiltration. It occurs most often in the first 3 decades and is found equally in men and women. According to the published data, the tumor is most common in the long tubular bones (56%), the mandible (26%), and the pelvis (14%)[1]. Important clinical feature is pain, swelling and pathologic fractures. The diagnosis of (DF) is difficult to make radiologically. Plain x-ray shows an osteolytic expansile, medullary lesion with well defined sclerotic margins. The CT scan is only useful to further demonstrate cortical breakthrough. MRI demonstrates the separation of the interosseus tumor from the bone. The treatment of DF is marginal or wide surgical excision. Rates of recurrence are 55-72% without resection and 17% with resection. Recently one study has recommended "aggressive curettage" as the surgical option.

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Case report

A 26 year old patient was referred from Ambejogai Medical College with the chief complaints of acute retention of urine, recurrent episodes of urinary retention since 3 months, lump in lower abdomen increasing in size since 7-8 months, dyspareunia since 3-4 months, and pain in abdomen off and on. She had a full term normal delivery 3 years back. She gave history of exploratory laparotomy done in a rural hospital 4 months back for the same symptoms with the diagnosis of pelvic mass. The mass could not be accessed and hence the abdomen was closed. She developed urinary infection.

Clinical examination

General and systemic examinations showed no abnormality. She had a midline scar of exploratory laparotomy and a suprapubic fixed hard mass possible as the mass was obliterating the vagina. On vaginal examination on a huge fixed mass was felt on the right side lateral to the vagina and hard in consistency. On rectal examination the rectum was found displaced towards the left by the mass and its mucosa free. Investigations showed that hemoglobin was 13g/dL. Renal and liver functions were within normal limits and routine urine examination was normal; HIV, VDRL and HBsAg were negative. Chest x-ray was normal. Abdominal sonography showed a well defined 12x10 cm round mass with mixed echogenicity in the right pelvic cavity with normal uterus and ovaries. Cervix

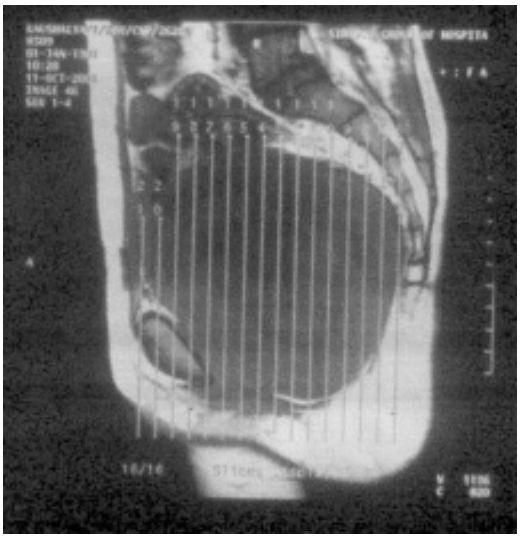

Figure 1. Pelvic MRI (Section).

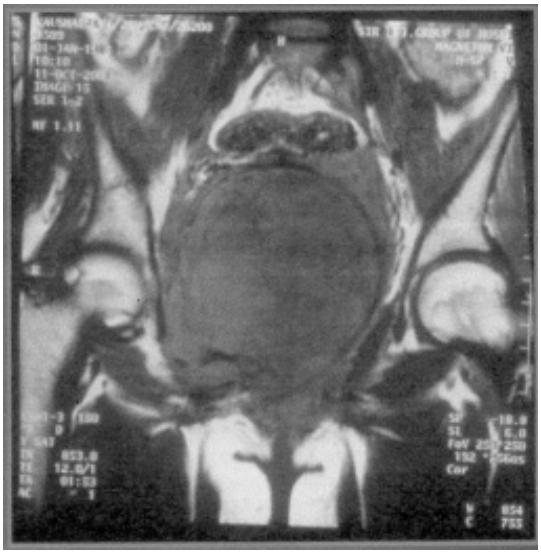

Figure 2. Pelvic MRI.

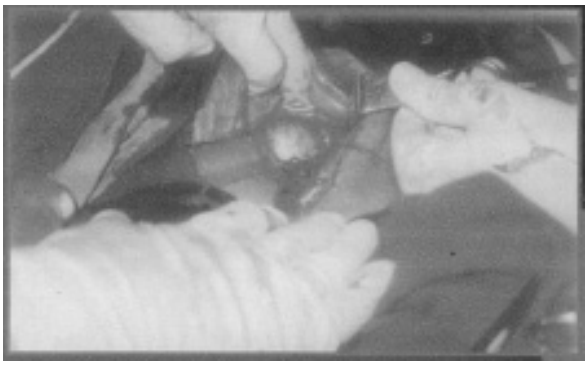

Figure 3. At laparotomy.

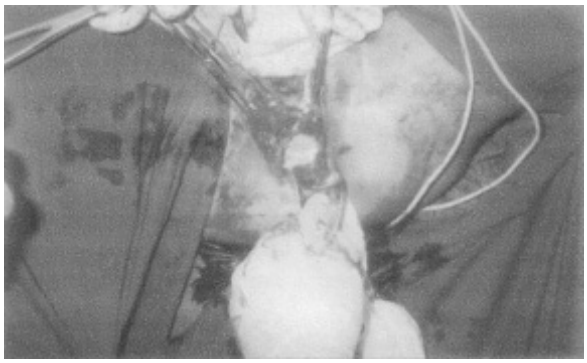

Figure 4. Perineal Resection.

