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Nerwiak osłonkowy (*schwannoma*) nerwu przeponowego: nietypowa przyczyna niebolesnego guza w obrębie szyi

Phrenic nerve schwannoma: an unusual cause of painless neck lump

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Streszczenie

Nerwiak osłonkowy (*schwannoma, neurilemmoma*) wywodzący się z nerwu przeponowego stanowi stosunkowo rzadki typ nowotworu. W pracy opisano przypadek nerwiaka osłonkowego nerwu przeponowego umiejscowionego w okolicy głowy i szyi u pacjenta, który zgłosił się z niebolesnym obrzękiem w obrębie szyi. Wyniki biopsji aspiracyjnej wskazywały na łagodny nowotwór wywodzący się z osłonki nerwowej. W celu potwierdzenia lokalizacji i typu guza wykonano badanie metodą tomografii komputerowej. Podjęto leczenie polegające na całkowitym wycięciu guza wraz z przyległymi włóknami nerwu. Po zabiegu u pacjenta wystąpiło porażenie prawostronne przepony, które leczono zachowawczo. Pooperacyjne badanie immunohistochemiczne pozwoliło na potwierdzenie rozpoznania nerwiaka osłonkowego komórkowego (*schwannoma*). Przedstawiony przypadek obrazuje wyzwania związane z diagnostyką i leczeniem nerwiaków osłonkowych umiejscowionych w okolicy szyi.

Słowa kluczowe: *neurilemmoma*, nowotwory wywodzące się z osłonki nerwowej, nerw przeponowy

Abstract

Schwannoma (neurilemmoma) arising from the cervical phrenic nerve is a relatively rare tumour type. We describe a case of phrenic nerve schwannoma in the head and neck region in a patient who presented with a painless neck swelling. Analysis of aspiration was suggestive of benign nerve sheath tumour. A computed tomography scan was done to confirm the location and entity of the tumour. The patient was treated with complete excision of the tumour including the maternal nerve fibres. He developed right hemidiaphragm palsy postoperatively, which was treated conservatively. Postoperative immunohistochemistry examination established the diagnosis of cellular schwannoma. The case highlights the challenges associated with the diagnosis and management of cervical schwannoma.

Keywords: neurilemmoma, nerve sheath neoplasms, phrenic nerve

INTRODUCTION

Schwannoma (neurilemmoma) is a slow-growing benign encapsulated tumour that originates from Schwann cells of the peripheral nerve sheath. It can occur in various parts of the body including the head and neck region, with an incidence of approximately 25–40% of all schwannoma cases[1]. Schwannoma of the head and neck may arise from the cranial nerves, commonly from the V, VII, IV, X, XI and XII, or the sympathetic and peripheral nerves[2]. However, the incidence of phrenic nerve schwannoma in the head and neck region is relatively rare compared to intrathoracic phrenic nerve schwannoma.

CASE REPORT

A 65-year-old male presented with a 4-year history of a slow-growing right neck mass. It was a painless, smooth, solitary mass with no obstructive or constitutional symptoms. Local examination revealed 8 × 8 cm swelling over level III and IV right cervical region. The mass appeared firm, mobile in all directions, and non-tender. Computed tomography (CT) scan of neck revealed a well-delineated mass with necrotic changes, measuring 5 × 4.6 × 7 cm. The mass was located posterior to the right sternocleidomastoid muscle, with a clear plane of demarcation. The carotid artery and jugular vein were displaced medially, but still patent. Superiorly. the mass extended as far as the level of the hyoid bone. A provisional diagnosis of fibrolipoma or enlarged nodes was made based on the CT findings. Fine-needle aspiration cytology (FNAC) results were suggestive of a benign spindle cell lesion. The patient underwent excision of the tumour under general anaesthesia. Intraoperatively, multiple nerve-like structures invaginating or in continuity with the tumour were identified, which probably originated from the phrenic nerve based on the anatomical location of the tumour, while the vagus nerve was identified intraoperatively, and well preserved.

