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Microsurgical approach to a Presacral Schwannoma with internal iliac venous encasement

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Abstract

Sacral schwannomas are very rare and encompass approximately 1%–5% of spinal schwannomas. Due to their benign nature and slow growth, these tumors are often giant and become difficult to treat. Their removal is a surgical challenge with different strategies reported in the literature. Presacral Schwannomas encased by the internal iliac vein are rare with not many instances of such a case found in literature. This is a case in which the venous encasement was found intraoperatively, following which the surgery was abandoned to avoid excess intraoperative bleeding and to enable further workup like CT scan and CT angiography to delineate the venous component. A multimodal approach was then taken for the excision of the tumour.

Keywords: Presacral, Schwannoma, Venous encasement, Microsurgery.

Introduction

Schwannomas are encapsulated, benign neoplasms which arise from the myelinated nerve sheath. They occur commonly in the thoracic region, followed by the cervical and lumbar regions. (1) Sacral schwannomas are very rare and encompass approximately 1%-5% of spinal schwannomas. In the sacral region they usually arise from the sacral nerves found outside the spinal canal and can present with a variety of symptoms as a result of pressure effects on abdominal organs or neural tissue and bony destruction. (2) The compliance retroperitoneal space delays their discovery. The clinical presentation is often misleading or even

absent although the radiological appearance indicates the diagnosis. This space is currently well analysed by computed tomography (CT) and magnetic resonance imaging (MRI). Their resolution in density provides a spatial, morphological as well as tissue analysis that helps determine the type of lesion and plan the surgery. This article reports a case of a presacral schwannoma where multiple modalities were used to assess the tumour preoperatively, following which a multimodal approach was taken to successfully excise the tumour.

Materials and Methods

A 34 year old male presented with complaints of generalized abdominal pain which was non radiating and non referring with no associated bowel or bladder complaints. Per abdomen was soft and nontender. The patient was managed on OPD basis following which the patient's symptoms were relieved and was asked to follow up with an ultrasonography of the abdomen.

USG was suggestive of a large well defined thick walled cystic lesion measuring 9.7x9.6x9cm in the left pelvic region (separate from bowel and prostate) with internal septations. The lesion was suspected to be a cystic schwannoma.

MRI was done suggestive of a large well defined solid cystic lesion measuring 9.5x8.6x8.5cm (APxTRxCC) seen in the left half of the presacral region with peripheral enhancing solid component and a few internal septations. Lesion causing a mass effect on the rectum, left distal ureter, left side seminal vesicles and urinary bladder with its displacement. The fat planes with the rectum maintained. No obvious bony sacral involvement present. No obvious intraspinal canal extension noted. Findings suggestive of neoplastic etiology possibly cystic schwannoma. (Figure 1)



Figure 1: Saggital cut of MRI demonstrating the schwannoma

Our patient underwent exploratory laparotomy for excision of the schwannoma. Anterior approach was taken for the same using a midline incision. Intraoperatively, it was found that the tributaries of the internal iliac vein were encasing the tumour forming an extensive mesh around it. (Figure 2) A tissue sample was collected to be sent for histopathological examination and the procedure was abandoned so as to not cause excessive blood loss and to adopt a multidisciplinary approach for further management of the tumour.

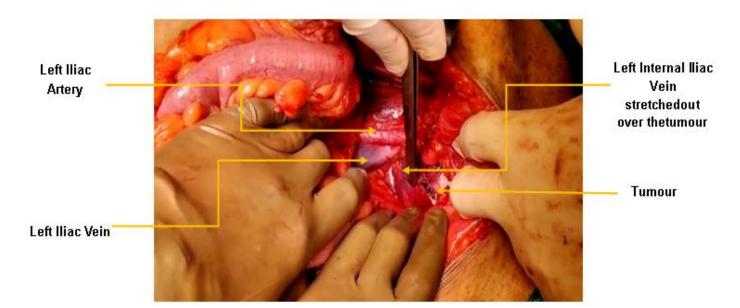


Figure 2: Intraoperative image of the tumour demonstrating venous encasement

The patient was then worked up further and a contrast CT scan (Video 1) was done for this patient post-operatively in order to delineate the iliac venous encasement around the tumour. CT revealed a well-defined peripherally enhancing cystic lesion measuring 7x7x8.2 cm in the presacral region predominantly on the left side with a 2.6 cm irregular crescenteric solid component along its left lateral aspect with a few calcific changes in the soft tissue component.

- It was found to be abutting the distal part of the ureter, sigmoid colon and urinary bladder which were displaced anterosuperiorly
- Posteriorly seen to be abutting the lower sacral vertebra and the sacral attachment of the left piriformis muscle with loss of fat plane. No sacral erosion or foraminal widening noted.
- Medially causing compression of the proximal and mid rectum displacing it to the right side

- Laterally abutting the obturator internus muscle, left internal iliac artery (IIA) and common iliac vein
 - The left IIA seen draping along the left lateral aspect of the lesion. No evidence of occlusion or thrombosis noted
 - The internal iliac vein (IIV) was not visualized ?thrombosis ? occlusion ?compressed ?involved
 - Superiorly it is seen to be abutting the left ureter anteriorly and the upper sacral vertebra posteriorly
 - Inferiority it is seen to be abutting the distal part of the rectum
 - However fat planes with the lesion are maintained

