CASE REPORT

Excision pelvic schawanoma under neurophyiological monitoring: A rare entity

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Abstract:
24 years old male man having the history of electrical pain and foot drop admitted under services of neurosurgery in King Abdul Aziz Medical City Jeddah. Patient underwent diagnostic workup and on CT scan reveals that large pelvic mass which is about 14x9 cm arises from the sacral foramen and nerve roots with highly vascular mass. It is confirmed on biopsy as pelvic schawonoma. The case was referred to surgical oncology team for further evaluation and management plan of care. This is highly challenging case and we had arranged multi-disciplinary team meeting including surgical oncology, neurosurgery, and spinal surgery interventional radiology and radiation oncology teams. After detailed discussion for management plan we had operated patient with anterio-posteior approach and did subtotal piecemeal excision of tumor, sacral and illac bone excision with lumbosacral spine fixation and excision of tumor from nerve roots under neurophysiologic monitoring and as result we did successful excision of tumor without any kind of neurological complications.
Conclusion: Pelvic schawonoma is the rare tumor, need proper evaluation with CT scan, MRI and histopathology and successful treatment is not possible without multidisciplinary team. The complete treatment includes excision of tumor, fixation of sacrum.

Keywords: pelvic schawanoma, multi-disciplinary approach, pelvic fixation, neurophysiologic monitoring, retroperitoneal tumours

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Introduction:
Schawanoma is the slow growing benign tumor arises from the nerve sheath of the peripheral nerve. The presentation of the schawonoma is late because of its slow growing nature and patient develops symptom when the size of the tumor increases and they causes pressure effect on the abdominlpelvic organs.[1] Retroperitoneal schwannoma (also termed neurilemmomas or neurinomas) is a rare entity consisting of only 0.5% to 12% of all retroperitoneal tumors.[2] Majority of schwannomas is found in peripheral nerve fibers in the limbs, head, and neck. In the retroperitoneal position, they occur most commonly between 40 and 60 years of age, with a male/female ratio of 2:3.[3]

Case Report:
24 years old military man having history of electrical pain radiation from back to right lower limb with weakness admitted under neurosurgery department King Abdul Aziz medical city Jeddah. On investigation we identified huge solid cystic mass arises from foramina L5 S1 S2 with high vascularity. On the basis of radiological finding it was suggestive of sacral plexus pelvic schawonoma with pressure effect on the pelvic organ. Patient was underwent open biopsy and it was diagnosed as pelvic schawonoma.

This is combine type of retroperitoneal intrasacral spine schawonoma. The multi-disciplinary meeting were carried out including surgical onolocogy team, neurosurgery team, spinal sur-

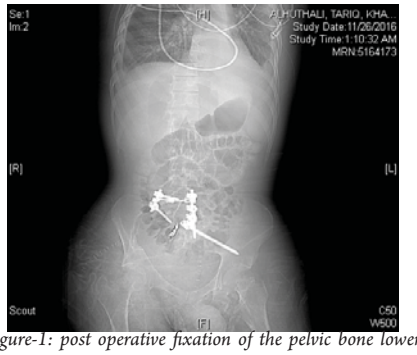

Figure-1: post operative fixation of the pelvic bone lower lumber and sacral vertibrae

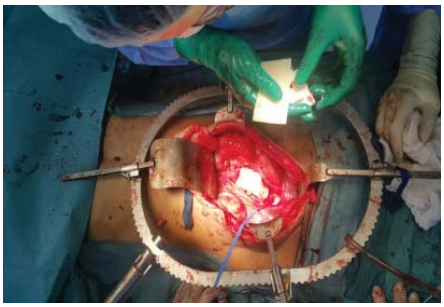

Figure-2: inside view of pelvic schawanoma

Figure-3: piece meal removal of the huge schawanoma

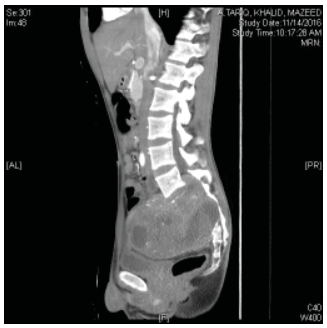

Figure-4: CT scan showing the huge tumor involving the secrum

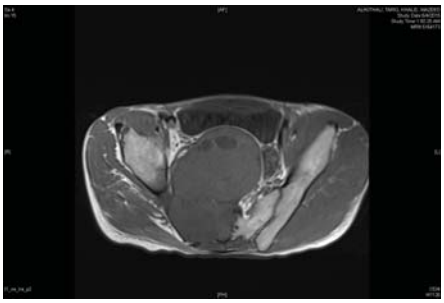

Figure-5: Coronal view of the tumor

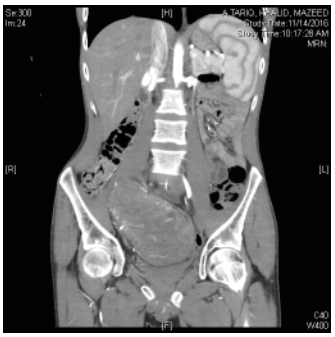

Figure-6: another view of CT scan showing huge pelvic schawanoma

gery team, and interventional radiology team and radiation.

