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OTHER ARTICLES

Multicentric Intra/Extracranial Cystic Facial Nerve Schwannoma: Case Report and Review of Literature

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Abstract To report a case of facial nerve schwannoma which presented a diagnostic and management challenge because of unusual multicentric cystic presentation. A 25-year-old female patient with a history of deep facial pain, parotid gland swelling and normal facial function showed multiple expansive cystic lesions of the temporal bone and parotid gland which turned out to be multicentric cystic facial nerve schwannomas. One should be keep this diagnosis in mind when dealing with a cystic lesions of the temporal bone and parotid gland.

Keywords Facial nerve schwannoma · Facial paralysis · Temporal bone cyst · Parotid gland tumor

Introduction

Facial nerve schwannoma is an uncommon tumour that can originate at any point along the nerve's anatomical course, from the cerebellopontine angle to its peripheral branches, and may involve more than one segment of the nerve [1, 2]. It is slow growing and originates from the neural sheath of Schwann cells [3, 4]. Schwannomas are usually benign, but cases of malignancy have been reported [1]. The cystic variant of facial nerve schwannoma is extremely rare, with only few cases described in literature [5, 6]. We report a case of a multicentric intra/extracranial cystic facial nerve schwannoma in a 25-year-old female which presented a diagnostic and treatment challenge because of its unusual presentation. To the best of our knowledge, this is the first report describing such a multi-compartment cystic facial nerve schwannoma disrupting almost the entire course of the facial nerve.

Case Report

A 25-year-old female patient presented to our Department with a 1-year history of deep facial pain and parotid gland swelling on the right side. Her medical history was non-contributory. Physical examination revealed a painful and swollen tail of the right parotid gland with normal facial nerve function. There were no signs of hearing loss, vestibulopathy or increased intracranial pressure. Clinical ENT examination showed no other abnormalities.

MRI of the head and neck region showed multiple expansive cystic lesions both extra and intra cranially. The largest expansive lesion was in the right parotid space, second large cystic lesion was expanding from the petrous part of the right temporal bone into the middle fossa and the third large cystic lesion was located in the right petrous apex (Figs. 1, 2). Bone algorithm MSCT of the head and neck region showed enlargement along the labyrinthine segment of the facial nerve canal into the geniculate fossa, thickening of the tympanic part of the facial nerve and dilatation of the mastoid part of the facial canal on the right (Fig. 3).

Initially, the patient underwent right parotid surgery and open tumour biopsy. The cystic, lobulated tumour showed an intimate contact to the facial nerve trunk distal to the stylomastoid foramen, and was resected completely

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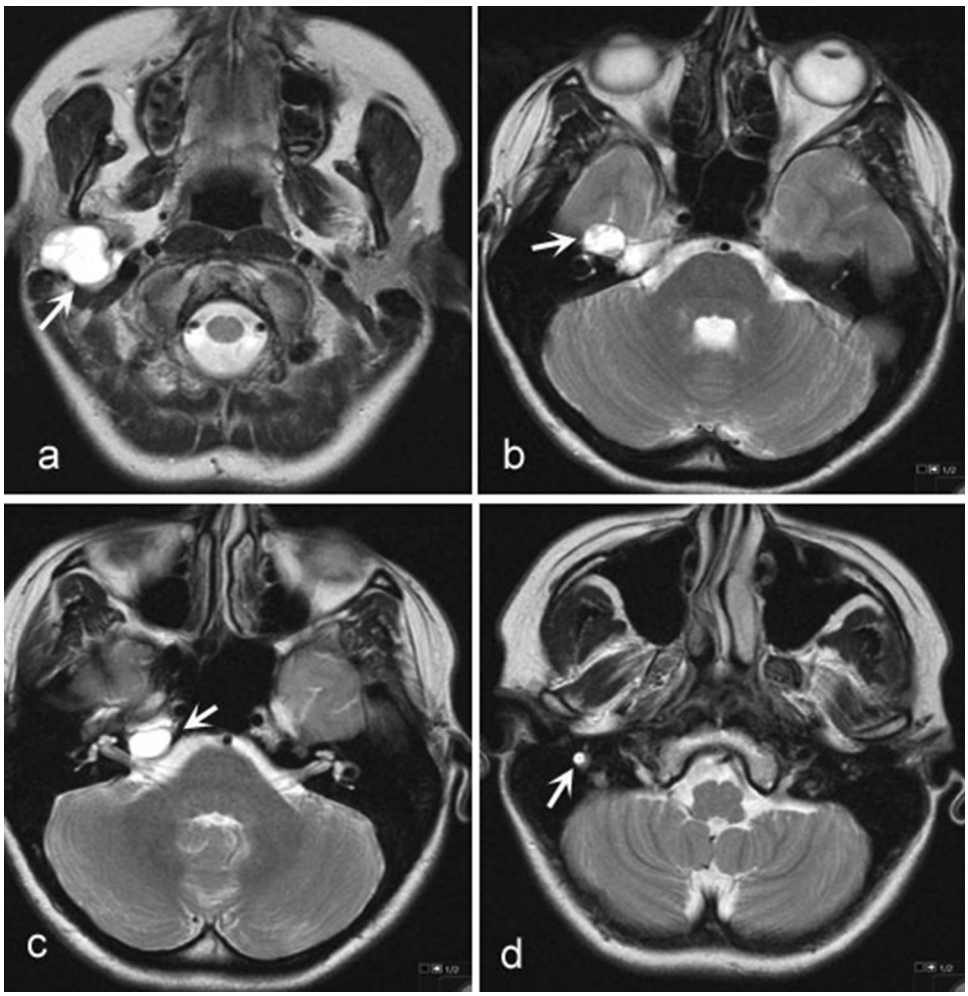