Grossly, the mass was well-encapsulated, ovoid in shape and greyish in colour, measuring 75 × 50 × 35 mm, with a glistering whitish cut surface. Microscopically, the lesion was cellular, composed of spindle-shaped cells arranged in a storiform pattern and short fascicles with occasional areas of hypocellularity. There was no atypia or significant mitotic activity. The tumour was strongly positive for S100 staining and negative for SMA and CD34, which was suggestive of cellular schwannoma. Postoperatively, the patient developed elevation of the right hemidiaphragm with reduced air entry over the right lower zone. He was treated with respiratory therapy until the symptoms improved with incentive spirometry. He was discharged after a week's stay and had good postsurgical outcome up to 6 months postoperatively.

DISCUSSION

Schwannoma (neurilemmoma) is a benign, slow-growing neurogenic tumour arising from the sheath of a spinal nerve root or any thoracic nerve. Schwannomas develop in patients aged 30–60 years[3], usually men[4]. Among these, only in a quarter of cases the nerve origin can be identified[3]. However, cervical schwannoma arising from the phrenic nerve is an extremely rare type, with only a few cases reported in the literature. In 1988, Grunstein et al. reviewed 16 reported cases of phrenic nerve schwannomas, of which only one was located in the cervical phrenic nerve, while the others were intrathoracic[5]. In 1998, Le Pimpec-Bartges et al. listed a total of 21 cases of phrenic nerve tumours, of which 13 were schwannomas, but none was located in the cervical tract[6].

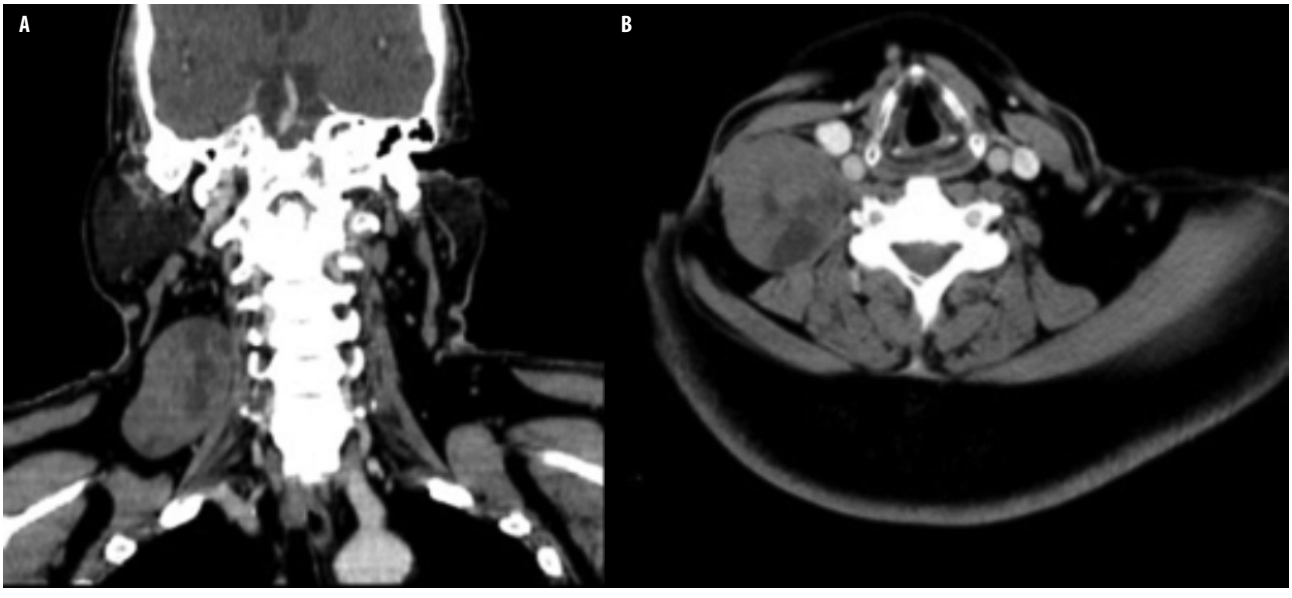

*Fig. 1. Axial view (**A**) and coronal view (**B**) of preoperative CT scan showing a soft tissue mass measuring 5 × 4.6 × 7 cm, with necrotic changes at the right lateral neck*

