International Surgery Journal
Ghatnekar A et al. Int Surg J. 2019 Nov;6(11):4135-4137
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20195139

Case Report

Rare case of giant retroperitoneal sarcoma

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Received: 09 August 2019
Revised: 26 September 2019
Accepted: 30 September 2019

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ABSTRACT

Schwannomas are benign nerve sheath tumors. Malignant schwannomas have been reported in literature but they frequently originate from the conversion of plexiform neurofibromatosis rather from the malignant degeneration of a benign schwannoma. Large retroperitoneal schwannomas are extremely rare tumours. They usually reach a large size when they arise retroperitoneally, which might raise suspicion for malignancy. Such tumors predominate in women schwannomas are usually solitary, slow growing and non- aggressive neoplasm usually found in head and neck region. Retroperitoneal schwannomas are usually large size are rare and are difficult to diagnose clinically. Nevertheless, the mainstay of management remains surgical removal and alternatively, the radio-therapy may prove to be effective. This case report of a 55 yrs male is based on such a patient in whom the diagnosis was established with great difficulty and intraoperative findings were something we had never encountered before.

Keywords: Retroperitonel sarcoma, Giant tumors, Sarcomas, Giant retroperitoneal tumors, Schwannomas, Malignant peripheral nerve sheath tumors

INTRODUCTION

Schwannomas are benign nerve sheath tumors. The malignant schwannomas have been reported in literature but they frequently originate from the conversion of plexiform neurofibromatosis rather from the malignant degeneration of a benign schwannoma.[1,2]

Principally, large retroperitoneal schwannomas are extremely rare tumours. They usually reach a large size when they arise retroperitoneally, which might raise suspicion for malignancy. Such tumors predominate in women. Due to atypical or non-specific clinical presentation or lack of specific laboratory data, a preoperative diagnosis and tumor's origin is difficult to achieve or define respectively.[3,4] Nevertheless, the mainstay of management remains surgical removal and alternatively, the radio-therapy may prove to be effective.

Regarding prognosis, 10 and 20-year survival among patients having onset of symptoms at younger age (<25 years) is 80%, 60% and 28%, respectively.[5] In contrary, the rate of survival among patients with onset of symptoms more than 25 years of age is 100%, 87% and 62%, respectively. Better survival rates are reported in patients with smaller size of tumor at the time of diagnosis (<2 cm in diameter).[6]

CASE REPORT

A 55 years old male came with complaints of lump in abdomen since 10 years, pain in abdomen since 2 months. Initially lump small in size then it is progressed to present severity. History of pain in abdomen aggravates on food intake. No history of bowel and bladder irregularities.

On examination: A visible lump seen in right iliac and right lumbar fossa, divarication of rectii seen.

A 20×15 cm well defined lump palpable, superiorly from 6cm below right costal margin, medially 2 cm lateral to umbilicus, inferiorly 4 cm above inguinal ligament, hard in consistency, smooth surface, non-ballotable, moves with respiration.

USG abdomen and pelvis: A large heterogeneously hypoechoic lesion measuring 20×20×12 cm appears to be retroperitoneal in origin and few satellite lesions noted with largest measuring approximate 6×5×4 cm.

Large mass lesion seen in pelvis measuring approx 10×11×9 cms.

CT scan abdomen and pelvis: Large mass lesion in right half of abdominal cavity predominantly in the right iliolumbar region. The lesion shows multiple lobulated satellite lesions at its periphery. The lesion measures about 25×16×16 cms.

Also it shows a lobulated heterogenous enhancing soft tissue lesion of 4.4x4 cms is noted in the left lumbar region within small bowel mesentry.

Patient has underwent excision of retroperitoneal mass.

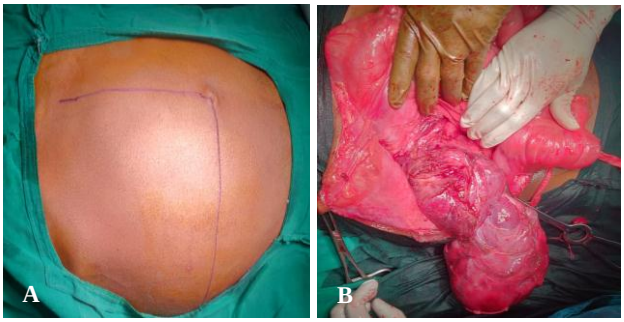

Figure 1 (A and B): Skin Incision marking, intraop picture of the tumor.

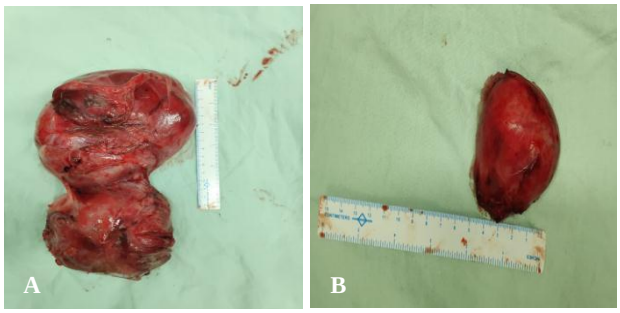

Figure 2 (A and B): Excised specimen of the tumor.

