

Dumbbell-shaped cervical spinal neurilemmoma presenting as neck mass

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Abstract

Cervical neurilemmoma may originate from any nerve sheath tissue in the neck including the vagus nerve, glossopharyngeal nerve, brachial plexus, sympathetic trunk and cervical spine. We report an unusual case of a dumbbell-shaped neurilemmoma arising from the cervical spinal roots in a patient who complained of having had a neck mass for several months. Computed tomographic scan and magnetic resonance imaging revealed a dumbbell-shaped tumour extending from the C4 spinal level through the intervertebral foramen into the right parapharyngeal space. Decompression surgery was performed first via the cervical approach. Five months later, the patient received laminectomy and a complete tumour excision. The symptoms and signs were significantly relieved without neurological sequelae. No evidence of recurrence was noted after one-year follow up. This two-staged operation could offer an alternative surgical approach yielding ideal therapeutic results in such a rare disease.

Key words: Neurilemmoma; Spinal Neoplasms

Introduction

Neurilemmoma, also termed schwannoma and neurinoma, originates from Schwann cells located in the nerve sheath. It is usually a slow growing, well-encapsulated and benign tumour. About one third of neurilemmoma occur in the head and neck region arising from the nerve sheath of the facial nerve, sympathetic trunk, vagus nerve, glossopharyngeal nerve, brachial plexus, and cervical spinal roots.¹ However, dumbbell-shaped neurilemmoma of the cervical spinal root is relatively rare. We report such a case who presented, unusually, with a neck mass.

Case report

A 37-year-old man came to our clinic with the chief complaint of having had an enlarging neck mass on the right side for several months. The patient also complained of radiation pain to his right shoulder. Physical examination revealed a 3 × 2 × 2 cm mass in level II beneath the sternocleidomastoid muscle. It was firm, fixed, and mildly tender. The remainder of the otolaryngological examinations were unremarkable. Neurological examination disclosed numbness at the C3 and C4 dermatome of the right shoulder. Oblique plain film of the cervical spine showed enlarged intervertebral foramen between the C3 and C4 vertebrae. Computed tomographic (CT) scan and magnetic resonance imaging (MRI) showed a dumbbell-shaped, intradural and extramedullary tumour extending from the spinal canal at the C4 level through the intervertebral foramen to compress the medial-posterior edge of the carotid sheath, which resulted in the separation of the internal carotid artery and internal jugular vein (Figure 1). An exploratory operation was done first via the cervical approach. A well-encapsulated mass connected

with two nerve fascicles was found (Figure 2). For pathological examination, preservation of nerve function, and achievement of later complete excision, decompression surgery with intracapsular removal of tumour was performed. The pathological diagnosis was neurilemmoma, which was consistent with typical Antoni A areas (Figure 3). The previous symptoms and signs were partially relieved after the decompression surgery.

Five months later, excision of the intraspinal portion of the tumour was performed via a posterior approach with C3–C4 laminectomy. The previous symptoms and signs were completely relieved without any neurological sequelae. MRI was performed twice with an interval of five months after the second operation, and the results revealed only post-operative change. No evidence of recurrence was noted at one-year follow up.

Discussion

Between 25 and 45 per cent of extracranial neurilemmomas occur in the head and neck region and about one third in this region arise in the neck.² Neurilemmoma of the neck is therefore not an uncommon tumour and should be considered in the differential diagnosis of a persistent solitary mass in this area. The reported origins of cervical neurilemmoma are cranial nerves IX–XII, sympathetic chain, cervical plexus and brachial plexus. In comparison, neurilemmoma of cervical spinal roots, although well documented, belong to a relatively rare disease entity. The majority of these lesions are intradural and extramedullary.³

Most neurilemmomas of cervical spinal roots present with radicular pain, neck or shoulder numbness, motor weakness and other neurological symptoms.^{4–6} A

