# Intranodal neurilemmoma presenting as parotid mass

D G GRANT, N BREITENFELDT\*, N A SHEPHERD†, D M THOMAS

# Abstract

Objective: We report a case of benign intranodal neurilemmoma, an extremely rare tumour arising from a nerve sheath within a lymph node.

Case report: A 67-year-old woman underwent surgery for a left-sided parotid mass. Histopathological analysis revealed a tumour arising from a lymph node within the superficial lobe of the parotid gland. The tumour demonstrated histological features of an intranodal neurilemmoma.

Conclusions: This case represents the first report of an intranodal neurilemmoma arising within a parotid lymph node, and supports the proposal that intranodal neurilemmoma be recognised as a distinct histological entity.

Key words: Neurilemmoma; Schwannoma; Parotid Gland; Lymph Nodes

# Introduction

Primary benign non-lymphoid neoplasms of lymph nodes are uncommon. Neurilemmomas arising within lymph nodes are rare and have only recently been differentiated from other spindle cell tumours.<sup>1</sup> A search of the literature found only two previously reported cases of true intranodal neurilemmomas.<sup>2,3</sup> We present a case of an intranodal neurilemmoma presenting as a parotid mass.

#### **Case report**

### Clinical aspects

A 67-year-old woman presented to the out-patients department with a six-month history of a slowly enlarging, non-tender left parotid mass. The patient had no other associated symptoms but was concerned about the cosmetic appearance of the lump. There was no significant past medical history, and no family history of any head and neck pathology or familial diseases. The patient was a non-smoker with a low alcohol intake.

Clinical examination revealed a 4 cm, ovoid mass in the left parotid gland. The lesion was well circumscribed, mobile and non-tender. The skin covering the tumour was normal and moved freely. The remainder of the examination, including flexible naso-endoscopy, was unremarkable.

A fine needle aspiration (FNA) was performed but the specimen was non-diagnostic. A second FNA was also non-diagnostic, so imaging was requested prior to surgical excision. A magnetic resonance imaging scan demonstrated a mass within the superficial lobe of the left parotid gland (Figure 1).

At surgery, an unusual, firm, multinodular lesion was found within the superficial lobe of the left parotid gland. A lateral conservative parotidectomy was performed. The well circumscribed tumour was excised complete with the superficial lobe of the parotid.

There were no post-operative complications, and facial nerve function was unaffected. The patient was discharged home the following day.

At followup, the wound had healed well. After eight months, there was no sign of local recurrence or further lesions.

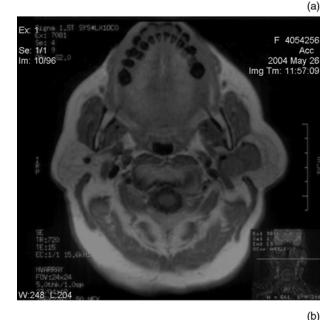
#### Histopathological findings

Macroscopically, a firm, whitish yellow,  $42 \times 28 \times 19$  mm, multinodular tumour was observed attached to the superficial parotid gland by a thin core of parotid tissue. Serial sectioning of the parotid gland revealed normal salivary gland tissue and a normal intraparotid lymph node.

Microscopy of the tumour revealed blunt-ended, spindled nuclei with cytoplasmic vacuolation, arranged in bundles and fascicles. At the periphery of the tumour, there was well defined lymphoid tissue with all the constituents of a lymph node, including a peripheral sinus and capsule (Figure 2). There was no evidence of necrosis, and mitotic activity was absent. Typical of a neurilemmoma, the tumour showed moderate cellularity and was composed of spindled, focally 'wavy' nuclei (Figure 3) with areas of Antoni A and Antoni B morphology. A diffuse if modest chronic inflammatory cell infiltrate and areas of hyalinisation were seen within the tumour. These features and foci of nucleomegaly were deemed typical of 'ancient neurilemmoma'.

Immunohistochemical analysis demonstrated that the tumour was strongly positive for S100 (Figure 4) and glial fibrillary acidic protein, and negative for desmin and  $\alpha$  smooth muscle actin. The tumour was also negative for epithelial markers, CD68, CD117, synaptophysin and HMB-45. This confirmed that the tumour was of nerve

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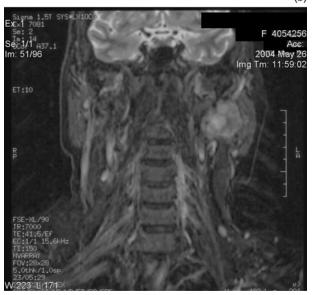


Fig. 1

(a) Axial, T1-weighted magnetic resonance image demonstrating a lobulated, intermediate-signal mass within the superficial part of the left parotid gland, displacing the retromandibular vein medially.
(b) Coronal STIR image of the same high-signal mass.

sheath origin, and a definitive diagnosis of a parotid intranodal neurilemmoma was made.