Intraop findings

E/o 6×5cms mass seen in the mesentry of ileocaecal junction.

E/o 3×4 cms polypoidal mass arising from ileal mesentry diverticulum.

E/o large 25×12 cms bilobed encapsulated solid cystic vascular mass arising from retroperitoneum.

E/o 6×5 cms solid mass arising from the retroperitoneum.

HPE of both the masses was s/o malignant preipheral nerve sheath tumor of moderate grade, and is S-100 positive.

DISCUSSION

The malignant peripheral nerve sheath tumours (MPNST) is defined as any malignant tumour differentiating into or deriving from the peripheral nerve sheath cells, with nonspecific symptoms, having high risk of recurrence locally and distant metastasis.[5,8] MPNST is particularly rare, affecting general population with an incidence of 0.001% and extremely rarespindle cell sarcoma in children, accounting for approximately 5-10% of non-rhabdomyosarcoma soft tissue sarcomas.[8,9]

Schwannoma are tumours that are arise from the Schwann cells of peripheral nerve fibre and are generally found in the head, neck and extremities.[10] Retroperitoneal schwannoma are more commonly present in the paravertebral spaces and pre sacral region.[12] If present in such locations, it is usually in association with Von Recklinghausen's disease. Among schwannoma that occur retroperitonealy, around 0.7% are benign and 1.7% were reported to be malignant yet producing vague symptoms such as abdominal pain and distension.[11] The Schwannoma are predominant in men among the age group 20 to 50 years. The commonest site is head and neck region and flexor tendon sheaths of extremities but sites may vary. The tumor at unusual locations such as the fallopian tubes, scrotum or urinary bladder has also reported in literature. Schwannoma in pelvis are very uncommon and comprises of only 1% of the cases.[13] These pelvic tumours usually present with non-specific symptoms like pain, rectal dysfunction or with palpable mass. Literature has also reported.

All MPNST characteristically demonstrate intense and uniform staining for S-100 protein.[5] These are more aggressive and are likely to have a high incidence of local recurrence after excision. Chances of distant metastasis are also high. Malignancy cannot be ruled out pre-operatively or by frozen sections, hence, it is recommended that complete excision with negative margins is achieved. Recurrence usually occurs within six months of surgical excision and recurrence rates vary from 16% to 54%.[12]

MPNST are usually solitary, slow growing, and non-aggressive neoplasm.[6] Majority of these tumours are asymptomatic and usually diagnosed incidentally or may present clinically with non-specific or vague symptoms.[2]

On routine clinical examination, they are usually detected as an abdominal mass as in this case. The predominantly cystic nature of the tumour can demonstrate by ultrasound and CT scan examination but the exact nature cannot be diagnosed.[1,9] Malignant transformation is rare and malignant schwannoma are large in size and are highly aggressive. They are painful, and may cause many different symptoms depending upon the location and size of the tumour.[13]

In retroperitoneal MPNST, these findings are not present which make the diagnosis quite challenging. Ultrasonography is useful in taking guided biopsy while CT scan helps in detecting associated malignant changes such as bony erosions.[3] If malignant change is present, irregular margins with infiltration into adjacent structures are seen on the scans.[10,12] In a study carried out in Japan, 94 (72.3%) cases of schwannoma turned out to be benign and 36 (27.7%) were found to be malignant.[12]

Quite infrequently schwannoma's undergo malignant transformation invading neighbouring organs for instance colon, kidneys, and adjacent viscera1. Tissue diagnosis is established with FNAC and excision biopsy CT guided biopsies and FNAC are not always reliable as it is required that the sample contains a significant amount of schwann cells.[9] FNAC from the areas of degeneration may lead to a misdiagnosis of malignancy. At the same time, FNAC increases the risk of haemorrhage, infection, tumour seeding and as a result, it is not the preferred choice of many surgeons. There is a lack of specific standardised criteria for diagnosing malignant changes except for certain characteristics such as dense fascicules in marble like pattern comprising of asymmetrically tapered spindle cells.[7]

Due to their insensitivity to radio and chemotherapy the malignant peripheral nerve sheath tumour carries a poor prognosis. Immuno-histochemical positivity for S-100 protein is a constructive diagnostic finding. Combinations of clinical, pathological and immune-histochemical studies usually help establishing a final diagnosis.[13]

In cases when the postoperative histology gives evidence of the tumour being malignant after marginal excision, local recurrence has to be expected in up to 72% of cases. However, recurrence of the tumour after resection involving wide surgical excision margins has been reported in only 11.7% of the cases.[3] In cases of unexpected event of proven malignancy, re-resection should be considered if a wide margin has not been achieved initially.

CONCLUSION

MPNST (malignant perineural nerve sheath tumour) has a poor prognosis because of their insensitivity to chemotherapy and radiotherapy so early diagnosis and complete excision is the mainstay of treatment to improve prognosis.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Ghatnekar A, Nichkaode P, Athavale VS, Tonape T, Patil K, Srilikhitha K, et al. Rare case of giant retroperitoneal sarcoma. Int Surg J 2019;6:4135-7.