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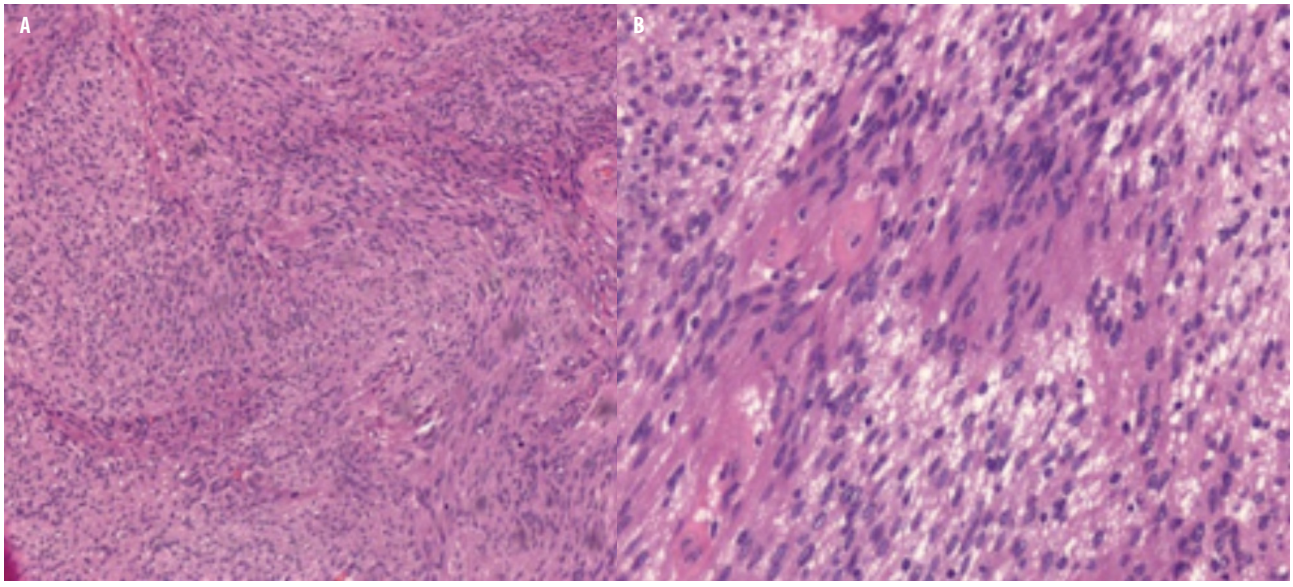

Fig. 2. **A.** *The tumour is cellular, composed of spindle cells arranged in short fascicles and showing a storiform pattern.* **B.** *Sparse hypocellular areas with Verocay bodies. H&E stain under high magnification (H&E, original ×400 magnification)*

As schwannomas are characterised by slow and progressive growth, patients remain asymptomatic with a long latent period before any manifestation occurs. The clinical symptoms vary depending on the location of the mass and nerve involvement. Most patients may have painless masses, and thus there is a need to differentiate them from other benign mesenchymal tumours. Other symptoms may include difficulty breathing, dysphagia, epistaxis, and hoarseness[7]. With regard to the phrenic nerve-related symptoms, they are also varied. Gilani and Danforth[8] reported intractable hiccups as presenting symptoms, whereas Moinuddeen et al.[9] and Mevio et al.[10] reported dyspnoea and chest discomfort which may mimic an angina attack. The lesion was identified as arising from the phrenic nerve intraoperatively in all of the above reported cases. In our case, the patient only presented with a painless mass, without any neurological deficits. The involvement of the phrenic nerve was confirmed based on postoperative right hemidiaphragm elevation.

