CASE REPORT

Ancient schwannoma mimicking a thyroid mass with retrosternal extension

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SUMMARY

We report the case of a middle-aged hypothyroid woman presenting with a neck swelling, voice change and breathing difficulty. On evaluation, she was diagnosed to have goitre with retrosternal extension. The mass was surgically excised and a histopathological report of ancient schwannoma was obtained. A review of literature on schwannomas of the head and neck was carried out. An ancient schwannoma should be considered as one of the differential diagnosis of a cervical mass, even though the clinical presentation point towards a thyroid swelling.

At 6 months follow-up, the patient was found to be asymptomatic except for a hoarse voice.

BACKGROUND

Schwannomas are benign nerve sheath tumours of Schwann cell origin arising from nerves covered with a neurilemmal sheath. The tumour is usually solitary and can arise from any cranial or peripheral nerve.[1] Many titles have been given to this tumour since Verocay[2] described its microscopic appearance in 1910, neurinoma, peripheral glioma, peripheral fibroblastoma, schwannoma and neurilemmoma, just to mention a few. About 25–40% of cases occur in head and neck region.[3 4] To date 100 cases of schwannomas of cervical region have been reported in literature.[5] Schwannomas originating from the vagus nerve are rare mediastinal tumours, accounting for 2% of all mediastinal neurogenic tumours, arising typically from the nerve sheath and extrinsically compressing the nerve fibres.[6] An ancient schwannoma should be kept in mind as one of the differential diagnosis of a cervical mass, even though the clinical presentation and relevant investigations point it towards being goitre.

CASE PRESENTATION

A female patient of around 30 years of age came to the outpatient department of our tertiary care hospital with symptoms of swelling in the neck of 3 years duration. She recently experienced a change in her voice for the last 2 months and breathing difficulty during the past 1 week. There was no history of difficulty or pain during swallowing, cough while taking liquids, change in appetite, body weight or menstrual cycles, intolerance to hot or cold environment or loss of hair. She had hypertension and hypothyroidism for which she was on treatment.

An examination revealed an obese woman with a short neck. There was 6×5 cm swelling on the anterior aspect of neck (figure 1), extending from the midline medially to the posterior border of the left sternocleidomastoid muscle laterally. Superiorly it was 7 cm below the chin and the inferior border could not be made out as it was extending behind the manubrium sterni. The skin over the swelling was normal. The swelling moved upwards on swallowing but there was no appreciable movement on protrusion of the tongue.

On palpation, the overlying skin was pinchable, the swelling was uniformly firm in consistency, smooth, mobile and there were no palpable lymph nodes. Lower border of the swelling was not palpable as it was extending behind the sternum. Trachea was deviated towards the right side. Laryngeal crepitus and bilateral carotid pulsations were present. Rigid laryngoscopy revealed a left vocal cord paralysis but the right cord was mobile and the glottis airway was adequate. At this stage we came to a provisional diagnosis of goitre with retrosternal extension.

INVESTIGATIONS

Thyroid function tests and routine blood investigations were normal. Ultrasound of the neck showed a large multiseptate hypoechoic lesion, measuring 5.3×4.1×7.2 cm arising from the lower pole of left lobe of thyroid gland. No obvious vascularity within the lesion was seen. Retrosternal extension was noted. A few enlarged bilateral level Ib, II and III lymph nodes were present, largest was 13×6 × 25 mm, present on the right side. Carotid and jugular vessels were normal. Impression given was of an exophytic colloid nodule of the left thyroid lobe. Sample was taken for fine needle aspiration cytology (FNAC) from the swelling, but it was reported to be inadequate for opinion.

Figure 1 Preoperative photograph of the patient showing a swelling in the anterior aspect of neck.

To cite: Pillai S, Agarwal AC, Mathew M, *et al*. *BMJ Case Rep* Published online: [*please include* Day Month Year] doi:10.1136/bcr-2013-200608

A contrast-enhanced CT (CECT) scan of the neck and thorax was performed, which showed a left thyroid mass measuring 8×5 cm with retrosternal extension up to the tracheal bifurcation (fourth thoracic vertebra). The trachea was compressed to 2×8 mm, at one point, by the mass (figure 2). Thyroid scan, after intravenous Tc^{99m} pertechnetate, showed enlarged right lobe of thyroid, and a left lateral palpable nodule, which was cold in nature, possibly an extra thyroidal mass.

TREATMENT

The patient was taken up for a thyroidectomy with a plan to do sternotomy if the lower extent of the gland was not accessible since the tumour was extending down to the level of tracheal bifurcation. A transverse cervical incision was made and the thyroid was exposed. A well-encapsulated mass was present in the region of left lobe of thyroid (figure 3). During the dissection of this mass from the surrounding structures, a cleavage plane was seen between the lower pole of the left thyroid lobe and the lesion, raising a suspicion as to whether it was a thyroid mass or another entity. Using finger dissection technique the lower extent of mass was reached, which was just above the arch of aorta. Since the mediastinal portion of mass was easily dissected off and pulled up into the neck, a sternotomy was avoided. As the mass was being dissected, the recurrent laryngeal nerve on the left side was seen to spread over the tumour, raising a suspicion of a mass arising from the nerve. All nerve fibres that were visible over the mass were isolated and freed from it. The postoperative period was uneventful and at the time of discharge, the patient's voice remained hoarse but there was no breathing difficulty.

