DESMOID-TYPE FIBROMATOSIS OF C6 VERTEBRAL BODY: RARE AGRESSIVE TUMOR INVADING CERVICAL VERTEBRA

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INTRODUCTION:

Desmoid-type fibromatosis is a benign, slow growing fibroblastic tumour but locally aggressive. It clinical course is unpredictable with high local recurrence rate. Spine and bony involvement is unusual. We report a case of severe cervical myelopathy secondary to desmoid fibromatosis treated surgically.

REPORT:

64 years old man having progressive worsening cervical myelopathic symptoms for 7 years. He became paraplegic with contracture deformity of bilateral lower limb 2 years prior to visit to clinic. Refusal for medical treatment is the reason of late presentation to hospital. Previously no history of neck trauma or surgery.

Patient was cachexic with bilateral hip adduction and knee flexion contracture deformity disabling us from assessing lower limbs power. Neurological level at presentation was at T1. Modified JOA (mJOA) score was 7.

Radiographs showed pathological fracture of C6. MRI revealed rim lesion with loculation at C6 vertebral body, retropulsion and compression of spinal cord causing demyelination (Figure 1).

ACCF of C6, reconstruction with tricortical iliac bone grafts and mesh cage and percutaneous bilateral hip and hamstring release was done. Intraoperatively noted C6 vertebral body destruction and whitish soft tissue mass anterior to dural with no dural adhesions. The tumour was removed intralesional.

HPE shows cellular monotonous fibroblastic cell proliferation in keloid-like collagenous background suggestive of desmoid type fibromatosis. Patient was started on early rehabilitation post operatively. One year later, patient functional level improved (mJOA score 9) and the lower limb contractures deformity resolved. Bilateral lower limb power improved to grade 4 and he able to stand semi-independently.

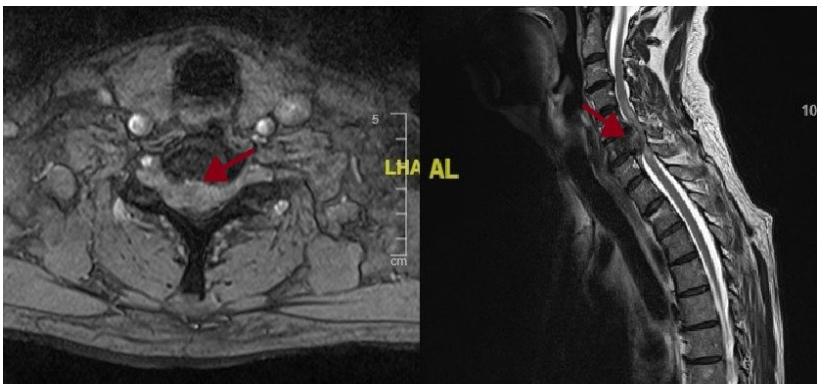

Figure 1: Arrow showing lesion severely compressing spinal cord

CONCLUSION:

Sporadic desmoid fibromatosis following spine surgery or resection of schwannomas has been reported. However, our patient has no prior history of trauma or surgery. The disease may progress slowly over years. Surgery is indicated for symptomatic patient with good outcome. Further follow up is essential to detect tumour recurrence.

REFERENCES:

1. Kim, So Jung, et al. "Desmoid type fibromatosis in the facet joint of lumbar spine: Case report and review of literature." *Korean Journal of Radiology* 14.5 (2013): 818-822.