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Giant sacral schwannoma mimicking teratoma. A case report

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ABSTRACT

Background: Sacral schwannomas are uncommon, constituting 1-5% of schwannomas. They present with vague symptoms. Schwannomas occurring in sacral and presacral regions present with enormous dimensions and are difficult to manage.

Case summary: We present a case of schwannoma occurring in the sacral region extending into the presacral region in a 45-year-old female which was diagnosed as teratoma on imageology. The lesion was excised and on histopathological examination, diagnosis of schwannoma was considered

Conclusion: Sacral schwannoma with cystic degeneration should be considered as a differential diagnosis for solid and cystic lesions of the sacral region. MRI and CT scanning will be helpful in diagnosis.

Keywords
schwannoma,
sacral,
spinal

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INTRODUCTION

Schwannomas are benign tumours originating from schwann cells. These tumours are more common in head and neck, posterior mediastinum and extremities. sacral intraspinal schwannomas are relatively infrequent and accounts for 1-5% of spinal tumours. [1] Schwannomas in spinal cord often results in extensive bony destruction and are detected when they attain enormous dimensions and are termed as Giant schwannoma. 3 types of sacral schwannomas have been described based on anatomical findings i.e. retroperitoneal schwannoma, intra spinal schwannoma and spinal schwannoma. Clinical features vary depending upon type of sacral schwannoma. Retroperitoneal schwannoma has slow growth and presents with non-specific symptoms. [2] Intraspinal schwannoma is an intraosseous lesion presenting with mild low back pain. Presenting with neurological deficit is unusual. [3] spinal schwannoma rise from spinal nerve root and present as dumb-bell tumour. Dumb-bell schwannomas occurring in sacral region comprise 4% of all spinal schwannomas. [4] We are presenting a rare case of giant sacral schwannoma arising from spinal cord mimicking sacral teratoma on imageology.

Case report

A 45 years old female came with chief complaints of pain in the right parasacral region since one month. Pain is radiating to right leg, aggravated on prolonged standing. Patient has history of irregular menstrual cycles from 2 months and dyspareunia. There is no history of abdominal pain, vomiting, fever and melena. Bowel and bladder movements are normal. No history of loss of appetite. Her obstetric history was P4L4.

On local examination (per rectum) induration is felt at 10'0 clock position.

Systemic examination: Per abdomen examination revealed ill-defined palpable mass in the pelvic region with variable consistency. Mild tenderness is present. Bowel sounds are normal.

Tone, bulk and power of both lower limbs are normal. Haematological parameters are with in normal limits (Haemoglobin: 13.1g%, RBC count 4.48 million/microliter, Total count – 6500/ microliter, ESR-16mm/hour, PCV- 39%, MCV – 87fl, MCH 27pg, Platelet count – 3.15 lakhs/microliter). Serum Urea – 13mg/dl, serum creatinine – 0.5 micromoles/L, Serum electrolytes like serum sodium – 142 mmol/L, Serum potassium – 4.1mmol/L, Serum chloride – 106mmol/L, Serum chloride 106mmol/L. random plasma glucose is 101mg/dL.

Contrast enhancing computed tomography (CECT) revealed large lobular sacral lesion with cortical destruction and presacral soft tissue component measuring 8.2X8.5X7.2cms suggesting teratoma.

Magnetic resonance imaging of pelvis shows well defined altered signal intensity lesion of size 8.5X7.1X7ms which is heterogeneously hyper intense on T2, hypo and isointense on T1 showing areas of restricted diffusion on DW1 and heterogenous enhancement post contrast with few cystic areas with in are noted (Figure 1). Lesion is arising from spinal cord at S2-S3 vertebral body region causing expansion of spinal canal at that region, involving S2 vertebral body extending into right neural foramina at S2-S3 level with extension into right presacral region. Lesion is causing displacement of uterus and rectum anterolateral with maintained fat planes. Clinical diagnosis of sacral teratoma is considered.

Excision of retroperitoneal tumor was done which is 8x8cms well encapsulated tumor at the level of S2-S3 with pedicle extending into right sacral foramina.

Grossly we received encapsulated soft tissue mass measuring 7X6X4.5cms. Cut section of lesion is lobulated, grey white with focal cystic and calcified areas (Figure 2).

