Primary neoplasms originating in the temporomandibular joint (TMJ) are extremely rare. Their clinical manifestations are usually related to the joint dysfunction, and include preauricular mass, pain, or trismus. However, as the TMJ is closely related to the external auditory canal and the petrous bone, invasion to such structures can occur and otologic manifestations might be associated, even as the presenting complaint.

A 65-year-old woman presented at our institution with a 10-month history of progressive right-sided mixed hearing loss, facial numbness, and dizziness. Otoscopy revealed a non-pulsatile brownish mass arising from the anterior and superior walls of the ear canal (Fig. 1A). The facial nerve function was normal, and the patient did not complain of any TMJ dysfunction. High resolution computed tomography (CT) revealed an enhancing soft-tissue mass centered in the right TMJ with extensive destruction of the petrous bone and intracranial extension (Fig. 2A and B). Magnetic resonance imaging (MRI) showed a well-defined infratemporal fossa tumor, with heterogeneous signal on enhanced T1-weighted images, and remarkably low signal on TSE-T2 weighted sequence (Fig. 2C and D). Transcanal biopsy of the external ear mass confirmed the diagnosis of pigmented villonodular synovitis (PVNS).

The patient underwent a Fisch type B infratemporal approach. A subtotal petrosectomy was performed and the external and middle ear extensions of the tumor were resected. At this level, the tumor was found to engulf and infiltrate the tympanic segment of the facial nerve (Fig. 1B), extending medially into the cochlea and labyrinth. Tough preoperative facial function was normal, it was judged that leaving any residual disease already infiltrating the nerve would presumably result in postoperative paralysis due to progression. Therefore, the affected segment was sacrificed to achieve a complete resection. The TMJ elements were then excised, exposing the tumor at the infratemporal fossa, which was safely resected. Finally, a temporal craniotomy was performed to remove the intracranial extension with the infiltrated dura. A great auricular nerve graft was used to repair the facial nerve, and a temporalis muscle flap and abdominal fat were used to obliterate the cavity. The postoperative recovery was uneventful and there are no radiological signs of recurrence at the 12 months follow up.

PVNS is a rare lesion that usually arises from large diarthrodial joints, such as knee or hip. It is extremely infrequent in the TMJ, with less than 80 cases reported (1,2). Its nature is still unclear. Though it is a histologically benign giant-cell proliferation, there is evidence of cytogenetic abnormalities, and some malignant cases with metastasis have been reported. These suggest that PVNS would be a neoplastic process (3,4).

The radiological appearance of PVNS on CT is a contrast-enhancing intra-articular lesion originating in the glenoid fossa, with focal areas of hyperdensity or cysts. It produces variable bony remodeling or erosion of the adjacent bone (3). On MRI, the most characteristic finding is a mass with low signal intensity on T1 and GRE-T2* weighted sequences, reflecting the deposition of blood degradation products. Occasionally, hyperintense areas on T1 or GRE-T2* sequences may appear due to the presence of lipids or cysts, respectively (5). Differential diagnoses include synovial chondromatosis, chondroblastoma, or parotid tumors (3,5).

Otologic manifestations are not common in this condition (26% of the reported cases), but may be related to extensive bony destruction of the temporal bone and intracranial invasion. Radical excision is the treatment of choice due to its locally aggressive behavior and high risk of recurrence, which ranges from 8 to 46% (1–4). This contrasts with less aggressive types of benign tumors, such as schwannomas, where wait-and-scan or subtotal resections may be reasonable management strategies for certain cases.
The surgical approach must be carefully planned to allow for a complete removal of the tumor while minimizing surgical trauma. However, due to its infiltrative nature and the complexity of this anatomical region, this goal may require the sacrifice of certain structures (1). In those cases with invasion of the petrous bone, infratemporal fossa approaches to the skull base as described by Fisch provide good access to the infratemporal fossa with optimal control of all the structures in this region. Postoperative radiotherapy may play a role in the management of those cases with incomplete resection or recurrences, but there is limited evidence on the usefulness and side effects of radiotherapy for PVNS in this location (2).

REFERENCES