Findings were suggestive of a neoplastic ethology of neurogenic origin, most likely a schwannoma. (Figure 3)

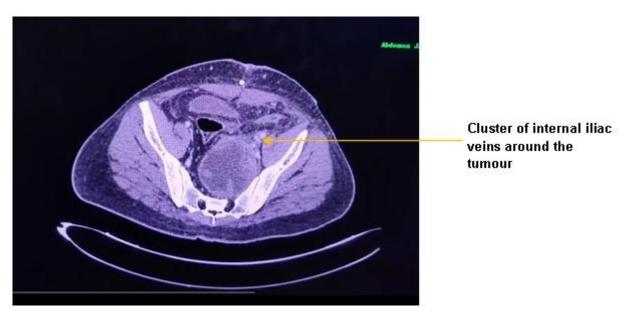


Figure 3: Cross sectional cut of CECT scan demonstrating a cluster of veins around the tumour

Further a CT angiography was done under interventional radiology to better delineate the vasculature around the tumour. It revealed that the distal left IIA and its anterior trunk was displaced laterally due to avascular mass lesion

corresponding to the CT. The venous phase appeared normal with no evidence of thrombosis and the left internal iliac vein was seen encasing the tumour. (Figure 4)

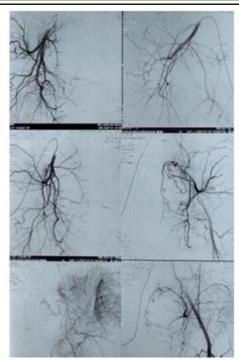


Figure 4: CT Angiography image of the tumour

The patient was then posted for surgery with cardiovascular surgery and neurosurgery teams. An anterior midline incision was taken following which the internal iliac vessels were secured by the CVTS surgeon. Dissection was done until the tumour was visualized. Dissection was started around the tumour to separate it from the surrounding tissue. There was venous bleed from the tributaries of internal iliac vein encasing the tumour which was controlled by ligating those tributaries. As the venous mesh around the tumour increased, the neurosurgeon was called and further dissection was continued under microscopic guidance by the neurosurgeon. The surgery was completed with complete excision of the tumour. The specimen and the cystic fluid within the specimen were sent for histopathological evaluation.

The patient tolerated the procedure well and was discharged after observation of 10 days.

Histopathology report of tissue was suggestive of a tumour showing Hypercellular (Antoni A) and hypocellular (Antoni B) areas. The tumour cells were arranged in bundles. The individual tumour cells were elongated with buckled nuclei and fine chromatin. Also seen were a few thick walled, hyalinised and congested blood vessels. No evidence of necrosis or mitosis present.

Examination of the cystic fluid revealed cells with degenerative changes.

Follow up of one year has shown him to be disease and symptom free.

Discussion

Schwannomas are benign neoplasms arising from nerve sheath. Although malignant schwannomas have been described, they commonly arise from the transformation of plexiform neurofibromatosis, and not from malignant degeneration of the schwannoma. Schwannomas occur predominantly in men aged 20 to 50 years. Sites may vary, but they most commonly occur in the head and neck region as well as flexor tendon sheaths of extremities. Unusual locations such as the bladder, scrotum and fallopian tubes have been reported. Schwannomas occurring in the pelvis are rare and account for only 1% of cases. Such tumors may present with non-specific pain, palpable mass or with rectal dysfunction. (4)

Preoperative diagnosis based clinical on examination is very difficult and so the role of imaging is important. Ultrasonography is a cheap modality for revealing a mass with semisolid or cystic areas, but it is not used widely due to specificity limitations. CT scan and MRI are more helpful in detecting specific characteristics of the tumor. Size, exact location, relationship with other organs and invasion can be accurately reproduced. In our case, preoperative diagnosis was made on the basis of the MRI report. Angiography has also been reported by some authors because of the hypervascularity of these tumors and possibility of embolization, but is not widely used. (5) We used angiography in our patient due to the extensive encasement of the tumour by the left internal iliac vein. Our case reported no thrombosis of the vein.

Surgical treatment of schwannoma aims at its complete removal. Problems with reaching the aim stem from the size of the tumour, its rich

blood supply and vicinity to important anatomic structures. The tumours in this area are usually supplied with blood through sacral arteries, medial and lateral arteries, lumbar arteries and internal iliac arteries. In our case the tumour was encased by the internal iliac veins. During the surgery, the surgeon should focus on preserving continuity of sacral nerve roots (S1, S2, S3) which exit from sacral bone through pelvic foramina, fused with the capsule of the tumour. Damage to the nerves results in dysfunction of vesical and anal sphincter muscles, disturbs sexual functions and leads to paresis of the gluteus maximus Microsurgical techniques have been employed in this case to preserve nerve roots as well as for vascular control. (6)

Hemorrhage is a serious intraoperative problem in cases that major vessels are situated nearby the tumor and there are several reports of unsuccessful tumor excision or even intraoperative death. ⁽⁵⁾ In our case, microsurgery was used to avoid excessive blood loss due to gross bleeding from the internal iliac tributaries leading to complete excision and no recurrence and resulting in the patient getting discharged after 10 days of surgery.

Conclusion

Vascular mapping is an essential step for planning excision of giant schwannomas. A multidisciplinary approach involving a vascular surgeon, neurosurgeon and microscopic dissection can be undertaken to facilitate excision and for a better outcome.

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