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KEY WORDS: Retroperitoneal sarcoma, liposarcoma, irreducible inguinal hernia, giant retroperitoneal mass

GIANT RETROPERITONEAL SARCOMA PRESENTING AS IRREDUCIBLE INGUINAL HERNIA

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ABSTRACT

INTRODUCTION - Retroperitoneal sarcomas constitute 15% of soft tissue sarcomas . Common histological type in the retroperitoneum is liposarcoma and leiomyosarcoma. These tumours displaces the intra abdominal organs rather than invading them. It is diagnosed on CT and MRI. Surgery is the mainstay of treatment. Adjuvant radiation therapy has shown improvement in outcomes. **CASE REPORT -** 45/M came with complaints of swelling in the right inguinal region for 3 years and was associated with abdominal distension and vague dragging type of pain. CT abdomen revealed fat density lesion 11 x 25 x 33cms from anterior pararenal space displacing bowel and herniating through right inguinal canal suggestive of retroperitoneal liposarcoma with right inguinal extension. Patient was taken up for surgery and a tumour was removed en bloc weighing 8.5kgs. Post operative histopathology showed liposarcoma well differentiated type grade 1 of size 46 x 36 x 13cms. **CONCLUSION -** Preoperative clinical suspicion of retroperitoneal involvement in a case of irreducible inguinal hernia is paramount for developing a surgical strategy. Unclear cases require preoperative diagnostic workup. Liposarcoma represents 60% of all retroperitoneal sarcomas. It is a malignant locally aggressive tumour. It is classified as well differentiated and dedifferentiated tumours. Prognostically these tumours have a predilection for relapse.

INTRODUCTION:

Retroperitoneal sarcomas are rare tumours accounting for only 1-2% of all solid malignancies(1). They constitute 10-20% of soft tissue sarcomas . Peak incidence is in the 5th decade, Although they can occur at almost any age.(1) Common histological type in the retroperitoneum is liposarcoma, leiomyosarcoma and undifferentiated and unclassified sarcomas.(4) These tumours grow to very large size without producing any symptoms. They present late because they arise in large potential spaces of retroperitoneum.(1) These tumours displaces the intra abdominal organs rather than invading them. (6) On occasion, patients may present with neurologic symptoms, resulting from the compression of lumbar or pelvic nerves, or obstructive gastrointestinal symptoms, resulting from the displacement or direct tumour involvement of an intestinal organ.(4) It is diagnosed on multiplanar imaging with CT and MRI with reconstruction. It is required not only for tumour detection , staging and surgical planning but also for guiding surgical biopsy of these tumours.(1).

Case Report:

We report a case of 45/M, who came with complaints of swelling in the right inguinal region for the past 3 years and vague dragging type of pain in the abdomen for past 6 months. Patient had no complaints of vomiting or constipation. There is no history of loss of weight or loss of appetite. There is no history of urinary disturbances. He was a non smoker and a non alcoholic. There is no history of scrotal pain. There no history of fever , night sweats or fatigue. On examination, abdomen was distended with umbilicus in the centre. All quadrants were moving equally with respiration. An ill defined mass was palpable in the right lumbar and iliac region that seemed to extend across the midline to the left lumbar region. It was non pulsatile and intra abdominally located. A swelling was palpable in the right inguinal region and was seen extending into the scrotum upto base. It was irreducible and no cough impulse palpable. External genitalia was normal. There was no generalised or regional lymphadenopathy. Patient was subjected to radiological investigation. CT abdomen and pelvis revealed fat density soft tissue lesion measuring 11 x 25 x 33cms arising from anterior pararenal space displacing bowel and herniating through right inguinal canal suggestive of retroperitoneal liposarcoma with right inguinal extension. MRI of the abdomen and pelvis showed large ill defined soft tissue density lesion with mixed intensity measuring 24x23x29cms and was reported as a case of retroperitoneal liposarcoma. Patient was taken up for laparotomy and the tumour was seen arising from the retroperitoneum pushing the bowel anterior and to the left side. It was seen crossing the midline extending into the mesentery of small bowel, sub hepatic region and right inguinal canal. There was no evidence of involvement of any vital abdominal organs or major blood vessels. There was no enlarged lymph nodes within the abdomen. Hence tumour was removed en bloc and right hernioplasty was done. It measured 44 x 35 x 11 cms and weighed about 8.5kgs. Post operative histopathology showed atypical lipomatous tumour/ liposarcoma well differentiated type

