Abstract  Despite the extensive branching of the trigeminal nerve, solitary neurofibromas along its branches are a rare finding. We report our management of a neurofibroma of the right auriculotemporal nerve in a 46-year-old woman. A chain of small nodules palpable in the right postauricular region was associated with increasing pain radiating into the postauricular and temporoparietal regions of her head. Magnetic resonance imaging and computed tomography showed several small ovoid lesions extending from the postauricular region to the infratemporal fossa. The lesions were removed surgically. The facial nerve adhered to the dorsal side of the largest nodule, but this could be removed without sequelae. The auriculotemporal nerve was identified as the nerve of origin and was removed together with the lesions. Histopathological examination was consistent with a neurofibroma with early plexiform cell formations. Clinical findings are discussed.

Key words  Neurofibroma · Auriculotemporal nerve · Facial nerve

Introduction

Extracranial solitary neurofibromas in the head and neck are rare findings [8]. Neurofibromas of the facial nerve have been described and mostly have been located intratemporally [9, 10, 13]. Neurofibromas of the vagus nerve, glossopharyngeal nerve, hypoglossal nerve, cervical sympathetic chain and trigeminal nerve have occurred occasionally [1, 4, 7, 12]. When they are situated along the branching of the trigeminal nerve, the nose, paranasal sinuses, infratemporal fossa, parapharyngeal space and mandibular region can be affected [1, 12].

Neurofibromas are diffuse or plexiform nerve sheath tumors. They develop from proliferating perineural fibroblasts and Schwann cells of peripheral nerves. They grow slowly within the nerve of origin, are usually well circumscribed and not encapsulated. They can occur as solitary lesions usually in the second and third decade of life without gender specificity. Neurofibromatosis due to von Recklinghausen’s disease (central type II and peripheral type I) may present clinically as multiple neurofibromas, but solitary neurofibromas can also be the first manifestation of general peripheral neurofibromatosis [2]. Malignancy rarely occurs in solitary neurofibromas but has been observed in 3–30% of cases with von Recklinghausen’s neurofibromatosis [4, 7, 9, 11, 12, 14]. Such malignancy is thought to be stimulated by hormone imbalances associated with puberty [7].

Clinical symptoms generally are due to tumor masses and increasing functional disorders or paresthesias as a result of nerve atrophy as tumor displaces nerve tissue. Diagnosis is based on clinical symptoms, computed tomography (CT), magnetic resonance imaging (MRI), and surgical biopsy with histological examination. Tumor must be resected completely but postoperative functional disorders or cosmetic loss may often make a complete excision difficult. Recurrence after complete excision is rare, but can occur due to poorly encapsulated tumor formations.

A housewife was treated at the department of otorhinolaryngology, University of Vienna, for solitary neurofibromas of the left auriculotemporal nerve. We now report on our experience in managing this patient.

Case report

A 46-year-old white Austrian housewife complained of a small, longitudinal growth behind and below her right ear. This had occurred for about 1 year with pain radiating into the periauricular and temporoparietal region of her head for about 6 months. A biopsy of this lesion was performed elsewhere and a histological diagnosis of neurofibroma was made.

When the patient came to our institution, a 3–4 cm long chain of small retroauricular nodules could be palpated. Each lesion was about 0.5 cm in diameter. The largest lesion was situated at the
caudal end of the chain. The retromandibular fossa appeared to be slightly flattened. The patient’s clinical findings were otherwise unremarkable. There were no signs of von Recklinghausen’s disease such as café au lait spots, Lisch nodules or another neurofibroma. There was also no evidence for facial nerve, gustatory or vestibular dysfunction or hearing loss.

Electromyography of the facial nerve showed symmetric muscular excitation. MRI demonstrated a well-defined 1-cm ovoid tumor in the right retromandibular fossa caudal to the external auditory canal on T2-weighted hyperintense and T1-weighted hypointense signals. Medial to this lesion, near the stylomastoid foramen, another tumor was found that was 1 cm in diameter and of the same signal weighting. Both lesions increased their signals after gadolinium enhancement (Fig. 1). CT showed no pathological bone erosion and no enlargement of the stylomastoid foramen.

Surgical removal of the nodules utilized a postauricular approach that extended along the anterior border of the sternocleidomastoid muscle. The great auricular nerve, anterior border of the sternocleidomastoid muscle, tip of the mastoid process, floor of the cartilaginous external auditory canal with its triangular process (“pointer”) were exposed, and the main trunk of the facial nerve was dissected free. A chain of retroauricular nodules, each 0.5 cm in diameter, extending to the parotid gland was removed.

An ovoid lesion of about 1 cm in diameter, was found cranio-medial to the main trunk of the facial nerve. At its cranial side the lesion adhered to a small nerve that was identified as the auriculotemporal nerve, which was presumed to be the nerve of origin. Both the nerve and lesion were removed. Another slightly smaller satellite nodule was found laterocaudal to this lesion and was excised from the floor of the external auditory canal. After surgery no changes in facial nerve function were observed, and the pain radiating into the temporoparietal region disappeared completely. The subsequent clinical course was unremarkable.

Histopathological examination of all lesions revealed multiple nerve bundles embedded in fat, connective tissue and skeletal muscle. The nerve bundles were partly circumscribed, and showed partly diffuse myxoid formations within perineural fibroblasts (Fig. 2). The nerve fascicles appeared split and contained structures possibly corresponding to early plexiform formations.

Discussion

With its sensory fibers the trigeminal nerve innervates the face from the chin to the parietal scalp, the underlying visceral organs and paranasal sinuses. Its motor fibers supply the masticator muscles. Although this nerve has the largest branching of the 12 cranial nerves, solitary neurofibromas along the trigeminal nerve have been described only in a few cases [1, 6, 12].

The auriculotemporal nerve arises from the third division of the trigeminal nerve in the infratemporal fossa, and splits to surround the middle meningeal artery before continuing behind the condylar process of the mandible. The nerve sends postganglionic parasympathetic fibers from the otic ganglion to the parotid gland, supplies sensory fibers to the anterolateral surface of the auricle, the anterior and superior wall of the external auditory canal, the anterior segment of the tympanic membrane and the skin of the preauricular and temporoparietal region. Clinical symptoms due to nerve dysfunctions include neuralgia, neuritis and gustatory sweating or hyperemia of affected skin [3].

The diagnosis of a neurogenic tumor and its nerve of origin is based on clinical findings and on CT and MRI results. The nerve of origin cannot be identified without surgical exploration [1, 4–6, 11–13]. Ultrasonography permits direct visualization of the vagus nerve, distinction between schwannomas of the cervical sympathetic chain and schwannomas of the vagus nerve and a relationship to surrounding blood vessels [11].