Fig. 1 Axial T2-weighted MR imaging, demonstrating a large cystic mass extending into the deep and superficial part of the right parotid (arrow in **a**), cystic lesion extending from the petrous part of the right temporal bone into the right middle fossa (arrow in **b**), cystic lesion of the petrous apex (arrow in **c**) and dilatation of the mastoid part of the facial nerve canal (arrow in **d**)

without disruption of the nerve continuity. Postoperatively, the facial function remained intact. Based on imaging and histology results, the final diagnosis of facial nerve schwannoma was made.

Because of the unbearable deep facial pain, another intervention with resection of the intracranial cystic lesions was indicated and the patient was informed that the facial nerve function will probably deteriorate. The procedure was conducted via the combined transmastoid/middle cranial fossa approach in order to preserve the hearing. The cystic tumour component invading the petrous apex was first mobilised and punctured, but as this did not result in sufficient decompression, it was resected, showing that the facial nerve was involved by the tumour from its course in the cerebellopontine angle to the stylomastoid foramen. The most challenging part of the surgery was peeling off the cystic tumour from the middle and posterior fossa dura which remained intact. The nerve with the tumour was resected, but a primary nerve reconstruction in the cerebellopontine angle was not feasible. The facial reinnervation is planned at a later stage via a masseter-facial or cross-facial anastomosis.

Discussion

Of all schwannomas, 25% originate from the head and neck, and of those most arise from the eighth nerve, but are relatively uncommon in the seventh nerve [1, 2, 7]. Schwannomas arise from the neural sheath of Schwann cells [5, 7]. In most cases they are solitary and encapsulated, eccentric to the nerve fascicles with only a few axons traversing the tumour [2, 7, 8]. A cystic subset of facial schwannoma is exceptionally rare. Rodrigues et al. reported two patients with cystic facial nerve schwannoma in the cerebellopontine angle that were treated with cyst decompression and marsupialisation [2]. Tsang et al. reported imaging of the cystic facial nerve schwannoma in the tympanic segment of the nerve [8]. In both reports there is a single cystic lesion in just one segment of the facial nerve