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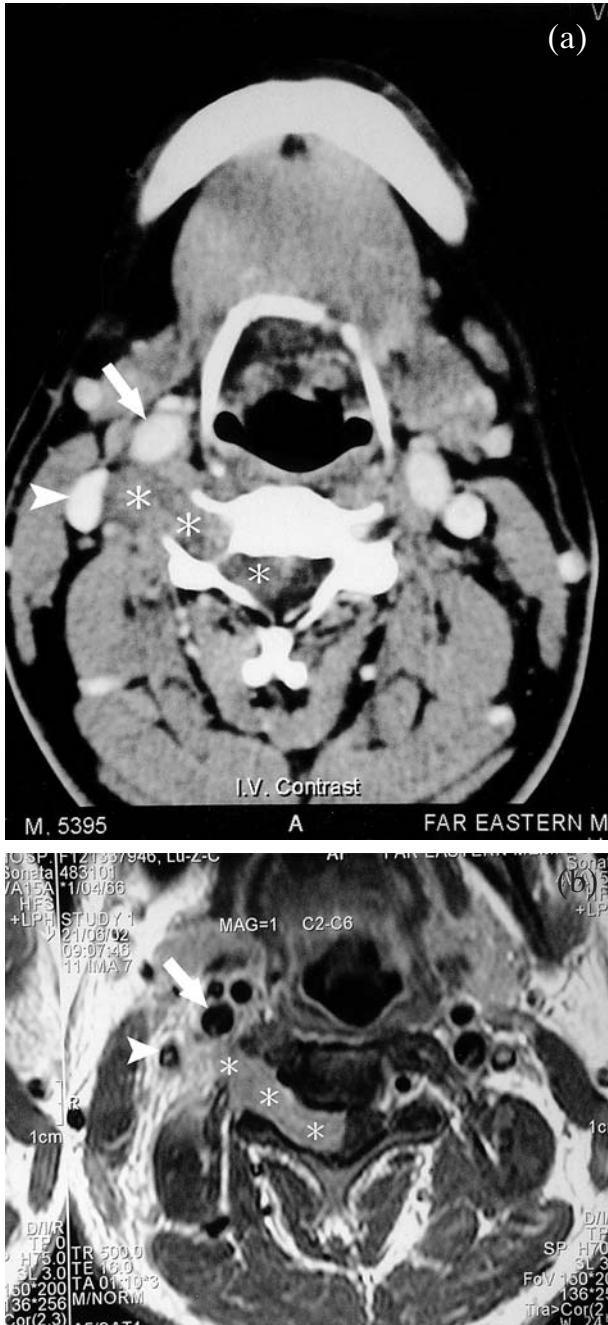


FIG. 1

(a) Computed tomographic scan imaging with contrast and (b) Magnetic resonance imaging T1WI with gadolinium shows a dumbbell-shaped tumour (asterisks) extending from the spinal roots through the intervertebral foramen which results in the separation of the internal carotid artery (arrow) and internal jugular vein (arrow head).

dumbbell-shaped form, one of the subtypes of spinal neurilemmoma, arising both inside and outside the spinal canal, usually presents similarly. There are only two reported cases in the Japanese literature of dumbbell cervical neurilemmomas with subcutaneous extension, from Asahi *et al.*⁷ To the best of our knowledge, our case is the very first case in the English literature of cervical dumbbell-shaped neurilemmoma presenting with a neck mass.

Although cervical neurilemmoma is not uncommon, those of a spinal origin should be carefully differentiated from others, due to the distinct management required. In

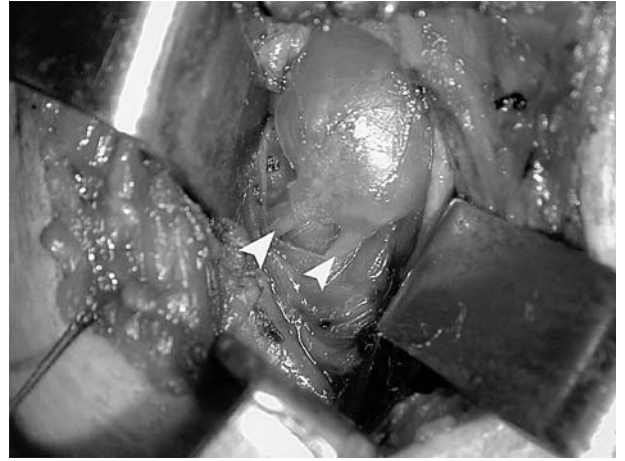


FIG. 2

The operative photograph shows a well-encapsulated tumour connected with two nerve fascicles (arrow heads).

