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Original research article

Treatment of endolymphatic sac tumour (Papillary adenocarcinoma) of the temporal bone

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ARTICLE INFO

Article history:
Received 7 October 2014
Accepted 18 June 2015
Available online 30 July 2015

Keywords:
Endolymphatic sac
Tumour
Temporal bone
Surgery
Papillary adenocarcinoma

ABSTRACT

Aim: To define a better treatment of sporadic endolymphatic sac tumours (ELST) analysing our experience and literature available data.
Background: ELST can arise as sporadic case (rare) or as a part of von Hippel–Lindau (VHL) disease. It is a low grade malignancy with local spread by continuity.
Materials and methods: we described our experience with 7 cases with up to date follow up.
Results: Five cases were free of disease after first surgical procedure. One case had recurrence in the temporal lobe after 12 years. One case had two surgical procedures followed by irradiation and died five years after radiotherapy with a slow disease progression.
Conclusion: With increasing expertise in the skull base surgery, complete tumour excisions are achieved in majority of the more recent cases and appear to be the treatment of choice. External irradiation is also used as palliative measures with doubtful effectiveness. Some recent reports showed encouraging results with gamma knife radiosurgery.
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1. Background

Adenomatous tumours of the temporal bone, since its description by Trietel in 1898, have been studied by various workers.[1–4] Subsequently, the primary neoplasm of the middle ear was subtyped as adenoma, papilloma, aggressive papillary middle ear tumour and mixed or papillary type.[5–7] In contrast to the mixed adenomatous tumours, the papillary tumours of the middle ear behave more aggressively with extension to the petrous apex and cranial cavity and frequently involve the facial nerve and the otic capsule.[7] In 1989, Heffner proposed that papillary tumours of the middle ear and temporal bone arise from the endolymphatic sac.[8] On the basis of histological and immunohistochemical studies of temporal bone, Heffner provided the conclusive evidence that papillary tumours of the middle ear do arise from the endolymphatic sac.[8] This article describes our experience in the management of ELST.

2. Materials and methods and results

All relevant clinical, radiological, operative and follow-up data of our cases are mentioned in Table 1.

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http://dx.doi.org/10.1016/j.rpor.2015.06.003
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Table 1 – Clinical data, results and follow up.

Sex, age (years), clinical features	Tumour location	Surgery and follow up
M/26, deafness, tinnitus, vertigo, otoscopically tumour in middle ear, X cr.nv. palsy, h/o operations	Posteroinferior surface of petrous pyramid	Combined petroccipital trans-sigmoid with translabyrinthine approach: complete tumour removal; improvement of X cr.nv. function; no recurrence
F/56, deafness, tinnitus, VII cr.nv. palsy, otoscopically tumour in hypotympanum, exploratory tympanotomy and biopsy	Jugulotympanic tumour which involves CPA, vertical part of ICA, JF and OC; extradural extension to MCF, labyrinth	Combined suboccipital transisgmoid and posterior infratemporal approach with subtotal petrosectomy with complete tumour removal. VII cr. Nv. Resected between IAC and mastoid and grafted with great auricular nv.; temporal lobe recurrence after 12 years. He refused any treatment
M/25 deafness, tinnitus, vertigo	CPA, endolymphatic canal, IAC, petrous apex, jugular foramen	Translabyrinthine approach with subtotal petrosectomy; sigmoid sinus and jugular bulb closed; no recurrence
M/70, deafness, hemifacial spasm	Posterior surface of petrous bone and CPA with IV ventricle dislocation; destruction and enlargement of IAC	Translabyrinthine transigmoid approach (closure of sigmoid sinus with surgicel); no recurrence
F/63, progressive hearing loss, h/o operation, iatrogenic VII cr.nv. palsy	Endolymphatic sac and duct, vestibular acqueduct, labyrinth, middle ear, petrous apex, mastoid, PCF and MCF	Previous surgery by transcochlear approach; revision surgery by same approach with temporal craniotomy; faciohypoglossal anastomosis; recurrence after 2 years with involvement of C1 and vertebral artery; supplementary irradiation Slowly and progressively tumour growth started 2 years after RT. Dead five years after RT
F/57, progressive hearing loss, tinnitus	Endolymphatic sac and duct, vestibular acqueduct, extension to the end of the IAC	Petro-occipital transigmoid approach (closure of sigmoid sinus with surgicel); no recurrence
M/45, deafness, VII cr.nv. palsy	>Petrous bone posterior surface, jugular foramen, IAC, middle cranial fossa	Translabyrinthine approach No recurrence

Cr: cranial; Nv: nerve; CPA: cerebellopontine angle; IAC: internal auditory canal; JF: jugular foramen; OC: occipital condyle; MCF: middle cranial fossa; PCF: posterior cranial fossa; ICA: internal carotid artery.