Microscopically, capsulated lesion was composed of hyper and hypocellular areas with spindle shaped cells. Few foci shows cells showing palisading with verocay bodies (Antoni A areas) (Figure 3). Many foci showed cystic spaces with adjacent hyalinised tissue (Figure 4). Foci of calcification are also noted (Figure 5). Congested blood vessels with hyalinised walls are noted. On immunohistochemistry, spindle shaped cells were positive with S-100. These histopathological features are consistent with schwannoma (Figure 6).

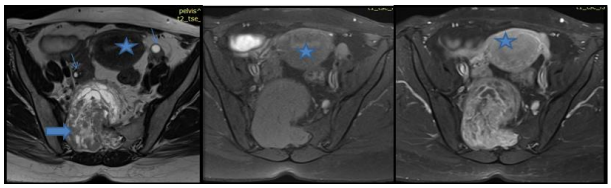

Figure 1. MRI pelvis - T2W, T1W and post contrast T1W axial images showing large heterogeneously enhancing lesion (thick arrow) in the spinal canal extending into presacral region through widened right neural foramina suggestive of neurogenic tumour. normal appearing uterus and ovaries (thin arrow) are seen separately

Discussion

Spinal schwannomas are more common in thoracic or lumbar region when compared to sacral region. [5] Retroperitoneal schwannomas are encapsulated tumors, often located in presacral and Para spinal areas. Pathogenesis of schwannoma is not clear but their occurrence may be associated with gene mutation. [6]

Peripheral nerve schwannoma arise from Schwann cells, involving one or two fascicles and displacing the other fascicles of involved nerve. They occur within endoneurium. Perineurium and fibrous epineurium surrounds the tumor and encapsulates it. [7]

Sacral schwannomas have low growth rate and have the ability to conceal due to the large pelvic space which leads to delayed diagnosis. Retroperitoneal schwannoma have mean growth rate of 1.9mm/year. Majority of these tumors are

solitary (96%) and only 4% are plexiform. [8] According to anatomic location, sacral schwannomas are divided into retroperitoneal, intraosseous, and dumbbell type. Retroperitoneal schwannomas are located in posterior peritoneum, outside the spinal cord. Intra osseous type are located in and invade spinal canal. Dumbbell type schwannoma extends outside the spinal cord through intervertebral foramen. [9]

Sacral schwannoma is asymptomatic when it is smaller in size but as the size increases patient presents with radicular pain to legs and reaches an enormous size at the time of presentation. Definite criteria for the term giant schwannoma is not defined but commonly accepted criteria is tumor extending into more than two vertebral bodies, extending into adjacent myofascial planes and extra spinal extension should be more than 2.5cms. [10]

Degenerative changes such as cyst formation can occur either because of degenerative changes in Antoni B areas or due to ischemic necrosis. Other degenerative changes are necrosis, fibrosis, haemorrhage, calcification and heterogenous cellularity which gives heterogenous signal intensities of contrast enhanced T 1 weighted images and T 2 weighted images. MRI features of typical schwannoma and giant sacral schwannoma are similar except when there is degenerative changes in giant sacral schwannoma. [11]

Patients with sacral schwannoma present with various clinical symptoms like nerve tissue or pelvic organ compression, sciatica, lower back pain, bone destruction, bladder and rectum compression and weakness in lower limbs. [12]

Common neoplastic lesions which have sacral origin are malignant tumors like chondrosarcoma, chordomas and metastatic lesion, while common benign tumors having sacral origin are giant cell tumor, osteoblastoma, aneurysmal bone cyst where as schwannoma are very rare. Schwannomas with degenerative changes can mimic other tumors in imageology as was in our case which was clinically diagnosed as teratoma. However histopathologically it is easy to differentiate the tumor from other entities

Complete resection of tumor is the treatment of choice to prevent the recurrence. However in giant invasive sacral schwannoma complete resection is difficult due to invasive growth pattern, hence piecemeal subtotal excision is choice and can achieve good outcome. Malignant transformation and local recurrence are extremely rare.

Conclusion

Sacral schwannoma are rare benign tumors and their clinical diagnosis is frequently delayed, as the symptoms are produced only when it reaches huge size. Sacral schwannoma with cystic degeneration should be considered as differential diagnosis for solid and cystic lesions of sacral region. MRI and CT scanning will be helpful in diagnosis. Specifically, CT scanning demonstrates bony destruction and helps for preoperative planning. Complete resection is treatment of choice but piecemeal subtotal excision in tumors with invasive growth pattern also achieves good outcome.

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