RESEARCH

Imaging findings of neurogenic tumours in the head and neck region

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Objective: The aim of this study was to describe the CT, MRI and ultrasonography findings of five cases of neurogenic tumours in the head and neck region.

Methods: Five neurogenic tumours were analysed with respect to their CT value, the presence of cystic change, target sign, lobulation, connection to the nerve and vascularity.

Results: The contrast-enhanced CT (ECT) of the schwannomas demonstrated either a mass with low enhancement (two out of three cases), which reflected the predominant Antoni B components, or a mass with cystic changes, which was an Antoni A-based schwannoma displaying cystic changes (one out of three cases). On MRI, all tumours showed homogeneous and isointense signals for muscle on T_1 weighted images (T_1 WIs). T_2 weighted images (T_2 WIs) and gadolinium (Gd)-enhanced T_1 WIs demonstrated target sign in both schwannomas. Ultrasound examination showed a well-defined, ovoid or round hypoechoic mass. The direct connection to the nerve was demonstrated in two of the five cases. Lobulation was observed in only one of the five cases and cystic changes were observed in one of the five cases. In all of the cases, no vascularity was seen in power Doppler images (PDIs) obtained percutaneously.

Conclusions: Low-enhanced areas on ECTs can be specific for schwannomas, which suggests the predominance of Antoni B components. The target sign on T_2 WIs and Gd-enhanced T_1 WIs can be specific, which can be used to differentiate the two different components (Antoni A and Antoni B). The direct connection to the nerve can be a specific finding for neurogenic tumours; however, at present the sensitivity is 40%.

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Introduction

The most common peripheral nerve tumours (PNTs) are benign schwannomas and neurofibromas.^{1–3} One of the most common sites of schwannomas is the head and neck region, but these tumours are rarely observed in the extracranial region.^{4,5} Among the extracranial schwannomas, lesions in the carotid sheath are common; however, schwannomas in other sites are extremely rare. Neurofibromas, which can occur throughout the body

either as a result of neurofibromatosis or (rarely) as an isolated lesion, also are known to arise in the head and neck.⁶ Neurofibromas in the temporomandibular joint (TMJ) region are extremely rare and to the best of our knowledge there has been only one report of neurofibroma in the articular disc of the TMJ, accompanied by von Recklinghausen neurofibromatosis (Type 1).⁷

We experienced a case of schwannoma in the oral floor and a solitary neurofibroma in the TMJ region. There have been some reports about neurogenic tumours but there have been very few studies that have analysed the imaging findings of these tumours in the head and neck region.^{3,4}

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The purpose of this study is to introduce the reader to extremely rare cases of neurogenic tumours in the head and neck region and to integrate the image findings of two cases with other cases of schwannoma in the carotid sheath.

Materials and methods

Five cases of neurogenic tumours in the head and neck region were retrieved from the files of the Department of Oral and Maxillofacial Radiology, Faculty of Dentistry, Kyushu University, Fukuoka, Japan, from 1989 to 2009. Patients were examined by CT, MRI and ultrasonography.

The CT values of the tumour contents and the presence of cystic changes were evaluated on CT images. MRI was used to analyse the tumours for the presence of target sign and lobulation. The definition of the target sign has varied in past studies.^{8–11} Koga *et al*¹² examined these previous articles, defined the characteristic target sign on MRI and clarified its sensitivity and specificity based on histology. In this study, we employed the definitions of a target sign developed by Koga *et al*:¹² (1) a biphasic pattern on T_2 weighted images (T_2 WIs) and gadolinium (Gd)-enhanced T_1 weighted images (T_1 WIs); (2) higher intensity peripherally and lower intensity centrally on T_2 WIs; and (3) higher intensity centrally by focal enhancement and lower intensity peripherally on Gd-enhanced T_1 WIs. In ultrasonography, the presence of connection to the nerve, lobulation and cystic change were examined and power Doppler images (PDIs) were obtained to evaluate vascularity.

Results

CT was performed in 4 cases (cases 1–4) and contrast-enhanced CT (ECT) was performed for 3 of them (cases 1, 2 and 4). MRI was performed in 3 cases (cases 1, 2 and 5), while ultrasound examinations were performed in all of the cases and PDIs were obtained for 3 patients (cases 1, 2 and 4). Table 1 summarizes the CT, MRI and ultrasound findings of the 5 cases of neurogenic tumours. The ECT of the schwannomas demonstrated either a mass with low enhancement, which reflected the Antoni B components (Figure 1a) or a mass with cystic changes that mimicked metastatic lymph nodes (Figure 2a). On MRI, the schwannomas showed homogeneous and isointense signal similar to muscle on T_1 WIs (Figure 1b) and demonstrated the target sign and lobulation on T_2 WIs and Gd-enhanced T_1 WIs (Figure 1c,d). The direct connection to the nerve is specific to neurogenic tumours; however, this finding was only detected in two of the 5 cases on ultrasound images (Figure 2b).

