Clinical symptoms are usually those of the thoracic component, which may include radicular pain in the territory of the involved intercostal nerve. The pointers to intraspinal extension and presence may be much more dramatic. These symptoms and signs are those of thoracic spinal cord compression with paraparesis or paraplegia, spasticity, hypoaesthesiae and sensory loss and sometimes hyperaesthesiae. There may be loss or impairment of urethral and anal sphincter function along with increased deep tendon reflexes and up going plantar responses. If the tumour is unilateral there may be a partial or complete Brown Sequard syndrome. As the pressure is usually quite slowly progressive, there is either a gradual spastic paraparesis or the patient may be asymptomatic inspite of quite extensive cord deformation and compression[3]. This is borne out by case 1 in our report. The reason may be the gradual compression of the cord with accommodation of the local vascular supply of the neural tissue.

Treatment of these tumours is surgical with either thoracotomy with excision of the posterior mediastinal mass and evacuation of as much tumour from the involved intervertebral foramen is performed. This is followed immediately by laminectomy and excision of residual intraspinal tumour[4]. However, a second approach is advocated by Findlay, who recommends initial excision of the spinal component of the tumour so that subsequent manipulations of the tumour will not be transmitted to the cord[5]. This is the approach preferred by us in case 2.

Spontaneous regression of these tumours has not been described previously, although it is well known in other tumours of neurectodermal origin such as neuroblastomas and mesodermal tumours, origin such as metastases of renal cell carcinoma.

In both our cases there is documented regression of tumour in the intraspinal compartment. This had been proven radiologically in case 1 (Figure 2) although there was recurrence of the intrathoracic component. This can be inferred symptomatically in case 2, where the patient became normal neurologically (both subjectively and objectively) inspite of treatment which can at best be described as placebo. The extent of cord compression found at surgery was not adequate in case 2 to be responsible for her severe neurologic symptoms two years prior to surgery. However it was quite compatible with her present neurologic state.

One can only presume that in both cases there was a vascular event leading to complete disappearance of intraspinal tumour in case 1 and reduction in size in case 2. Such tumour necrosis is well known. Evidence for this may be the presence of hyaline change seen in the tumour of case 2.

Regression of intraspinal extradural nerve sheath tumours has not been reported previously in the literature. Our discovery of two such cases emphasizes the need for vigilance on the part of clinicians dealing with these tumours so as to add to the knowledge of the behavnour of such neoplasms.

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Fig. 2.

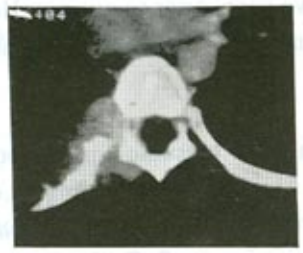

Fig. 2: Axial C.T at D5 with intravenous contrast enhancement showing no intraspinal tumour and intrathoracic recurrence (Case I).

Case II: A 36 year old right handed Asian female presented to a physician two years ago with the complaints of paraesthesiae in both feet, weakness in both legs and a bilateral foot drop. She had a complete loss to pinprick sensation upto the level of D5 and reduced kinaesthetic and touch sensation to the same sensory level. She had bilaterally increased tone and deep tendon reflexes, absent abdominal reflexes and bilaterally upgoing plantar responses. She was advised further investigation and informed about the probabiity of spinal surgery. However she opted to have medication mainly vitamin preparations. However she gradually improved over the next six months starting to have first increase in power in the lower limbs and then the sensation improved to normal. Now two years later she was seen in a general surgical clinic for a right sided intrathoracic mass detected on a chest x-ray performed for intractable cough. She complained of occasional paraesthesiae in her lower limbs. On examination she was seen to be grossly overweight but her neurological examination was entirely normal. A C.T. scan of the chest and thoracic spine was performed revealing a dumbbell tumour with an intraspinal component at D3 with enlargement of the intervertebral foramen and erosion of the body (Figure 3). During subsequent D3 and D4 laminectomy the lamina of D3 was found to be deformed posteriorly by underlying purely extradural tumour. The thecal sac had only minimal compression and was freely pulsatile distally (Figure 4).

Fig. 3.

Fig. 3: Axial C.T at D3 showing intraspinal lesion (Case II).

Fig. 4.

Fig. 4: Thecal sac following tumour removal (small arrow) and tumour bed (Large arrow) (Case II).

The intraspinal tumour was excised, the intrathoracic portion being left to be dealt with at a later stage. Frozen and paraffin section showed this tumour to be a schwannoma with Antoni A and B areas. There was moderate hypercellularity but no mitotic areas were seen. Some hyaline change was also seen.

Discussion

Dumbbell neoplasms of the spine arising from nervous tissue may be of two types. One is the intra and extradural variety. These are mainly neurofibromas, which arise from dorsal nerve roots, although some examples originating from the motor roots are also known[1]. The second form affects the intercostal nerves in the posterior mediastinum. These are mainly schwannomas and extend to the intraspinal compartment by enlargement of the corresponding intervertebral foramen (53% of cases described by Ringertz & Lidholm[2]).

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SPONTANEOUS INTRASPINAL REGRESSION OF DUMBBELL NERVE SHEATH TUMOURS

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Abstract

We report two cases of posterior mediastinal benign nerve sheath tumours of the dumbbell type. One of these had intraspinal regression following posterior mediastinal surgery. There was symptomatic regression in the second. Vascular events may have been the cause of this decrease in size of intraspinal tumour in both cases.

Key Words: Dumbbell tumour, extradural compression, hourglass tumour, intraspinal tumour, spontaneous regression

Case Reports

Case I: A thirty five year old Asian female presented to the thoracic medicine department at our institution with the complaints of right sided chest pain for seven years. This was initially mild and intermittent and had now become severe and constant. Chest X-Rays revealed a right sided intrathoracic and posterior mediastinal mass. A chest C.T. scan showed a 12 x 8.0 x 8.0 cm tumour lying in the right chest cavity and posterior mediastinum, with an intraspinal component, severe cord distortion, and enlargement of the intervertebral foramen at D5 (Figure 1). A diagnosis of dumbbell neurofibroma was made. She underwent a right posterolateral thoracotomy through the fifth rib at another facility. Complete excision of the intrathoracic tumour was performed. No attempt was made to remove the intraspinal component and she was advised spinal surgery after recuperation. Histologically this excised tumour was seen to be a "simple" neurofibroma with no malignant features.

Fig. 1.

Fig. 1: Axial C.T at D5 with intravenous contrast enhancement showing cord compression (Case I)

She presented 9 months later to our neurosurgical clinic, not having undergone spinal surgery. She complained now of severe right sided chest wall pain and hyperaesthesiae in the thoracotomy scar. There were no other neurological signs especially those of myelopathy. A repeat X-Ray showed tumour recurrence within the thorax. A chest C.T. scan was then performed, with and without intravenous contrast. This confirmed on 8 x 7.5 cm tumour with erosion of adjacent vertebrae but no intraspinal component (Figure 2). This was further confirmed by a thoracic myelogram performed via the lumbar route. No indentation of the contrast column was found. She then had a right posterolateral throacotomy through the fourth interspace with excision of a fibrous well encapsulated tumour. At operation the only difficulty encountered was tumour adherence to the posterolateral chest wall. On histology this recurrent tumour was seen to be a benign myxoid neurofibroma. She had an uneventful postoperative recovery.

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