A case of facial nerve schwannoma with positive octreotide scintigraphy

*Un cas de schwannome du nerf facial avec scintigraphie à l'octréotide positive*

A 72-year-old woman with a 10-year history of grade VI left peripheral facial paralysis according to House Brackmann’s classification, not investigated by imaging at the time of onset, was referred following the discovery of a tumour obstructing the external auditory canal, without earache or otorrhoea. She had initially experienced vertigo, hypoacusis and pulsatile tinnitus gradually deteriorating over a period of about two years. She had no other notable personal or family history.

Pure-tone audiometry revealed mixed hearing loss of 70 dB in the left ear with a speech reception threshold of 67 dB.

CT scan of the ear showed opacification of the tympanic and external auditory canal. MRI showed a strongly gadolinium-enhanced mass measuring $31 \times 46$ mm, in close contact with the dome of the jugular bulb (Fig. 1). The radiologist concluded on a possible tympanic glomus paraganglioma. Octreotide scintigraphy demonstrated uptake by the lesion, strongly supporting the suspicion of paraganglioma (Fig. 2). No uptake was observed on MIBG scintigraphy. The absence of abnormal contrast enhancement on arteriography was an argument against paraganglioma. Surgical exploration revealed a tumour of the second and third parts of the facial nerve, corresponding to a schwannoma.

Schwannomas of the facial nerve are rare tumours, as fewer than 500 cases have been reported in the literature [1]. They have an extremely variable clinical presentation, as they can involve any segment of the nerve [1]. They originate from the labyrinthine segment in 43.5% of cases, the tympanic segment in 42.8% of cases and the mastoid segment in 36.7% of cases [1]. Multisegment lesions are common [2]. The most common clinical sign is progressive peripheral facial paralysis (73% of cases) [2]. Schwannomas protrude through the external auditory canal in only 11% of cases [2].

Tympanic paragangliomas are neuroendocrine tumours, usually benign, arising from paraganglionic tissue [3] derived from the neural crest. Tympanic paragangliomas generally arise in the promontory along the tympanic nerve. They usually present as a pulsatile raspberry tumour, raising the tympanic membrane, and are accompanied by otological (hearing loss, tinnitus) and neurological signs (glossopharyngeal, vagus, and accessory nerve lesions, and late peripheral facial paralysis).

$^{111}$Indium-labelled octreotide scintigraphy has a sensitivity of more than 95% for the diagnosis of paraganglioma [3]. Octreotide is a somatostatin analogue [4], which binds to somatostatin surface receptors. Somatostatin is essentially expressed by neuroendocrine tumours, Hodgkin's lymphomas, as well as most meningiomas and astrocytomas [4]. The original case of schwannoma reported here showed intense octreotide uptake, which has never previously been reported in the literature.

Octreotide scintigraphy has even been used in some studies for the differential diagnosis between meningioma and schwannoma [5].

This case suggests the possible presence of somatostatin receptors on the surface of some schwannomas and also highlights the fact that no complementary investigation is able to clearly establish a definitive preoperative diagnosis.

Fig. 1. Non-enhanced and contrast-enhanced MRI T1-weighted sequence of the lesion showing homogeneous contrast enhancement at the arterial phase.
Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References


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