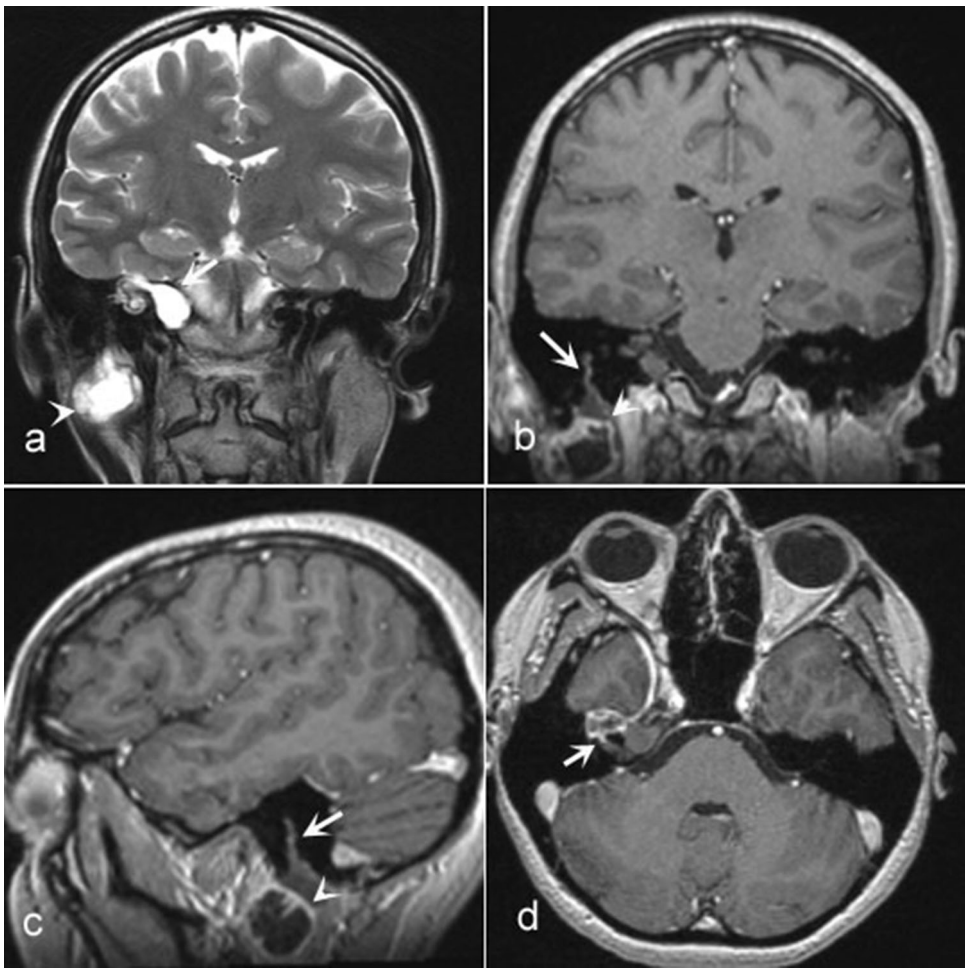

Fig. 2 T2 weighted MR imaging, demonstrating a large cystic mass into the right parotid (arrowhead in **a**) and large cystic lesion into the petrous apex on the right (arrow in **a**). Contrast-enhanced T1-weighted imaging shows dilatation of the mastoid part of the facial nerve canal on the right (arrow in **b** and **c**), peripheral contrast enhancement of the lesion into the parotid (arrowhead in **b** and **c**), peripheral contrast enhancement of the lesion expanding from the geniculate fossa and contrast enhancement of the labyrinthine segment of the facial nerve (arrow in **d**)

and diagnosis was fairly easy to make due to impaired facial nerve function.

The clinical presentation of facial schwannoma depends on the tumour location and its relationship to the surrounding structures [1, 4, 7]. Small lesions may be asymptomatic [4]. According to some authors, the most common presenting symptom is facial weakness, which occurs due to the interrupted motor component of the facial nerves. Onset may be acute or slow [7]. Recurrent facial paralysis, with intermittent periods of incomplete recovery, is a pathognomonic sign in facial nerve tumours [4]. Our patient, despite multi-compartment tumours and significant nerve involvement, presented only with deep facial pain and headache and completely normal function of the facial nerve.

It is considered difficult to set preoperative diagnosis of the facial nerve schwannoma due to its rarity and there is a limited range of clinical and radiographic features to suggest the diagnosis of schwannoma [3, 4]. Bone algorithm MSCT examinations can demonstrate bony canal enlargement, bony scalloping and remodelling of surrounding bony boundaries. MRI can show solid or cystic mass along the course of the facial nerve, hyperintense or heterogeneous on T2, iso- to hypointense on T1-weighted images. Post contrast-enhanced MR images usually demonstrate homogeneous enhancement, or peripheral enhancement in cystic lesions, as we showed in our case [9, 10]. Often, if facial nerve schwannomas are restricted to the cerebellopontine angle and/or internal auditory canal, they can be mistaken for vestibular schwannoma [7].

