Abstract. Sporadic schwannomas of the skull base are rare. We present two cases with sporadic schwannoma of this region that pose diagnostic and therapeutic problems. The first patient (female, 40 years of age) developed an extensive vagal schwannoma with deviation of the internal carotid to the medial side. A transoral extirpation of the tumour was chosen to allow for surgery without visible scars. A second patient (female, 63 years of age) developed a skull base tumour some months after resection of oral squamous cell carcinoma from the floor of the mouth. This tumour was not visible on computed-tomographic scans taken prior to ablative surgery for oral cancer. A lateral pharyngotomy was chosen in order to allow for extension of the resection in case of carcinoma spread. Healing was uneventful in both patients. Both patients developed solitary schwannomas-only, without any hint of type 2 neurofibromatosis or schwannomatosis during a follow-up of several years. Exclusion of a tumour predisposition syndrome is recommended in patients with peripheral nerve sheath tumours.

Schwannomas are benign neoplasms of peripheral nerve sheaths. They grow slowly but can become symptomatic due to expansive growth. Therapeutic concepts have to be individualized in order to achieve optimum results in delicate anatomical regions such as the skull base. Imaging techniques such as magnetic resonance imaging (MRI) (1) and positron emission tomography (PET) (2) are helpful in the differential diagnosis of schwannomas and neurofibromas from malignant peripheral nerve sheath tumours, particularly in patients with a hereditary background for nerve sheath tumour development. However, these diagnostics do not provide evidence for tumour biology. Intermediate stages of peripheral nerve sheath tumours that show findings of both benign and malignant growth are problematic (3). In particular, extensive tumours have to be differentiated from malignant peripheral nerve sheath tumours that have become symptomatic over a short period of time (4, 5). This report adds two cases of sporadic skull base schwannomas to the current literature, emphasising on imaging techniques and the surgical approach.

Case Reports

Case 1. A 40-year-old female reported a slowly increasing pressure in the region of her right mesopharynx and a roundish, firm tumour close to her right mandibular angle. Impairment of swallowing had developed over a period of several months. The inspection of the skin of the affected region was unsuspicious and the skin was easily moveable above the tumour. Mouth opening was not impaired (distance between upper and lower incisal ridge: 40 mm). Oral inspection revealed a protrusion of her right soft palate. Prior to our investigation, a biopsy had been taken from the soft palate tumour at another hospital. Histological diagnosis was ‘neurofibroma’. However, type 1 neurofibromatosis was excluded following the National Institute of Health (USA) diagnostic criteria (6). On computed-tomograms (CT) a 4.2x2.8x4 cm\(^3\) encapsulated, oval tumour was depicted, which had displaced the internal carotid artery to the medial side. This tumour had obviously developed from the right vagus nerve. The tumour showed a high uptake of gadolinium with cystic compartments. MRI confirmed the localization of the tumour to the mesopharynx and the skull base. Angiography disclosed the irregular course of the medially-shifted internal carotid artery. The patient urged for a surgical approach invisible from the outer aspect. Therefore a transoral resection was performed. Following the cutting of the soft palate, the anterior aspect of the firm tumour was


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exposed. The tumour adhered tightly to the surrounding tissues and was stepwise detached from the artery. The cavity was filled with collagen fleece and the defect was closed in layers. Healing was uneventful. Histological diagnosis revealed a schwannoma. Repeated MRI up to 10 years after surgery revealed no local tumour recurrence.

Case 2. A 63-year-old female had been treated for squamous cell carcinoma of the floor of the mouth. Ablative surgery included segmental resection of the mandible, resection of the floor of the mouth and bilateral supra-omohyoidal lymphadenectomy. Five months following surgery, a computed-tomogram was taken and a tumour was diagnosed of the left side of her neck, in the paravertebral region on level C2/C3. This lesion was not detectable on a CT scan performed prior to ablative surgery for the oral cancer. This lesion had unclear borders and was enhanced and only visualized after application of contrast material. B-Scan ultrasound depicted an oval space-occupying lesion lateral to the vertebral column. Neither the shape nor the internal structure of the lesion was diagnostic. Topography of the lesion was not in accordance with the typical drainage of the floor of the mouth. In order to clarify the type of lesion, the patient was hospitalised. A lateral pharyngotomy was chosen to access the tumour. This approach would allow an easily practicable extension of surgery in the case of malignant spread of a carcinoma. In situ, a sharply demarcated, solid tumour was dissected, which proved to be an intramuscular schwannoma. Healing was uneventful and no local recurrence was noted during 5 years of follow-up, neither for carcinoma nor for schwannoma. The patient had no signs of a neurofibromatosis.

Discussion

Schwannomas are rare tumours. However, about one out of three sporadic schwannomas is diagnosed in the head and neck region (4). Localisation in the skull base is very rare (7). Synchronous detection of oral cancer and schwannoma appears to not have been reported before. However, schwannoma of the neck may mimic distant metastasis in PET/CT (8). Differential diagnosis of sporadic nerve sheath tumours from nerve sheath tumours indicative for a hereditary disease is essential in every single case (9). MRI is the diagnostic tool of choice to reveal peripheral nerve sheath tumours (1), in particular skull base tumours (7). MRI allows no histological differentiation of these tumours but is an aid in assessing their biological behaviour (1, 2). Angiography is essential in depicting lesions following the course of the carotid arteries (10, 11). Postoperative assessment of the skull base relies on the reference to preoperative images for comparison (10). The surgical approach is tailored to the site of the lesion and should take into consideration the individual situation of the patient.

References

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Figure 2. Case no. 1. A: Anterior view of the angiogram depicting the almost bow-shaped deviation of the internal carotid to the medial side. B: Lateral view of the angiogram depicting the anterior displacement of the internal carotid. C: Intraoperative aspect during transoral resection of the schwannoma. The firm tumour is presented through the laterally pulled muscles of the soft-palate. D: Axial magnetic resonance imaging scans 5 years following extirpation of the skull base schwannoma. A small-contrast enhanced area is detected, which remained unchanged during the follow-up period.

Figure 3. Case no. 2. A: A computed-tomogram (CT) showing the mandibular defect after ablative tumour surgery (arrow). B: Axial CT scan 5 months after resection of a floor-of-the-mouth carcinoma. On the right side of the vertebral column and medial to the main vessels, a roundish non-homogeneous tumour is depicted (arrow). C: Ultrasound imaging of the region shows a pre-vertebral oval tumour with poor internal echoes. Dorsal enhancement of the ultrasound can be seen (GP, parotid gland, UK, mandible, WK, vertebral body). D: Following lateral pharyngotomy and dissection of the pre-vertebral muscles, the tumour was extirpated by blunt dissection. Caudal to the tumour the internal carotid was entangled with a loop.