CASE REPORT

Cystic schwannoma of the recurrent laryngeal nerve: a rare finding posing diagnostic difficulties

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SUMMARY

A 49-year-old woman with a painless mass in the neck was examined by the surgeon. Imaging and cytology prior to surgery suggested the mass to be either a thyroid cyst or a branchial cleft cyst. After surgery, the patient reported a hoarse voice and the pathologist confirmed the removed lesion to be a cystic schwannoma of the left recurrent laryngeal nerve. The inconclusive imaging results, combined with colloid-like material in the punctate should prompt the investigator to include cystic schwannoma in the differential diagnosis. With the probability of a neurogenic origin of the mass in mind, nerve-sparing surgery can be performed. As a future prospect, positron emission tomography scans are mentioned as a modality with possibilities to discriminate a cystic schwannoma from other common cystic lesions.

BACKGROUND

In the present case report we describe the difficulties we have encountered in diagnosing this condition and how to possibly prevent them.

Although this article describes an uncommon situation, it is important for other clinicians to recognise the combination of signs and symptoms, as an uncertain diagnosis lead to a patient with a permanent hoarse voice. When recognised in advance, appropriate measures can be taken to prevent permanent damage to the patient.

The described case is of practical use for not only the surgical professions, but also for the pathologist and the radiologist; experienced physicians were 'tricked' in each step of the diagnostic process in our case. This case report describes all the common pitfalls which lead to a wrong diagnosis and suboptimal treatment.

CASE PRESENTATION

A 49-year-old woman was referred to the surgeon's office by the ear, nose and throat (ENT) specialist with a painless mass in the neck. The patient explained that she first noticed the mass a year earlier and that it had varied in size over the following year. On physical examination a solid mass of 5×2 cm was seen, which was mobile to the skin but not to the deeper layers. The mass was located just lateral to the left hemithyroid and did not move during swallowing.

INVESTIGATIONS

Ultrasound examination showed a 25 mm diameter thick-walled cystic structure with turbid contents (figure 1). There were no suspicious cervical lymph

nodes. A CT scan was performed to further differentiate the origin of the cystic lesion, but the results were not conclusive (figure 2). Fine-needle aspiration cytology (FNAC) yielded a clear yellow cystic content, with some amorphous material and possibly a trace of colloid, not indicative of a specific type of cyst. The patient chose to have the neck mass removed surgically.

DIFFERENTIAL DIAGNOSIS

The most probable diagnoses in the differential diagnosis were either a thyroid cyst or a branchial cleft cyst (type II or III).

TREATMENT

During surgery a large thick-walled cyst was excised just ventral to the sternocleidomastoid muscle at the height of the thyroid gland. The cyst had a tail extending upwards in the direction of the tonsillar fossa, which was concordant with the probability diagnosis of a branchial cleft cyst (figure 3).

OUTCOME AND FOLLOW-UP

Postoperatively the patient had a hoarse voice for which the ENT specialist was consulted. Fiberscopy showed a complete paralysis of the left vocal chord. On examination of the excised material a schwannoma was diagnosed, which showed signs of progression towards a malignant

Figure 1 Ultrasound of the neck mass showing a thick-walled cyst with clear contents located just lateral to the left hemithyroid. The thick wall is best visible on the superficial aspect of the cyst. No flow is observed in the cyst or in the wall of the cyst on ultrasound Doppler examination.

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Figure 2 CT of the neck region showing a cystic structure lateral of the left hemithyroid. The cyst is located between the internal and external branches of the carotid artery.

schwannoma. Extensive courses of linguistic therapy did not improve the hoarseness, which led to the conclusion that the recurrent laryngeal nerve was the affected nerve.

DISCUSSION

Schwannomas are benign tumours of neurogenic origin, arising from Schwann cells surrounding the peripheral nerves. Their appearance is variable from a solid tumour to a cystic mass. Although quite rare in absolute numbers, up to 45% of schwannomas occur in the head and neck region.¹ These tumours are difficult to diagnose preoperative as they can present at a multitude of sites, mimicking a multitude of other neck masses.²

Diagnostic modalities described in the literature and other case reports are MRI, CT and ultrasound imaging, mostly combined with FNAC.^{3–5}

CT, MRI and ultrasound are used to differentiate between solid and cystic lesions and to investigate the anatomical relations of the lesion to the surrounding tissues. However, these techniques are unable to differentiate between a schwannoma and other cystic neck masses. Only one radiological sign on MRI, the so-called target sign, has been described to be 100% specific for a schwannoma.⁶

Figure 3 Intraoperative view of the cystic lesion. The tail extending upwards is clearly visible.

Another technique that is worth considering in case of a diagnostic dilemma is positron emission tomography (PET). It has not been described as an imaging modality to differentiate a branchial cleft cyst and a schwannoma, but it has been investigated in the context of differentiating malignant peripheral nerve sheath tumours and schwannomas.⁷ Both benign and malignant tumours exhibited uptake of the glucose analogue fluoro-D-glucose (FDG), with a correlation between signal strength and cellular fraction of the lesion. Schwannomas, especially of thoracic and abdominal origin, are commonly described as incidental findings on PET-CT scans, which were made to track down metastasis of malignant tumours or to locate an unknown primary tumour. Only one case report describes the use of a PET scan on a confirmed branchial cleft cyst.⁸ The branchial cleft cyst showed no FDG uptake at all when compared with the surrounding tissues, whereas a schwannoma would exhibit some uptake at least.

In our case, the differential diagnosis consisted of a thyroid cyst or a branchial cleft cyst. Both the CT scan and the FNAC were 'inconclusive', although the pathologist reported traces of colloid in the aspirate. It is reported that the amorphous contents of a schwannoma can mimic the appearance of colloid and that such a result demands a further workup.⁹ If confronted with a similar case, we would consider an additional MRI and PET-CT.

To conclude, cystic schwannomas of the neck are a rare finding and are therefore very difficult to diagnose preoperative. All of the mentioned diagnostic modalities have their own shortcomings to differentiate between cystic lesions of different origins. The combination of a cystic lesion at a location not typical for a specific disease and punctate material resembling colloid should prompt for considering a cystic schwannoma in the differential diagnosis, warranting further investigations such as MRI or PET-CT and selecting the appropriate (nerve sparing) surgical technique. In addition, use of an endotracheal tube with nerve integrity monitoring could have detected the neurogenic origin of the cyst.

Learning points

- ▶ Schwannomas are uncommon benign neurogenic tumours.
- ▶ Forty-five per cent of these schwannomas occur in the head and neck region.
- ▶ An unclear originating structure of the lesion and inconclusive fine-needle aspiration cytology warrant further investigation with, for example, MRI or positron emission tomography CT.
- ▶ If schwannoma is considered, a nerve-sparing procedure together with the neurosurgeon can be performed.

Contributors All authors contributed equally in the clinical treatment of the patient and drafting and reviewing of the manuscript.

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