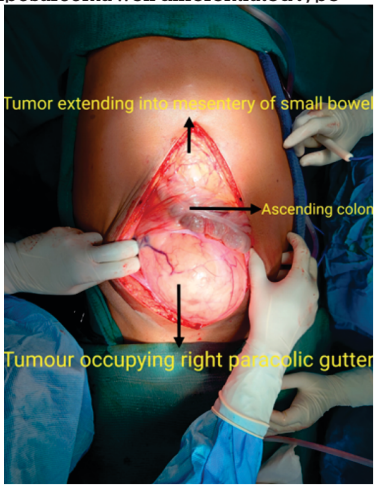

Fig 1. Tumour Pushing The Bowel Anteriorly

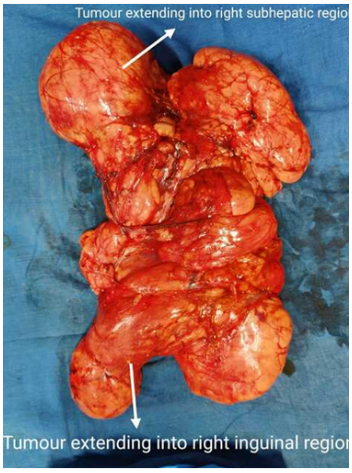

Fig 2.Tumour Excised In Toto 1 (fnclcc).

DISCUSSION :

Retroperitoneal sarcoma presenting as inguinal hernia is a rare condition. A retroperitoneal tumour especially a liposarcoma should be though of as a differential diagnosis in a case of large , painless , slow growing inguinal mass that is irreducible associated with abdominal symptoms. Since the retroperitoneal sarcomas arise in a area of large extent without any anatomical barriers they grow upto huge size. Deep inguinal ring and spermatic cord structures act as a potential doorway for tumours to grow from retroperitoneum into the scrotum.(4) Liposarcoma, is the most common variant of retroperitoneal sarcomas and it represents 60% of all retroperitoneal sarcomas. It is a locally aggressive tumour composed of adipocytes.They carry cells with nuclear atypia and hyperchromasia. They are classified as well differentiated liposarcoma and dedifferentiated liposarcoma. Abdominal radiography provides minimal information regarding the retroperitoneal sarcomas. Ultrasound scanning helps identifying the size and location of the tumour. A more precise determination of the size, extension and its relation to adjacent organs is done using computed tomography. It helps in adequate preoperative evaluation and planning the surgery. (6) Over-expression and/or amplification of genes localised to chromosome 12q are cytological feature of liposarcoma including MDM2, S100 and CDK4. Prognostically these tumours have a predilection for relapse. In well differentiated tumours, upto 40% tumours will recur locally. Average reported time for recurrent disease is 2 years. The risk of local recurrence depends partly on tumour biology and surgical factors. Most would agree that negative margins optimise outcomes for patients(1). The goal of surgical management for patients with retroperitoneal sarcoma is removal of all gross disease with en bloc resection of adherent organs. Surgical removal is considered the gold standard in treatment of retroperitoneal liposarcoma in both primary and recurring tumours(8'9'10) . The resectability of the tumour does not rely upon its size, subtype or histologic grade.(8) The difficulty in achieving complete and curative removal of the tumour lies in its relations to neighbouring structures.(7) In order to obtain a complete removal of the tumour, 50% of the patients must remove some part of adjacent organs, such as the kidney, ureter, and portions of the large intestine. (6) Current strategies involving radical compartment resections for liposarcoma have reported to improve local recurrence rates. The presence of local recurrence is observed both patients with negative and positive gross margins, which excludes gross surgical margins as a factor predictive of recurrence. (6,11) A large case-control study using a national database found an association with improved survival with the use of radiation therapy, either pre- or postoperatively, compared to resection alone.However, retroperitoneal sarcomas pose a challenge due to their larger size and toxicity to adjacent organs (e.g., kidney, spine, liver, or small bowel), which limits its use in the adjuvant setting. (2) In unresectable or metastatic disease use of anthracycline based regimens has been associated with modest improvements in short term survival. (3)

CONCLUSION :

Pre operative clinical suspicion of retroperitoneal involvement is paramount for developing of a surgical strategy and in unclear cases demands extended preoperative diagnostic work up. (5)Following appropriate patient management is crucial to prognosis.

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