Preoperative imaging is vital for identifying the location and origin of the tumour. In the majority of case studies, the patients were asymptomatic, thus the identification of nerve origin based on physical examination remains difficult. Imaging studies play a key role in the differential diagnosis of intracranial tumours. Particularly in cases where schwannoma is suspected, CT is routinely performed, while other diagnostic modalities such as magnetic resonance imaging (MRI), dynamic MRI, and angiography may be performed additionally[11]. However, MRI represents the gold standard for the identification of this neoplasm type. With the help of cytology analysis, spindle cell features may be indicative of a nerve sheath tumor, and the presence of a mitotic figure may indicate transformation to a malignant peripheral nerve sheath tumour. However, histochemical analysis is required for definitive diagnosis.

Grossly, schwannoma is typically encapsulated, but tumours affecting the temporal bone tend to be non-encapsulated. The tumour is usually attached to an identifiable nerve, and it is firm and rubbery, tan-white to yellow in colour. Microscopically, the tumour consists of a mixture of Antoni A and Antoni B areas. Antoni A areas are formed by short fascicles of closely packed monomorphous spindle cells with fibrillar cytoplasm. The cells sometimes form a palisaded arrangement around the acellular, collagenized foci known as Verocay bodies. Antoni B areas are composed of spindle cells haphazardly arranged within a loose myxoid stroma. Occasionally, cellular variants characterised by the lack of Antoni A and Antoni B areas are seen. On immunohistochemistry, schwannomas are strongly S100-positive, whereas neurofibromas tend to be weakly positive and also contain neurofilament-positive fibres[12]. Even though many clinical textbooks and literature describe the difference between schwannoma and neurofibroma, occasionally the distinction between these tumour types remains problematic, especially for tumours with hybrid features combining the characteristics of more than one type of benign peripheral nerve sheath tumour, i.e. neurofibroma, schwannoma and perineurioma. As in our case, cellular schwannoma has a predominantly cellular growth with the lack of Antoni B areas and Verocay bodies. This may be mistaken for neurofibroma. However, with the lack of neurofilament protein and absence of clinical features of neurofibromatosis, the diagnosis of neurofibroma was revised to cellular schwannoma after thorough discussion among the pathologists and surgeons, highlighting the difficulty that may arise.

Total surgical excision of the tumour with nerve preservation remains the treatment of choice. Kim et al. described a good outcome for intracapsular enucleation, with six out of seven patients maintaining normal postoperative

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neurological function[11]. Some authors have suggested that incomplete tumour resection may be advisable to preserve function in cases where irreparable damage to an important nerve can be expected after total excision[13]. However, Valentino et al. reported that in a case series of cervical schwannoma 64% of patients with nerve preservation developed permanent neurologic deficits[14]. This rate shows that despite efforts to preserve vital nerves, there is still a high risk of postoperative neurological deficits. In the same study, the authors also reported that they frequently encountered tumours that splayed the nerve fascicle over the entire surface of the tumour, making identification and preservation of nerve origin difficult and often impossible, as shown in our case report. Our clinical experience is contrary to the generic descriptions of neurilemmomas by a number of authors who claim that tumour excision can be done by careful dissection from the capsule without any damage to the nerve fibres[11].

CONCLUSION

The diagnosis and treatment of peripheral nerve sheath schwannoma is always a challenge, especially due to its rarity in the cervical region. A neurogenic tumour can be suspected based on clinical and FNAC results, however the final diagnosis relies on the immunohistochemistry results. Despite some distinctive features differentiating schwannoma and neurofibroma, occasionally the distinction is problematic, especially in cases involving cellular schwannoma, so good communication is required between the surgeon and the pathologist. Although complete surgical excision is the treatment of choice, the possibility of vital nerve involvement should always be kept in mind.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organisations that could negatively influence the content of the publication or claim rights to the publication.

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