A case of an invasive schwannoma developing at gibbus due to an old tuberculous spine

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Summary
The invasive schwannoma in the spine is a rare clinical entity. There have been no reports of schwannoma arising from tuberculous spine. The authors report on a case in which invasive cystic schwannoma arose at old tuberculous spine and paraplegia progressed by the tumor. The tumor excision and resection of the sequestrum achieved satisfying neurological improvement. Although development of cystic tumor in the tuberculous spine may be extremely rare, it should be considered in the differential diagnosis, especially when systemic symptoms of tuberculosis were absent.

Introduction
An invasive schwannoma of the spine is an extremely rare clinical entity and there are no reports of schwannoma arising from a tuberculous spine. This is a report describing a rare case of a giant cystic schwannoma arising at the site of gibbus due to previous tuberculosis.

I. A case report
A 75-year-old woman presented with a history of spinal tuberculosis, but no evidence of active disease for 42 years. Her old medical records were not available. According to her memory, she had left sciatica from the age of 24. Spinal tuberculosis was diagnosed at another hospital when she was 25 years old. Thereafter, through several years of bed-rest and drug treatment (streptomycin), a kyphosis gradually developed in the lumbar spine. At 32 years, a left iliopsoas abscess spontaneously drained and required surgical drainage several times. She had also undergone a posterior lumbar spinal fusion at the age of 33. The post-surgical
course was unremarkable and, thereafter, she was not hospitalized further. She came to our institution because of a 6-month history of increasing lower limb weakness and bladder dysfunction. About one year prior to being seen at our hospital, she suffered back pain at mid-lumbar level. Three months later, she noticed muscle weakness on her left leg, which developed gradually and spread to both legs. Six months later she could not walk and had received an indwelling catheter.

On physical examination, she had an obvious gibbus at mid lumbar level. Neurological examination disclosed a flaccid paralysis below the iliopsoas. Muscle power was graded at fair on her right leg and between zero and poor on her left leg. Rectal sphincter tone was diminished. She suffered sensory disturbances below the L1 dermatome in both legs and analgesia below L3 on the left leg. Bilaterally, knee and ankle reflexes were absent. Symptoms were more marked on the left side. She did not suffer systemic symptoms of tuberculosis such as fever and weight loss. Laboratory reports were unremarkable. Her white blood cell count was 5,700, with a normal differential count. Erythrocyte sedimentation rate was 18 / hour. Purified protein derivative test (Mantoux) was 15mm in diameter at 48 hours. Sputum, gastric washing and urine culture for tuberculosis were negative. C-reactive protein was 0.52 mg/dL. Cerebrospinal fluid was xanthochromic with a positive Pandy, Nonne-Apelt and tryptophan test.

Chest radiography revealed no evidence of tuberculosis. Lumbar lateral radiograms revealed a diagnosis of severe osteoporosis
and a 56 degree kyphosis at the L2-3 level with fragmented, localized sclerotic and destructive bone lesions (not shown). Magnetic resonance images showed that the L2-3 vertebra was completely fused and detected a large cystic lesion occupying the spinal canal at L2 and eroding and extending into the L2 vertebral body, which had low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. There was rim enhancement after an intravenous gadolinium-diethylene triamine penta-acetic acid injection (Figure 1). Computer tomography showed that the cystic lesion had eroded into the surrounding bony tissues with remaining sequestra in the lesion (Figure 2). Myelography failed because injection of small amounts of contrast medium induced unbearable sciatic pain.

We were uncertain if the lesion was a tuberculous abscess or cystic tumor, however, decided to perform decompressive surgery because of the progressive muscle weakness. Surgery was performed through a posterior approach. The previous surgical wound was reopened and a very thin fused lamina was resected from L1 to L4. The bone removal was extended laterally to include the L1/2 and L2/3 facet joints to facilitate resection. The dural tube was widened to double in width, thin and attenuated and was opened from L2 to L4. Two different cysts were identified; one was located in the dural tube and the other was in epidural space. The cyst in the dural tube was ruptured. A grayish tumor was mingled with the cauda equina and was removed piece-meal. It was not possible to perform radical excision because the tumor lacked a capsule that would help in dissecting it from the surrounding structures. The origin of the tumor was assumed to be from the cauda equina. It was found that the anterior dural tube had been pushed posteriorly by the an-
gular deformity and bony sequestrum at the L2-3 vertebral bodies. Next the anterior dural tube was cut longitudinally and sequestra were resected. After the tumor resection, the thin dural tube was sutured without any problems. The tumorous tissue and sequestra obtained at surgery were submitted for microbiological and histopathological studies. Histology of the tumor showed typical appearance of schwannoma without inflammatory cell proliferation (Figure 3), and no tumor cells in the sequestra. The results of the bacterial stains and culture studies were negative for acid-fast and any other bacilli.

After surgery, bed-rest for 2 weeks was ordered after which mobilization was allowed after application of an external brace. Postsurgical course was satisfactory. Lower extremity strength and sensory disturbance was recovered, especially on the right leg. Although urinary disturbances were not recovered, ambulatory disturbance improved gradually. Two years after surgery, the patient could walk with a cane and left orthosis.

II. Discussion

Cauda equina nerve sheath tumors are usually single, small, well encapsulated tumors that are relatively simple to remove and are associated with a good surgical outcome. On the other hand, giant invasive schwannomas in the spine are encountered uncommonly and differ from the other schwannomas in that they may lack a well-defined capsule and may extend into the adjacent tissue without having undergone a malignant change histologically. In the case of our patient, the multi-lobulated lesion eroded anteriorly into the vertebral bodies and posteriorly, thinning and attenuating the dura and the lamina.

MR findings in tuberculous patients generally are more typical of neoplasm than of infection and are sometimes difficult to differentiate from soft tissue tumor. Low signal intensity on T1-weighted images and high signal intensity on T2-weighted images are commonly seen in tuberculous abscesses. Marginal enhancement on post-contrast MR images and post-contrast CT images are typical findings in tuberculous abscesses, and are sometimes seen in other abscesses and in tumors. Gupta et al. emphasized the presence of bone fragments is specific for tuberculosis. In our patient, because of enhancement of the tumor wall in contrast enhanced MRI and the presence of bone fragments, it was very difficult to preoperatively diagnose intradural schwannoma.

To our knowledge, there are only two case reports dealing with an association between tuberculosis and a tumorous lesion. One is a case of chiasmal osteoma and the other is a large calcified mass occupying the same location after tuberculous meningitis. There is no report of neural tumor after tuberculosis. Therefore, the exact mechanism of tumorigenesis in the present case is unknown. However, schwannoma formation
may be a sequela of longstanding chronic inflammation.

Because complete removal had the risk of sacrificing many nerve roots, controversy still exist whether these slow-growing and histologically benign but neurologically devastating tumors should be aggressively resected or not\(^1,6\). Considering her age and the risk of recurrence is supposed to be low, the need for adequate tumor removal was balanced against preservation of nerve function. The tumor excision led to improvement in the patient’s neurological functions. Our patient, however, requires life-long follow up because total excision was not possible.

Although development of this entity in the tuberculous spine may be extremely uncommon, it should be considered in the differential diagnosis, especially when systemic symptoms of tuberculosis such as fever and weight loss are absent and laboratory studies are unremarkable.

References

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