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Case Report 5

Hemangiopericytoma of lumbar spine: A mystery behind low back pain

Case history

A 45 year old previously healthy lady presented with mechanical type of low back pain of 4 months duration. She was treated by a general practitioner with analgesics and a course of physiotherapy but did not help with the pain. The onset was gradual and slowly progressing pain dull in nature. There was no radiation to the lower limbs but recently she noticed numbness and weakness of both legs two weeks prior to the admission. She denied recent falls or trauma to the back, she had no issues passing urine and no changes in the bowel habit. With time her symptoms affected badly and she had to depend on walking aids for mobility. There were no features suggestive of an infective process as well as no symptoms related to any primary malignancy such as lung, thyroid, breast, bowel and genitourinary.

On physical examination, her gait pattern was affected due to the back pain. There was midline and tenderness over the lumbar spine with limited spinal movements. Decreased sensation over bilateral lower limbs from mid thigh downwards was noted. Further neurological assessment revealed bilateral paraparesis of medical research council (MRC) power 3/5. There was no saddle anaesthesia and anal sphincter tone was preserved. Bilateral knee and ankle reflexes were weak and planter reflex was equivocal. Laboratory investigations

didn't reveal any significant alteration. Erythrocyte sediment rate (ESR), C-reactive protein (CRP), and tumour markers were within normal range. Chest radiograph was unremarkable.

Plain X ray of lumbo sacral spine anteroposterior and lateral views were obtained and it showed significant height loss of L2 vertebra with lytic lesion within the body (Figure 1). Disc space was maintained and grade I spondylolisthesis was also reported at L5-S1 level. Magnetic resonance imaging (MRI) was then requested and it showed an extra medullary enhancing lesion extending anteriorly and posteriorly causing significant cauda equina compression with severely collapsed L 2 vertebral body (Figure 2). Computerized tomography guided percutaneous biopsy was performed and the histological analysis showed a very vascular spindle cell tumour with slit like and staghorn vascular spaces, suggestive of haemangiopericytoma.

A decision was obtained to operate the patient in view of severe cord compression, progressive neurological deficit and to obtain a tissue sample for confirmation of the diagnosis. Considering vascular control during surgery a preoperative angiogram was performed and it showed the tumour receives blood supply from both L2 lumbar segmental arteries and angio embolisation was performed two days prior to the surgery.

Surgery was performed through anterolateral retro peritoneal approach with the patient in right lateral position. L2 vertebral body was identified. Segmental arteries were ligated. L2 corpectomy was performed (Figure 3,4) and the reddish fleshy mass of tumour was removed.



Figure 1

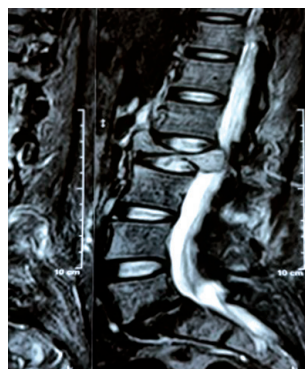


Figure 2

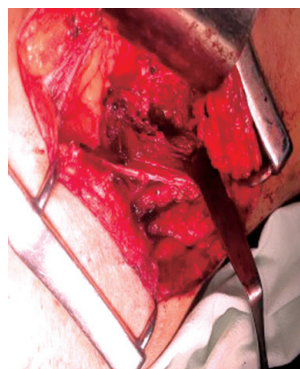


Figure 3

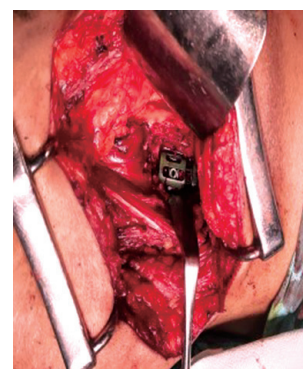


Figure 4

Discussion

Haemangiopericytoma is a rare vascular tumour originates from Zimmerman's pericytes which are contractile spindle cells lining the capillaries and post capillary venules. This was first described by Stout and Murray in 1942. It accounts for less than 1% of all vascular tumours. It exhibits both benign (80%) and malignant forms (20%). It may arise from any part of the body where capillaries are present, but most common sites are lower extremities, retro peritoneal region followed by head and neck region. Spinal haemangiopericytoma is extremely rare and only 80 cases have been reported worldwide. There is no sex preference but a slight predominance in males is reported. The soft tissue form commonly occurs in fifth and sixth decades whereas the osseous form commonly arises in fourth and fifth decade.

The tumour can be locally aggressive and it has a very high potential for local recurrence (80%). The malignant form is highly aggressive and preferably metastasizes to lung and bone (23%). Histologically it is characterized by dense blunt spindle cell proliferation with a richly vascular stroma. The capillary vessel proliferation

Spinal stability was achieved with expandable lumbar cage and lateral plating. Post operatively significant pain improvement neurological recovery was observed. Histology report confirmed the diagnosis of haemangiopericytoma without evidence of malignant potential.

often acquires a staghorn configuration. Each pericyte is characteristically surrounded by rich reticulin network. The histological feature that favours malignant potential includes mitotic count more than 4 per high power field, nuclear atypia and necrosis.

Clinical manifestations may vary depending on the size of the tumour and the location. Pain may not be reported in soft tissue form whereas pain is the first symptom commonly experienced in osseous type of tumour. Neurological deficit may result from vertebral tumours causing cord compression or as a result of pathological fracture.

There are no specific radiographic features of these tumours. However, characteristic signs that may help in the diagnosis. A "spider shaped" appearance in the arterial phase and dense well demarcated round or oval tumour staining in the venous phase are the characteristic features.

Surgical resection remains the first choice of treatment for all type of tumours when feasible. Spinal tumours should be resected as en bloc so as to relief neural compression. However, radical resection is often impossible without causing significant neurological deficit. Pre operative

angio embolisation is an efficient strategy to minimize intra operative bleeding. Angiographic studies help in identifying the feeder vessels. Complete resection of benign tumours is sufficient whereas in malignant cases additional radiotherapy or chemotherapy may be considered especially in high grade tumours, large tumours and resection with positive margins. Radiotherapy alone is indicated in unresectable tumours, similarly chemotherapy alone is indicated in unresectable tumours and metastasis. However, the effectiveness of adjuvant chemotherapy and radiotherapy is still uncertain. The prognosis of the tumour largely depends on resectability and the histologic grading. Usually the benign tumours have a good outcome but in contrast malignant counterparts have a high rate of local recurrence and propensity to metastasize.

Conclusion

Haemangiopericytoma is an extremely rare vascular tumour which can mimic a destructive metastatic lesion in the spinal column. Therefore the primary goal in the management of a lytic lesion in patients over 40 years, is to exclude secondary metastatic deposits from common primary sites. Haemangiopericytoma of spinal column should be considered potentially malignant and therefore be treated promptly and aggressively. Surgical resection is always the mainstay of treatment and should be attempted whenever feasible. The role of radio therapy and chemotherapy is still uncertain and therefore may be considered in high risk patients. Both benign and malignant tumours should be followed up for long term for the early detection of recurrence.

References

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