The neurofibroma in the temporomandibular region did not demonstrate either the target sign on MRI or connection to the nerve on ultrasound images; therefore, the findings were non-specific (Figure 3a–c).

Discussion

Retrospective observation of ECT images of cases 1 and 2 revealed a biphasic pattern of enhancement, which was present in the low-enhanced compartment at the periphery and the slightly more enhanced compartment at the centre of the mass. The low-enhanced area was

Table 1 Summary of the CT, MRI and ultrasonography appearances of five cases of neurogenic tumour

		Case 1	Case 2	Case 3	Case 4	Case 5
Age		62	76	27	41	37
Sex		Male	Male	Female	Male	Female
Chief complaint		Painless swelling	Mass accidentally detected	Painless swelling	Painless swelling	Swelling when opening mouth and tenderness
Location		Left oral floor	Left carotid sheath	Right carotid sheath	Left carotid sheath	Right temporomandibular region
Diagnosis		Schwannoma	Schwannoma	Schwannoma	Schwannoma	Neurofibroma
CT findings	CT value (HU)	14	35	Not enhanced	Central 20 Peripheral 77	(NP)
MRI findings	Cystic change	–	–	–	+	–
	Target sign	+	+	(NP)	(NP)	–
	Lobulation	+	–	–	–	–
Ultrasound findings	Connection to the nerve	–	+	–	+	–
	Lobulation	+	–	–	–	–
	Cystic change	–	–	–	+	–
(Power Doppler)	Vascularity	+	–	(NP)	–	(NP)
		(intraoral)	(percutaneous)		(percutaneous)	

NP, not performed; + indicates presence of finding; – indicates absence of finding

Figure 1 (case 1) A 62-year-old male had been aware of a painless swelling in the left floor of his oral cavity for 1 month. Histopathological diagnosis was a schwannoma. (a) The solitary mass in the left oral floor was oval, well circumscribed and heterogeneous with low contrast-enhancement on the contrast-enhanced CT (approximately 14 HU). (b) T_1 weighted images (T_1 WIs) showed homogeneous isointensity. (c) T_2 weighted images (T_2 WIs) showed peripherally high intensity and centrally relatively low intensity, whereas gadolinium-enhanced T_1 WIs showed centrally high intensity by focal enhancement and peripherally low intensity, the so-called “target sign” (d). Lobulation is seen in (c) and (d). (e) Power Doppler images by the intraoral approach demonstrated moderate vascularity in the mass

considered as corresponding to the Antoni B area and the slightly more enhanced compartment was considered as corresponding to the Antoni A area (Figure 1a).¹² Figure 4a,b show the pathological images of a schwannoma (case 2). The central area consisted of Antoni A, which, microscopically, was a hypercellular portion with some capillaries and spindle cells with nuclear palisading (a), and the peripheral area consisted of Antoni B, which was a hypocellular portion with edematous or myxomatous stroma (b).

Case 4 was a schwannoma displaying marked cystic changes. The findings of ECT mimicked metastatic lymph nodes, but no primary tumour was detected by the systemic imaging survey (Figure 2a). Schwannomas with cystic changes have been reported previously,^{13,14} so radiologists should consider schwannoma based on their knowledge of the anatomy and the physical characteristics of nearby nerves when a well-defined, circular-type mass with an enhanced wall and cystic components with no invasion of the mass into the adjacent tissues is found by imaging.

MRI was performed in 3 cases (cases 1, 2 and 5). In all three cases, the mass lesion showed homogeneous and isointense signal to muscle on T_1 WIs (Figure 1b). Two cases of schwannoma (cases 1 and 2) presented the “target sign” (Figure 1c,d). In our study, the lower intensity on T_2 WIs and higher intensity on Gd-enhanced T_1 WIs reflected the cell-rich portion (Antoni A type components), whereas the higher intensity on T_2 WIs and lower intensity on Gd-enhanced T_1 WIs reflected the myxoid portion (Antoni B type components) of the mass.