Observation is normally recommended for patients with preserved facial function, and is preferred until it deteriorates to House-Brackmann grade III. In the long term, however, it is unlikely for tumours not to be surgically treated [6, 7]. The main objective of surgery in case of small lesions is to slow or halt the deterioration of facial function, or to remove the tumour and then reconstruct the nerve in advanced lesions. The rationale for the first surgery in our patient was the establishment of a diagnosis and the removal of the extracranial tumour. Although the outcome was favourable and the facial function remained intact, the patient insisted upon a second intervention due to unbearable pain from severe headache, fully aware of the possibility of facial function worsening. The surgery

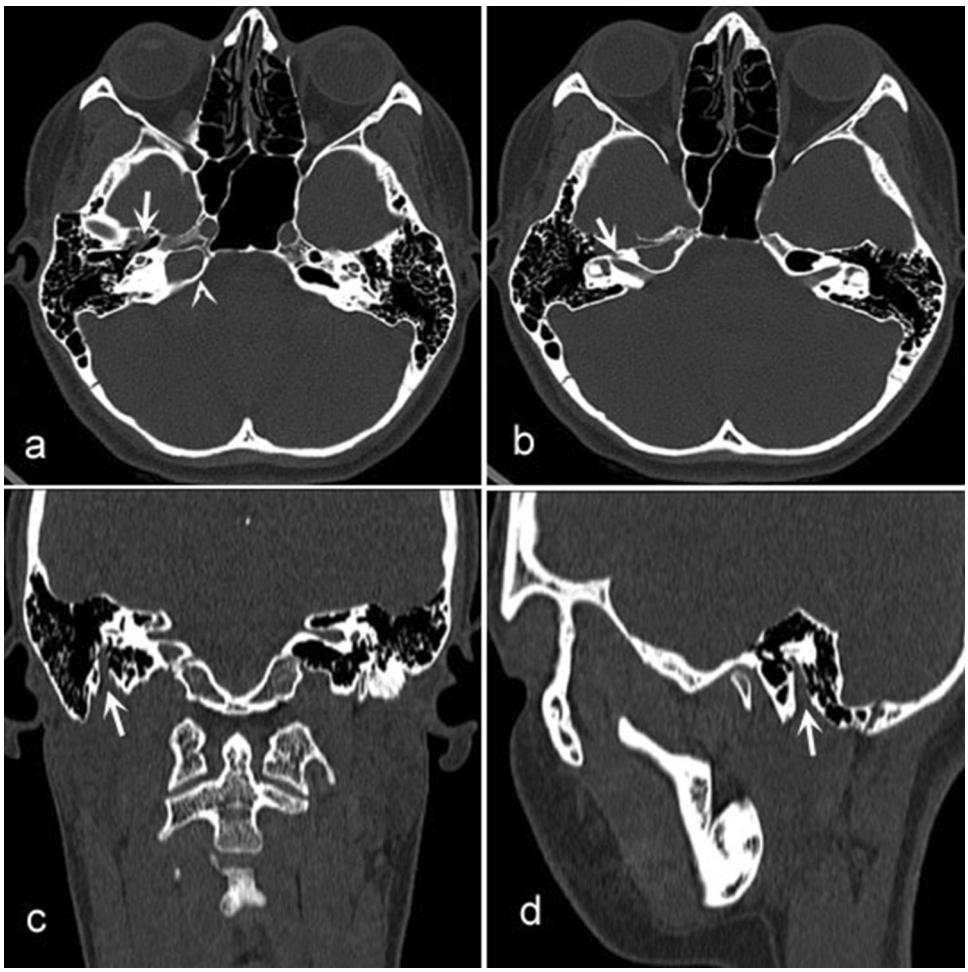

Fig. 3 MSCT (bone window) in axial plane,demonstrating expansive lytic lesion of the petrous apex (arrowhead in **a**) and enlargement of the tympanic part of the right facial nerve (arrow in **a**), enlargement along the labyrinthine segment of the facial nerve canal into the geniculate fossa (arrow in **b**), enlargement along the mastoid segment of the facial nerve canal in coronal (arrow in **c**) and sagittal plane (arrow in **d**)

was successful in removing the tumour at the cost of facial function, which requires further management.

Conclusion

Facial nerve schwannomas are rare but should be considered in the diagnosis of slowly growing lesions involving the parotid gland, temporal bone or petrous apex. A cystic variant of the facial nerve schwannoma, as presented in this report, is exceptionally rare and associated with a less favourable surgical outcome and can be mistaken for other lesions in before mentioned areas. Due to its rarity and imaging resemblance to other brain tumours, preoperative diagnosis of schwannoma is challenging. In complete tumour removal surgery, nerve fibres are usually damaged. Hence nerve reconstruction should be performed, usually resulting in a moderate level of dysfunction.

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Declarations

Conflict of interest We declare that we have no conflict of interest.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from the patient included in the study.

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