Case Report

Neurocytoma of the Thoracic Spinal Cord

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Summary

Central neurocytoma is an unusual tumour that arises in the supratentorial ventricular system of young adults. Similar lesions, termed simply neurocytoma, have been described at a variety of locations outside the ventricular system. Here, we report the case of a 50-year-old man who presented with pain and a rapidly progressive myelopathy due to a neurocytoma of the upper thoracic spinal cord. The literature on spinal neurocytoma and its relation to central neurocytoma are discussed.

Keywords: Spinal cord neoplasms; central neurocytoma; neurocytoma.

Introduction

In 1982, Hassoun et al. used the term ‘central neurocytoma’ to describe two cases of a distinctive intraventricular tumour [11]. Central neurocytoma (CN) is now a well-established diagnosis and is defined as a tumour composed of small round cells with neuronal differentiation that is typically located in the supratentorial ventricular system near the foramen of Monro [8]. CN comprise approximately 0.5% of all intracranial neoplasms, usually affect young adults and often present with symptoms of increased intracranial pressure [12]. The possibility of distinctive genetic abnormalities in CN has been raised [14, 29, 34].

Early experience suggested that CN were essentially low-grade [11, 19]. It is now known that a proportion of CN, particularly those with Ki-67 (MIB-1) labelling indices of more than 2%, have a less favourable prognosis in terms of time to recurrence or progression after surgery [1, 17, 23]. Craniospinal dissemination from otherwise typical CN has also been described [7]. Furthermore, primary tumours resembling CN have been found at sites outside the ventricular system including the cerebral hemispheres, thalamus, pons, cerebellum and spinal cord [1, 4, 6, 9, 16, 20–22, 24–28, 30]. These tumours are not ‘central’ and, although the terminology is disputed, many authors refer to them simply as ‘neurocytomas’ [8]. It is not yet clear how their clinical behaviour relates to that of classical CN. Here, we report a primary neurocytoma of the thoracic spinal cord and review the literature on this rare tumour.

Case Report

History

A 50-year-old man presented with three months of cervical, interscapular and right shoulder pain that was made worse by lying flat. Ten days before admission he developed progressive weakness of the left lower limb, poor urinary stream and constipation. His health was otherwise good and there was no significant family history.

Examination

There was a partial Brown-Séquard syndrome with ‘pyramidal’ weakness of the left lower limb (graded 3/5 overall) and a spinothalamic sensory level at T5 that
was more marked on the right. The cranial nerves, upper limbs and general examination were normal.

Radiology

Magnetic resonance imaging (MRI) showed an intrinsic tumour of the upper thoracic spinal cord and an associated syrinx. The tumour was isointense on both T1- and T2-weighted sequences, enhanced vividly and had an irregular margin (Fig. 1). The brain and lumbar sacral spine were normal.

Surgery

At operation the spinal cord was swollen and tense. The pial surface was intact but a midline dorsal myelotomy exposed a vascular red-brown tumour, which was internally debulked using an ultrasonic aspirator. Dorsally, there was a good plane of cleavage from the cord but ventrally and at the rostral margin significant bleeding was noted. Several adherent tumour fragments could not be excised but at closure the cord was fully decompressed and pulsatile. The blood pressure remained stable throughout the procedure. Spinal evoked-potential monitoring was not used.

Course

The post-operative examination revealed paralysis of both lower limbs and anaesthesia below the T4 dermatome. Unfortunately, the patient’s paraplegia did not improve and he was later transferred to a spinal injuries unit for rehabilitation. MRI performed two months after the operation showed persistence of the cervical syrinx, residual or recurrent cord tumour at the level of the T2/3 disc and atrophy of the distal spinal cord. In view of these appearances and the complete neurological deficit, the remaining tumour was removed and the rostral syrinx drained by spinal cord excision between the T2 and T6 vertebral levels. Two years after presentation the patient is fully independent in a wheelchair and has no evidence of recurrent disease on imaging.

Histopathology

The first specimen and subsequent residual tumour had similar features (Fig. 2). The tumour was composed of sheets of uniform small round cells and areas of delicate fibrillary neuropil. The cells had finely-speckled vesicular nuclei and clear perinuclear haloes. Their nucleoli were not prominent. Hyalinisation of small blood vessels was noted. No tumour astrocytes, ganglioid cells or rosettes were seen and there was no mitotic activity, necrosis, vascular endothelial hyperplasia or calcification. In the second specimen the tumour had a well-circumscribed non-encapsulated margin with the cord.

Immunohistochemistry for synaptophysin showed strong positive staining in fibrillar areas and in some cells. The cells were also positive for MAP-2 (microtubule-associated protein 2) and negative for Neurofilament-200kD and GFAP. The Ki-67 index was 2%. These findings suggested the diagnosis of neurocytoma. EM was not performed.

Discussion

The presence of a well-characterised tumour at a highly unusual location should always be questioned. For example, two recent reports have emphasised that oligodendrogial or glioneuronal tumours can be mislabelled as extraventricular neurocytoma if diagnostic criteria are not strictly applied [10, 31].

Neurocytomas have bland morphology and are composed of uniform round cells together with irregular fibrillary regions that resemble neuropil. The relative proportion of these two components varies from case to case [8, 12]. The diagnosis also requires evidence of neuronal differentiation, which is usually demonstrated on electron microscopy or by positive immunohistochemical staining for synaptophysin, a normal protein of pre-synaptic vesicles [8, 32]. This should label fibrillary areas of the tumour since focal cytoplasmic staining alone is not specific [31]. Other neuronal markers, such as MAP-2, may also be present. Our tumour conforms to this pattern and, in the absence of an intracranial lesion, is a primary neurocytoma of the spinal cord.

This case and five previously reported spinal neurocytomas (SN) are summarised in Table 1 [1, 4, 16, 24–27]. On MRI five were discrete, brightly-enhancing, intramedullary lesions. Two were associated with a syrinx and there was evidence of intratumoural haemorrhage in one. Therefore, the appearances of SN are similar to those of spinal cord ependymoma [13]. Curiously, one SN was an intradural extramedullary tumour of the cauda equina that resembled a schwannoma on imaging and at operation [25]. Three other