our case, neck mass was the chief presenting sign. However, simultaneous neurological symptoms such as radiation pain to the shoulder combined with C3 and C4 dermatome numbness drew a suspicion of a neurologically associated tumour. Oblique plain film of the cervical spine showed enlarged intervertebrae foramen between the C3 and C4 vertebrae. CT scan then disclosed a roughly dumbbell-shaped tumour extending from the spinal canal through the intervertebrae foramen and compressing the adjacent structures. MRI further revealed an enhancing intradural and extramedullary tumour. Although aspiration biopsy has been recommended as a conservative pathology technique, a precise diagnosis is only made in 25 per cent of cases.⁸ Hence, initial exploratory surgery for tissue sampling is often necessary for accurate diagnosis and for planning further treatment.

If the lesion is proved to be a neurilemmoma, a clinically and histologically benign lesion, the advocated treatment for these tumours is complete surgical extirpation. With regard to neurilemmomas originating from outside the spinal canal, adequate surgical exposure via a trans cervical approach is the most appropriate way for tumour excision, as it allows easy control of the great vessels and a wide operation field allows the avoidance of injury to vital structures.^{9,10} The surgical approach for cervical dumbbell-shaped tumours originating from the spinal roots is more

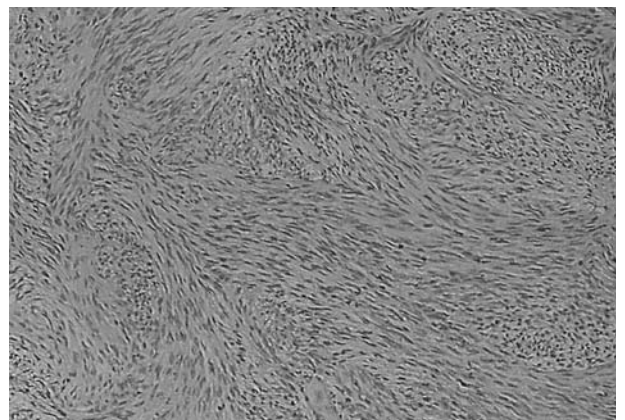


FIG. 3

Photomicrograph of the surgical specimen shows Antoni type A area with nuclear palisading characteristic of the neurilemmoma (haematoxylin and eosin stain, x100 original magnification).

challenging. Anterior or posterior cervical approach combined with vertebrectomy or laminectomy is traditionally enough for tumour excision.⁶ Moreover, a two-staged operation has been proposed by Love¹¹ and Habal *et al.*¹², with laminectomy as the stage I operation followed by further tumour removal. In our case, a two-staged operation was performed. But, unlike in the previously mentioned method, our first step was internal decompression surgery aimed at symptom relief, tissue sampling and reduction of tumour volume before the second-stage complete excision. Operative findings of nerve fascicles connected to the tumour provided the hint of neurilemmoma which was confirmed by the pathological examinations. This newly designed surgical planning provides a good alternative for the treatment of such a special tumour.

In conclusion, dumbbell-shaped neurilemmoma of the cervical spinal roots, although rare, can present as a neck mass. Careful history taking, physical examination and detailed imaging studies are essential for the diagnosis. A two-staged operation with an internal decompression followed by further tumour excision could be a good alternative surgical approach which may yield ideal therapeutic results.

- **This case report describes a spinal neurilemmoma presenting as a mass in the neck**
- **Computed tomography and magnetic resonance imaging revealed a dumbbell-shaped tumour**
- **Treatment was surgical in two stages. The possibility of spinal pathology presenting in the neck must be taken into account when assessing neck masses**

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