3. Discussion

3.1. *Origin and extension of the tumour*

The endolymphatic sac is located in the posteromedial surface of the petrous pyramid roughly midway between the sigmoid sinus and the internal auditory canal.[9] The sac consists of a proximal and a distal segment.[9] The proximal or rugose portion of the sac is contiguous with the endolymphatic duct and lies within the posterior portion of the petrous bone.[9] This portion of the sac is covered in part by the bony operculum.[9] The distal portion of the sac is located within the dura mater at the posterior cranial fossa.[9] Both pathological and radiological findings suggest that the proximal portion of the sac gives rise to the endolymphatic sac tumour.[8,10]

From its origin, the tumour growth can affect the petrous bone itself and its posterior surface in the posterior cranial fossa, facing the cerebellopontine angle.[11] From the endolymphatic sac, the tumour may erode the vestibule, the posterior semicircular canal and the mastoid cavity.[9] Subsequently, the mastoid tumour can involve the jugular bulb and the facial nerve from where it can spread anteriorly to the middle ear.[9] Middle ear tumour can, then, spread to the middle cranial fossa through the tegmen, through the tympanic membrane to the external auditory canal, and medially to the otic capsule.[9] Advanced tumours also extended anteriorly into the cavernous sinus or inferiorly to involve the skull base in the vicinity of the jugular foramen.[9,10,12]

3.2. *Surgical management*

ELST is grows slowly. Most of the tumours are usually large at time of the diagnosis. The majority of the endolymphatic sac tumours are sporadic in nature whereas some tumours are associated with von Hippel–Lindau (VHL) disease. Sporadic tumours are usually cystic, hemorrhagic and invade adjacent structures, while VHL associated cases infiltrate the bone structure and have a fibrous portion.[13] The complete tumour resection should be attempted in all cases of ELST.[8] Subtotal resection carries a high risk of bulky or multifocal recurrence.[13] The following factors should be considered before attempting a complete removal of an extensive ELST: (i) existing preoperative morbidity, (ii) expected short term and long term prognosis without treatment or with limited tumour resection, (iii) growth rate and aggressiveness of the tumour, (iv) probability of complete tumour resection versus expected procedure-related morbidity, and (v) the general medical condition and life expectancy of the patient.[9] The surgical approaches required for these tumours vary according to the size and extension of the tumours.[14] Smaller tumours localized to the endolymphatic sac area and adjoining the posterior fossa can be dealt

by transmastoid–retrolabyrinthine or by retrosigmoid approach.[14] Transmastoid–translabyrinthine is a convenient approach for tumours which involved the inner ear and mastoid bone while an infratemporal fossa approach is preferred for bigger lesions when the tumour involved the facial nerve, the jugular bulb and the middle ear.[14] A transcochlear approach is needed when complete exenteration of the otic capsule and the exposure of petrous carotid artery is necessary in extensive tumours.[14] A combined transtemporal–retrosigmoid approach give adequate exposure for tumours involving posterior and middle cranial fossae.[14] Various skull base approaches used in different series were: transmastoid, suboccipital craniotomy, translabyrinthine, infratemporal fossa, transcochlear, combined suboccipital and transmastoid, combined translabyrinthine and suboccipital, combined infratemporal and transcochlear, combined infratemporal and translabyrinthine, combined translabyrinthine and transcochlear, retrolabyrinthine presigmoid, retrolabyrinthine retrosigmoid, temporoparietal craniotomy, combined petrooccipital with transsigmoid and translabyrinthine, combined suboccipital with transsigmoid and posterior infratemporal, and combined translabyrinthine and transsigmoid.[8,13–20] With a complete resection of the tumour the long term prognosis of these patients are good.[8,13–20]

3.3. *Radiotherapy*

The selection of radiotherapy should be standard external irradiation or gamma knife radiosurgery.[13] Both pre- and postoperative external irradiation (50–60 Gy) was given to several patients in some series.[8,14–20] Heffner[8] reported 3 of 4 patients developing tumour recurrence after subtotal resection with adjuvant irradiation. In a series of 7 patients by Poletti et al., only a 63-year-old female who received irradiation where the target was defined on a contrast-enhanced MRI taken with stereotactic localization system and coregistered with simulation CT.[14] A total isocentric dose of 54 Gy in 2 Gy daily fractions was given by means of 5 non-coplanar 6 MV photon beams.[14] Field conformation was achieved by means of a 5-mm multileaf collimator.[14] Radiation was well tolerated by the patient without significant acute toxicity.[14] At 18 months after treatment, the patient is free from late complications and post irradiation MRI showed necrotic areas and a slight reduction of tumour size.[14] (Figs. 1 and 2). Two years after irradiation, the tumour progressively increased in size. The patient died five years after radiation with lower cranial nerve paralysis and worsening neurological condition. Kim et al.[21] reported 1 patient who underwent a successful salvage surgery for maintaining hearing and controlling vertigo after failing primary gamma knife therapy for ELST at an outside institution. Two patients with residual tumour underwent external beam irradiation therapy.[21] Serial imaging studies indicated that one tumour is stable and the other has been growing.[21] The tumour increased in size even following preoperative external irradiation.[18] The analysis of the published series showed that 50% of the patients who received postoperative radiotherapy following subtotal resection of ELST had further growth of the residual tumour on follow up, and 20% of those who received radiation following a complete removal of the