OUTCOME AND FOLLOW-UP

The tumour was sent for histopathological examination and was reported as a mass composed of cellular spindle cells with wavy nuclei, interspersed hypocellular and cystic areas, extensive hyalinisation, haemosiderin laden macrophages and hyalinised thick-walled vessels. No mitotic figures were seen (figure 4). An immunohistochemistry analysis of the specimen was conducted, which showed tumour cells positive for vimentin and S-100 protein, focally positive for epithelial membrane antigen and creatine kinase , negative for smooth muscle actin, human melanoma black 45 and CD34, supporting a diagnosis of ancient schwannoma. The patient came for a follow-up after 6 months and was found to be asymptomatic except for a hoarse voice. Rigid angled telescopy of larynx was performed, which showed left vocal cord palsy persisting.

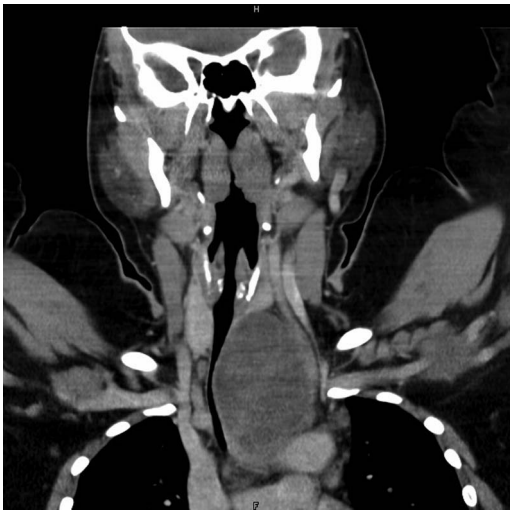

Figure 2 Contrast-enhanced CT of the neck and thorax, coronal view, showing tracheal compression by the tumour.

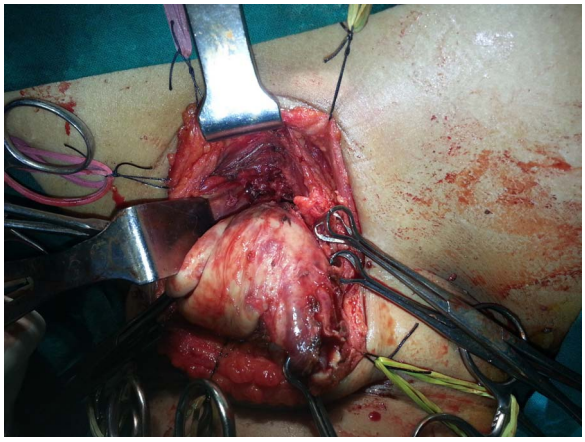

Figure 3 An intraoperative photograph showing the tumour exposed.

DISCUSSION

Neurogenic tumours can be divided into nerve sheath, ganglion cell and paraganglionic cell neoplasms.[6] Schwannoma, (neurilemmoma), is an infrequent, benign, encapsulated tumour arising from nerve sheath cells.[7] This tumour is usually solitary and can arise from any cranial or peripheral nerve.[1] It is stretched over the tumour rather than running through it as in neurofibroma.[1] Histologically five variants of schwannomas have been described namely common, plexiform, cellular, epithelioid and ancient schwannoma.[4] The term ancient neurilemmoma was first suggested by Ackerman and Taylor[8] in a review of 48 neurogenic tumours of the thorax.

Histologically a schwannoma is composed of an intimate mixture of spindle cells forming highly cellular Antoni A areas and less cellular, myxoid Antoni B areas.[2 3 9] An ancient schwannoma is a long-standing tumour, which undergoes degenerative, 'ancient' changes, dominated by large, cystic, myxoid areas with calcification, haemorrhage and hyalinisation.[4 10 11]

Schwannomas are more commonly encountered in the extremities,[4] though 25–40% of the cases originate from the neural structures of the head and neck region.[3 4] In this region

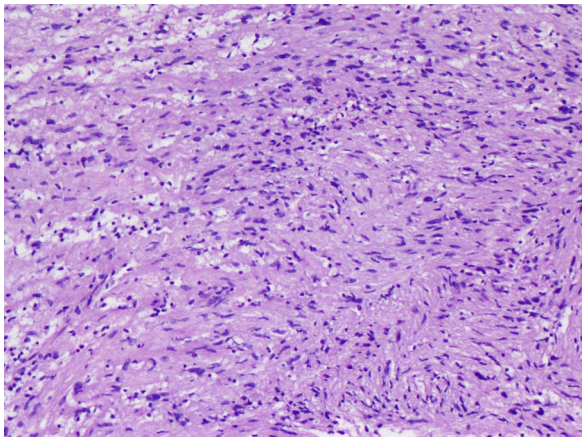

Figure 4 Tumour composed of spindle cells with wavy nuclei. H&E ×200.