Oncology: As a result of conclusion of the meeting we decided to excision of tumor with combine team efforts and aim of management is complete resection of tumor, spine fixation, excision from intra, extradural and nerve root. This is the challenging case with possibility of high risk of complication like urine, fecal incontinence, motor and sensory deficit, bowel injury etc. Tumor is high vascular and there is high chances of per operative bleeding so we decided to do angio-embolisation prior to surgery by interventional radiologist. Angio-embolisation of right internal illac artery which is main branch of tumor was carried out with side branch of left internal illac artery.

The surgery was done under neurophysiologic monitoring

Step-1 midline laparotomy (done by surgical oncology team). Mid line laparotomy was done. Identification of ureter done and we save it. Then ligation of internal illac artery. Excision of tumor (piece meal excision of tumor (near total) up to illac bone into pre sacral area. Then posterior approach through right buttock incision and we clear out all tumor. All these excision done under neurophysiologic monitoring.

Step-2 (dorsal mil line incision from L4 to S2 done by spinal surgery). Excision of Sacral and iliac bone followed by fixation

Step-3 (microscopic review and excision of tumor from nerve roots and L5 S1 laminectomy (done by neurosurgery team)

1. Resection of affected part of sacrum
2. Duratomy to ressect intradural tumor (hnbno tumor)
3. Extradural tumor resection in step wise under neurophysiologic monitoring
4. Decompression of sciatic nerve
5. Tumor arises from S1 nerve root in the right side which is non functional was excised.

The estimated time of the whole surgery is 15hrs. The outcome of the patient is very good with multi-disciplinary team effort without neurological deficit.

Finally the patient was referred to radiation oncology for radiotherapy.

The purpose of report this case is to develop awareness among the physician that multi-disciplinary team effect is required to manage this kind of challengeable case.

Discussion:
A retroperitoneal pelvisacral schawonoma is rare slow growing tumor and presents delay symptom. The common symptom of this cases is pain and weakness in the lower limbs.[4] The tumor is benign in nature but when it appear with von Recklinghausen's disease it can be malignant.[7]

The percentage of pelvic schawonoma is 1-3 %.[6] This case showed that patient having the pain in the lower limbs radiates from back due to rediculopath of one or more sacral nerve plexuses.

The radiological imaging technique like CT scan abdomen and pelvis and MRI pelvis play decisive role for the diagnosis regarding the anatomical original, size, extent of tumor relation with pelvic organ and feeding arterial supply to the tumor.[4] Typically schawonoma is well characterize tumor having solid, cystic component with some area of necrosis with vascularity.

A pre-operative iliac artery angiography and relevant artery embolization may helpful to reduce the blood lose and prevent uncontrolled hemorrhage which may necessitate the premature termination of the operation.

The surgical management is depend upon the anatomy of the tumor like intra sacral and retroperitoneal extension of the tumor. The anterior trans abdominal approach is good for the tumor present in the presacral area to gain the vascular control of the tumor and protection of intrapelvic organ. And posterior approach should be used in cases of tumor involved spinal canal and intrasacrum.

Reconstruction of bony structures should be considered and pre-operatively planned depending on the degree of sacral destruction and sacroiliac joint involvement. CT can be used to accurately detect the bony destruction caused by the tumor and should be part of the preoperative evaluation in all patients.

Multi-disciplinary teams including oncology surgeon, neurosurgeon anorectal surgeons, vascular surgeons, spine surgeons and anesthetists should be available since complications might be anticipated. It is of utmost importance to avoid injury to the adjacent visceral, vascular, and nervous structures, which may result in complications like hemorrhage and neurological deficits.

Local recurrence and malignant transformation are extremely rare, so a piecemeal subtotal excision or simple nucleation should be the treatment of choice.[7]

We did piecemeal subtotal excision of tumor under neurophysiologic monitoring.

Conclusion:
Pelvic schawonoma is the rare tumor, need proper evaluation with CT scan, MRI and histopathology and successful treatment is not possible without multidisciplinary team.

The complete treatment includes excision of tumor, fixation of sacrum and removal of tumor from microscopic nerve root under neurophyiological monitoring.

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Role and contribution of authors:
Dr Arshadullah Khan, conceive the idea and collected the data and references and did the initial write-up

Dr Jehad Al Shawi, collected the data and references, and helped in discussion writing.

Dr Sumayya Tajwar, collected the data and references and helped in introduction and discussion writing.

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