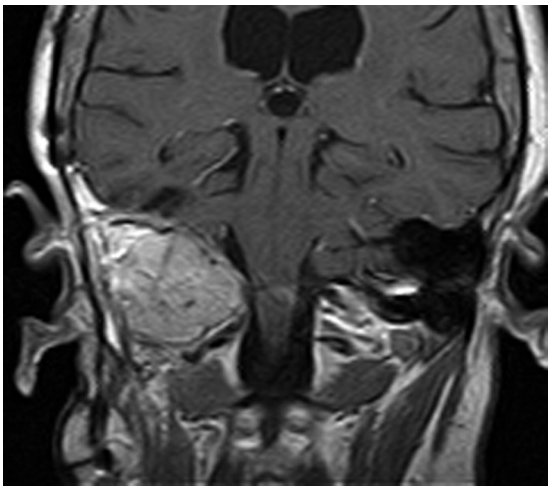

Fig. 1 – MRI before radiotherapy of the fifth case.

disease had recurrence within one year.[22] Consequently, the role of postoperative radiotherapy (fractionated or stereotactic) in the treatment of ELST is still controversial.[22]

Carlson et al.[23] reported a unique case of definitive stereotactic radiosurgery (marginal dose 15 Gy) for a 1.9-cm ELST in a medically infirm patient with VHL affording durable tumour control almost 8 years later. Hence, stereotactic radiosurgery should be considered in a poor surgical candidate or in cases of focal intracranial recurrence when the morbidity of salvage surgery is high.[23] Two patients (aged 11 years 43) underwent gamma-knife radiosurgery for recurrent tumours.[24] Both patients have a follow-up of 7 years.[24] Although not disease free, they remain neurologically stable.[24] A recurrent ELST patient received a dose of 15 Gy which was given to 50.00% with a confirmative index of 1.4.[25] Twenty shots were administered using 3 collimators (8, 14, and 18 mm).[25] The cochlea received less than 10 Gy of radiation. The patient was subsequently asymptomatic at a 2.5 year follow-up and imaging revealed regression of the tumour size. Hence, gamma-knife may be beneficial or should be considered to control areas of

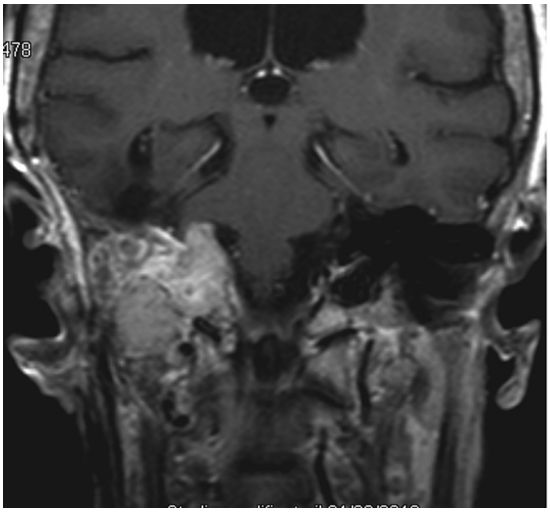

Fig. 2 – MRI two years after radiotherapy of the fifth case.

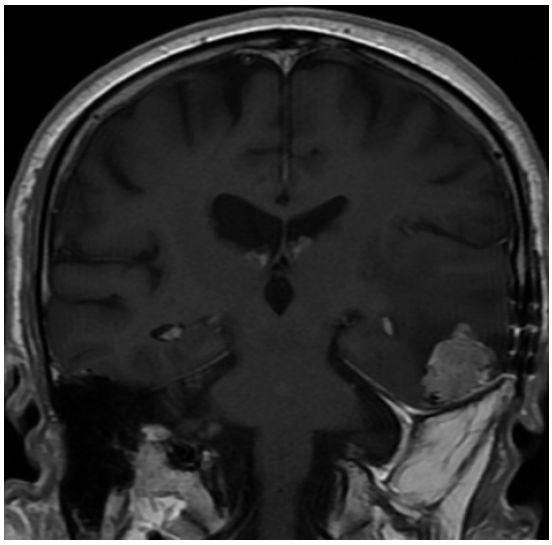

Fig. 3 – MRI showing the late recurrence in temporal lobe 12 years after surgery of the second case.

local tumour invasion, or small areas of residual or recurrent ELST.[24,25]

4. Conclusion

Complete surgical removal (with combined skull base approaches in bigger tumours) result in lasting cure without any recurrence during follow up. It is important to have a long-term follow-up to rule out delayed recurrence (Fig. 3). Lastly, the adjuvant therapy, like external irradiation, has no role in the management of these tumours. Gamma knife radiosurgery may provide longer recurrence free interval following surgery.

Conflict of interest

None declared.

Financial disclosure

None declared.

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