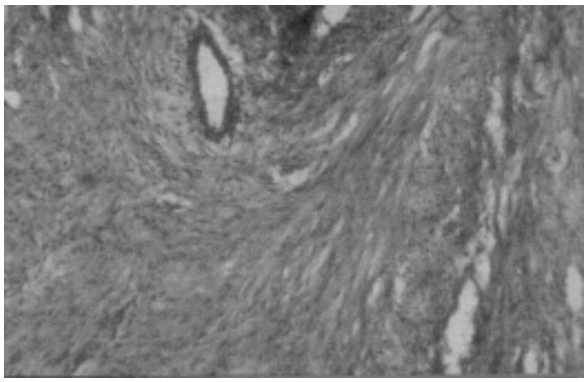

Figure 5. Histology.

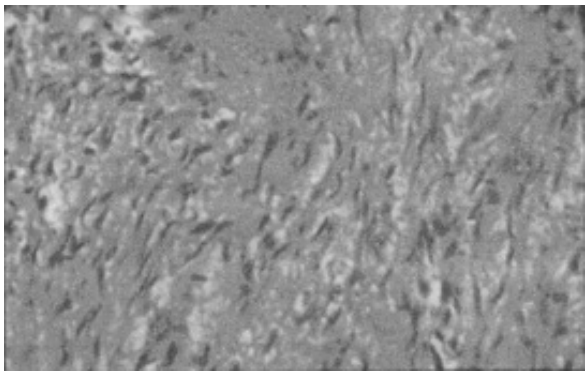

Figure 6. Histology.

and vagina were not seen separately from the mass. CT scan showed a large well defined mass appearing to involve the right obturator muscle and normal lymph nodes.

MRI (Figure 1 & 2) showed a large encapsulated heterogeneous mass pushing the bladder and the uterus anteriorly and superiorly to the left, and extending into the introitus. Right obturator muscle appeared hyper intense. Right ureter was displaced laterally. Fine needle

aspiration cytology suggested a diagnosis of either neurofibroma or desmoplastic fibroma. CA 125, α feto proteins, βhCG and CEA were normal.

She was admitted to Sir JJ Government Hospital on 10th October, 2001 for surgery. At laparotomy (Figure 3) done on October, 2001 the pelvic cavity was seen completely obliterated and a fetal head size mass was occupying the cavity. Uterus was displaced to the left lateral pelvic wall, right round ligament was lengthened and stretched to approximately 20 cm due to the large mass which was retroperitoneal in origin. The mass was 14x12x10 cm in size. The following structures were adherent to the mass a) bladder in the trigone area with the mass compressing the bladder and the urethra against the pubic bone b) right ureter which was compressed against the right pelvic wall and c) rectum, sigmoid and mesocolon. The posterior peritoneum was opened on the right side to dissect and enucleate the mass. But it was not possible to completely dissect out the mass. Keeping in mind the vaginal protrusion, Schuchardt's incision was taken on the right side and the mass dissected inferiorly by sharp dissection (Figure 4). It was impossible to dissect the left side of the mass due to presence of the uterus and cervix superiorly as it occupied the whole pelvic cavity and was retroperitoneal. Attempts to pull the mass with myoma screw were done but failed and the decision for hysterectomy taken as the dissection on left side was impossible. Bilateral ligation of internal iliac arteries was done prophylactically to ensure less intraoperative blood loss. After the completion of dissection the mass had a pedicle with its origin from the inferior margin of the right pubic bone; it was cut by a chisel.

Since the mass adherent anteriorly to the bladder wall eroding its musculature was close to the trigone, the bladder got opened during the dissection. The bladder was closed in three layers, ensuring the patency of the ureteric orifices. Post operatively a 5x5 mm vesicovaginal fistula near the right ureteric orifice was noticed on the 15th day probably due ischemic necrosis. It was repaired 3 weeks later. It healed well and the patient was discharged on 25th November, 2001.

The mass was arising from the inferior margin of the right pubic bone attached by a pedicle. It was cut by sharp dissection with the help of a chisel.

Discussion

DF was described as a distinct entity by Jaffe[2]. It is a primary benign fibrous tumor of the bone, histologically identical to the more common soft tissue desmoid tumor (Figure 5 and Figure 6). DF is a rare, nonmetastasizing but locally aggressive tumor[3]. The aggressive nature of this tumor was emphasized by Inwards et al[4] and Bohm et al[5]. Although considered a benign lesion, it is locally destructive, and can extend into the soft tissues, having a high rate of local recurrences after incomplete surgical excision. Therefore cognition of this entity is important to ensure proper surgical treatment.

Most lesions are subcutaneous but approximately 25% involve skeletal muscles. Grossly, they are pearl gray firm and homogenous, and appear well circumcised but microscopically they can be seen to infiltrate the surrounding fat or muscle[7]. Most authors[4-6] emphasize that low grade fibrosarcoma poses the most difficult problem in histological differential diagnosis of DF, even for the experienced pathologist because the features of both tumors merge imperceptibly with one another[3-4]. In the era of advanced imaging modalities of CT and MRI, pelvic masses can be properly pelvic masses can be properly diagnosed. Pelvic masses shoud no longer be an 'Enigma' in future. Operate on these patients in view of the local invasiveness of these tumors and their adhesion to important neighboring structures.

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