Jee *et al*¹⁵ reported that the presence of target signs on T_2 WIs suggests a high probability of neurofibromatosis, although a fascicular appearance and thin hyperintense rim were also detected in some cases. On the other hand, Koga *et al*¹² suggested that the target sign predominantly suggests the possibility of a schwannoma. Therefore, the MRI findings of the neurofibroma were controversial. In case 5, the biphasic pattern was absent and the mass showed a wholly homogeneous and hyperintense signal on T_2 WIs (Figure 3a,b). A histopathological examination

Figure 2 (Case 4) A 41-year-old male had been aware of a painless swelling in his left upper cervical region for approximately 1 month. Histopathological examination indicated that the tumour was an Antoni A-based schwannoma displaying cystic changes. Contrast-enhanced CT images revealed a round-shaped mass lesion in the left carotid sheath with well-defined boundaries and a homogeneous low-density area roughly in the centre of the mass. (a) The CT value for the central region of the mass was approximately 20 HU and for the periphery was 77 HU. (b) Ultrasonography showed a mass that was ovoid, smoothly margined, hypoechoic and primarily homogeneous with a slight internal echo, but had no echogenic region in the centre (arrowheads). Power Doppler images did not reveal internal vascularity. A longitudinal sonogram showed a thickened nerve that was producing tapering at the end of the mass (arrow)

revealed the tumour to be a neurofibroma displaying spindle cells and collagenous fibres that were homogeneously distributed.

Ultrasonography was performed in all the cases. The images typically showed a well-defined, ovoid or round, hypoechoic, primarily homogeneous solid mass with or without a moderate posterior acoustic enhancement. Two cases (Cases 2 and 4) showed a direct connection to the nerve (Figure 2b), which was seen at the abutting

margins of the mass in the sonogram longitudinal to the carotid. Identification of such a nerve entering and exiting a mass is pathognomonic for a peripheral nerve sheath tumour^{10,16} and the nerve immediately adjacent to the PNTs may be thickened and have loss of anisotropy, thus producing a tapered appearance to the oval tumours.

Lobulation was seen in Case 1 (Figure 1c,d). Fornage¹⁷ explained that neurofibroma can be lobulated

Figure 3 (Case 5) A 37-year-old female noticed swelling when opening her mouth and tenderness in her left temporomandibular region. Limitation of her mouth opening was not seen (42 mm of maximum interincisal opening). A histopathological examination indicated that the tumour was a neurofibroma displaying spindle cells and homogeneously distributed collagenous fibers. (a) T_1 weighted images demonstrated a well-circumscribed oval mass lesion that was homogeneous and isointense to muscle in front of the right temporomandibular joint. (b) T_2 weighted images showed the lesion to be homogeneous and hyperintense. (c) Ultrasonography images showed a well-defined, ovoid, hypoechoic and primarily homogeneous, but with a slight internal echo, mass lesion with posterior acoustic enhancement

Figure 4 The pathological images of a schwannoma (Case 2). (a) The central area consisted of Antoni A, which was microscopically a hypercellular portion with some capillaries and spindle cells with nuclear palisading, and (b) the peripheral area consisted of Antoni B, which was a hypocellular portion with edematous or myxomatous stroma

and King *et al*¹⁸ reported that lobulation was seen in schwannomas and neurofibromas. Case 1 was a schwannoma and our findings agree with the features reported by King *et al*.¹⁸

King *et al*¹⁸ also reported that schwannomas are highly vascular tumours with an abundance of vessels and blood flow that can be easily obliterated with light pressure from a probe. They also noted that colour Doppler sonography with compression might prove valuable in differentiating schwannomas from neurofibromas. However, in the present study, two of the three cases of schwannoma (Cases 2 and 4) did not show internal vascularity on PDIs. In the other case (Case 1), no vascularity was seen upon examination by the percutaneous approach, whereas moderate-to-significant internal vascularity was detected from the intraoral approach (Figure 1e). The vascularity of schwannomas has been reported to range from minimal to abundant vessels;¹⁹ therefore, we concluded that the depiction of the vessels by imaging depended on the amount and diameter of the vessels. Moreover, depiction of the vessels depended on the attenuation of the ultrasound. This may be the reason why the percutaneous approach failed to detect the vascularity in Case 1.

When undertaking a differential diagnosis of a neurogenic tumour in the carotid sheath, metastatic lymph node and paraganglioma may be considered. The CT appearance of paraganglioma reflects the hypervascular nature of the tumour, resulting in homogeneous and intense enhancement after administration of intravenous contrast material. The MRI appearance is that of

a lesion exhibiting low signal intensity on T_1 WIs and proton density-weighted images and a high signal intensity on T_2 WIs. Multiple areas of high and low signal intensity, the so-called “salt and pepper appearance”, can be seen within the lesion, representing fast and slow flow.^{20,21}

Conclusion

The imaging findings of five cases of neurogenic tumours in the head and neck region were presented. The ECT of the schwannomas demonstrated either a mass with low enhancement, which reflected the Antoni B components or a mass with cystic changes, which mimicked a metastatic lymph node. On MRI, the tumours showed homogeneous and isointense signal to muscle on T_1 WIs and the target sign was specific to the schwannoma. Ultrasonography showed a well-defined, ovoid or round hypoechoic mass with or without a moderate posterior acoustic enhancement. While a direct connection to the nerve is specific for a neurogenic tumour, the findings of our current study were not sensitive enough to use this method for differentiation purposes.

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