#### Discussion

Neurilemmomas arise from the nerve sheath and consist of Schwann cells in a collagenous matrix. The two main growth patterns are described as Antoni types A and B. Type A tissue has elongated spindle cells arranged in irregular streams and is compact in nature. Type B tissue has a looser organisation, often with cystic spaces intermixed within the tissue.<sup>3</sup> The cystic spaces can result in high signal intensity on T2-weighted magnetic resonance imaging. Tumours originating in Schwann cells can be

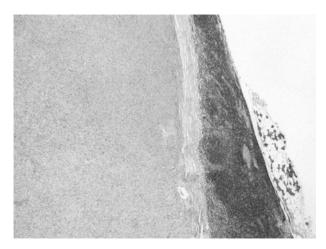


Fig. 2

Low power photomicrograph demonstrating a small amount of parotid parenchyma (right), the edge of a lymph node and the very well circumscribed tumour (left) (Hematoxylin-eosin;  $\times 12.5$ ).

detected at immunohistochemical examination by virtue of their positive staining for S100 antigen.

Weiss *et al.* reviewed a number of cases previously thought to be neurilemmomas arising in lymph nodes.<sup>1</sup> Further investigation revealed strong immunohistochemical reactivity to smooth muscle actin and desmin and negativity to S100. This, along with the presence of stellate areas of collagen deposition ('amianthoid fibres'), suggested a diagnosis of palisaded myofibroblastoma, a distinct spindle cell tumour. The authors proposed that true S100-positive neurilemmoma was a distinct and much rarer histological entity.

Neurilemmomas arising within lymph nodes are very rare.<sup>4</sup> A literature search of Medline revealed only two previous case reports of true intranodal neurilemmomas.<sup>2,3</sup> One report was of a two-year-old infant with a neurilemmoma arising in a subscapular lymph node. The other reported an elderly patient with a neurilemmoma arising within a mesenteric node, found incidentally following bowel resection for an adenocarcinoma.

Our case demonstrates the features typical of a true intranodal neurilemmoma, namely, a tumour of neural origin with ill-defined Antoni A and B areas consisting of

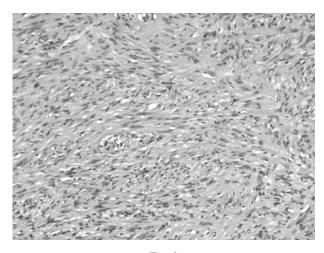


Fig. 3

High power photomicrograph of the tumour, demonstrating moderate cellularity with spindled and focally 'wavy' nuclei (Hematoxylin-eosin; ×100).

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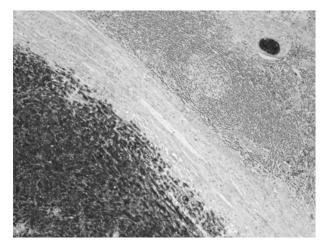


Fig. 4

S100 immunohistochemical analysis, demonstrating strong positivity of the tumour and also positivity of a small peripheral nerve (top right) ( $\times$  50).

spindle cells strongly positive for S100, encapsulated by normal lymphoid tissue. The tumour showed no evidence of amianthoid fibres, distinguishing it from the more common palisaded myofibroblastoma. A further intriguing feature in this case was that the tumour arose within a lymph node, itself within the parotid salivary gland, thus presenting as a parotid gland mass.

- We report a 67-year-old woman who underwent surgery for a parotid mass
- The mass was found to be a true benign intranodal neurilemmoma, an extremely rare tumour arising from a nerve sheath within a lymph node
- These tumours can be differentiated from other intranodal neoplasms by their characteristic microscopic appearance and positive staining for \$100 antigen
- This case report is the first of its kind, and supports the proposal that intranodal neurilemmoma be considered as a distinct histological entity

Primary benign non-lymphoid neoplasms of lymph nodes are uncommon.<sup>5</sup> The differential diagnosis of intranodal spindle cell tumours includes proliferative lesions derived from smooth muscle cells, such as leiomyoma, angiomyolipoma, leiomyomatosis and lymphangiomyomatosis, all of which show immunoreactivity to smooth muscle actin. It is important to distinguish these benign tumours from lymph node metastases of carcinomas, melanomas and other malignant tumours, which usually show marked cellularity, mitotic activity and cytological pleomorphism, together with different immunohistochemical staining patterns. The treatment of a benign neurilemmoma therefore relies on accurate histological diagnosis. Complete surgical excision is adequate as recurrence is rare, and neurilemmomas rarely show malignant transformation.<sup>6</sup>

#### Conclusion

We report a case of a neurilemmoma arising within a parotid lymph node. The lesion showed characteristic morphological and immunohistochemical features of an intranodal neurilemmoma. This finding supports the proposal of a histological entity distinct from the more common palisaded myofibroblastoma.

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