schwannomas comprise a significant number of tumours of the parapharyngeal space.[12] During a review of literature we found that schwannomas mimicking a thyroid mass are rare.[1 13–15] To the best of our knowledge a case of ancient schwannoma of the left recurrent laryngeal nerve mimicking a thyroid mass with retrosternal extension producing vocal cord paralysis has not been reported in the literature, but there has been a case of schwannoma of middle mediastinum arising from left recurrent laryngeal nerve.[16]A significant percentage of ancient schwannomas are located in deep locations such as the retroperitoneum.[10]

FNAC has a questionable value as far as the diagnosis of these tumours is concerned as it can be non-specific.[1 4] CECT scan and MRI scan are mostly helpful in the preoperative planning and to a lesser extent in knowing the nature of a lesion. CECT scan is sensitive to the cystic changes that frequently accompany these tumours. MRI is capable of reliably imaging not only the tumour and its capsule but also the nerve from which the tumour arises.[3] Histopathological examination of the tumour provides the confirmatory diagnosis, as was seen in our case.

Surgical excision is the treatment of choice and if the lesion is known to be a schwannoma, it is possible to open the capsule and shell out the tumour, thereby leaving the capsular nerve fibres undisturbed and possibly avoiding functional deficit.[3] But in our case, the main mass of the tumour was found to be substernal and it was not surgically possible to dissect out the tumour from the nerve without a sternotomy which would have increased her morbidity. This case is unique because the mass was present in the neck of a hypothyroid patient, was moving with deglutition and had produced vocal cord palsy. This led to a clinical impression of a thyroid mass. As to why a schwannoma of recurrent laryngeal nerve moved with deglutition, the only possible answer we could conclude is the mass may have been attached to the pretracheal fascia.

Learning points

- During the evaluation of a neck mass, a wide possibility of lesions should be kept in mind including nerve sheath tumours.
- A cervical mass with retrosternal extension is amenable to surgical excision without a sternotomy, provided the surgical technique is suitably tailored.
- Immunohistochemistry of the lesions provides a conclusive diagnosis.

Contributors SP was the principal investigator with the other authors as co-investigators. SP and DRN were the surgeons in this case and was assisted by ACA. MM was the pathologist. The manuscript was prepared by ACA and reviewed by all the authors.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

1 Irabor DO, Ladipol JK, Nwachokor FN, *et al*. Schwannoma of the left brachial plexus mimicking a cervicomediastinal goiter in a young Nigerian lady. *West Afr J Med* 2002;21:195–6.
2 Chang SC, Schi YM. Neurilemmoma of the vagus nerve. A case report and brief literature review. *Laryngoscope* 1984;94:946–9.
3 Ku HC, Yeh CW. Cervical schwannoma: a case report and eight years review. *J Laryngol Otol* 2000;114:414–17.
4 Bayindir T, Kalcioglu MT, Kizilay A, *et al*. Ancient schwannoma of the parotid gland: a case report and review of the literature. *J Craniomaxillofac Surg* 2006;34:38–42.
5 Ahmad M, Butt RM, Ahmad N. Jugular foramen schwannoma—a very rare entity. *J Ayub Med Coll Abbottabad* 2009;21:174–5.
6 Rammos KS, Rammos SK, Foroulis CN, *et al*. Schwannoma of the vagus nerve, a rare middle mediastinal neurogenic tumor: case report. *J Cardiothorac Surg* 2009;4:68.
7 Conley JJ. Neurogenous tumors in the neck. *AMA Arch Otolaryngol* 1955;61:167–80.
8 Ackerman LV, Taylor FH. Neurogenous tumors within the thorax; a clinicopathological evaluation of forty-eight cases. *Cancer* 1951;4:669–91.
9 Gilmer-Hill HS, Kline DG. Neurogenic tumors of the cervical vagus nerve: report of four cases and review of the literature. *Neurosurgery* 2000;46:1498–503.
10 Bindra R, Gupta S, Gupta N, *et al*. Ancient schwannoma of the neck mimicking soft tissue sarcoma. *J Can Res Ther* 2010;6:234–5.
11 Ugokwe K, Nathoo N, Prayson R, *et al*. Trigeminal nerve schwannoma with ancient change. Case report and review of the literature. *J Neurosurg* 2005;102:1163–5.
12 Maniglia AJ, Chandler JR, Goodwin WJ, *et al*. Jr. Schwannomas of the parapharyngeal space and jugular foramen. *Laryngoscope* 1979;89:1405–14.
13 Mikosch P, Gallowitsch HJ, Kresnik E, *et al*. Schwannoma of the neck simulating a thyroid nodule. *Thyroid* 1997;7:449–51.
14 Badawi RA, Scott-Coombes D. Ancient schwannoma masquerading as a thyroid mass. *Eur J Surg Oncol* 2002;28:88–90.
15 Jong YN, Lee JJ, Chan YJ, *et al*. Neurilemmoma of the thyroid gland. *Intern Med* 2012;51:1641.
16 Sasaki K, Kohno T, Mun M, *et al*. Thoracoscopic removal of middle mediastinal schwannoma originating from recurrent nerve. *Thorac Cardiovas Surg* 2008;56:375–7.