Primary parapharyngeal space neoplasms are rare among the tumors of the head and neck [1]. The majority of parapharyngeal space neoplasms are benign. Pleomorphic adenoma (PA) is the most common primary neoplasm arising in the parapharyngeal space (PPS) [2]. Benign parapharyngeal PA typically is well-demarcated and displaces the adjacent structures without direct invasion [3-4]. To our knowledge, intracranial extension of benign parapharyngeal PA has not been reported in the literature. Only one case of benign parapharyngeal PA with a small tongue-like skull base extension has been mentioned [5]. We report a rare case of a benign primary parapharyngeal PA with wide skull base remodelling and bulky intracranial extension, demonstrated by magnetic resonance imaging (MRI).

CASE REPORT

A 69-year-old male presented with right aural fullness for months. He visited a local hospital and middle ear effusion was found. He underwent grommet insertion, but the symptom persisted. He then visited our hospital for further management. Oropharyngoscope examination revealed a bulging submucosal mass underneath the right oropharynx. Magnetic resonance imaging (MRI) showed a large, lobulated, well-defined mass in parapharyngeal space with erosion of the base of the middle cranial fossa and extension to the paracavernous sinus region. The possibility of a benign PA should be included in the differential diagnoses when encountering a bulky parapharyngeal mass with intracranial extension.

ABSTRACT

Parapharyngeal pleomorphic adenoma (PA) is a benign tumor of the salivary glands. We report a rare case of giant benign parapharyngeal PA with intracranial extension in a 69-year-old male who presented with aural fullness. Magnetic resonance imaging (MRI) showed a large, lobulated, well-defined mass in parapharyngeal space with erosion of the base of the middle cranial fossa and extension to the paracavernous sinus region. The possibility of a benign PA should be included in the differential diagnoses when encountering a bulky parapharyngeal mass with intracranial extension.
Figure 1

Figure 1. a. Axial T1WI shows a large, lobulated mass of intermediate signal intensity in the right parapharyngeal space with posterolateral displacement of the right ICA, but preservation of a cleavage plane between the tumor and the deep lobe of the right parotid gland. b. Axial T2WI and c. coronal T2WI with fat saturation show that the lesion is predominantly hyperintense with streaky hypointensities inside. Extension of the lesion to the base of right middle cranial fossa is also noted (arrow). d. Post-contrast coronal T1WI with fat saturation shows moderate, heterogeneous enhancement of the tumor (arrow).
Parapharyngeal pleomorphic adenoma with intracranial extension

The patient underwent debulky surgery to remove the subcranial portion of the mass en bloc from preauricular subtemporal approach. Histopathology showed mixed epithelial and myoepithelial cells in a myxoid stroma with benign-looking nuclei of variable sizes and shapes, consistent with pleomorphic adenoma (H&E staining, ×400).

**Figure 2.** Photomicrograph reveals that the tumor contains mixed epithelial and myoepithelial cells in a myxoid stroma with benign-looking nuclei of variable sizes and shapes, consistent with pleomorphic adenoma (H&E staining, ×400).

**Figure 3.** Postoperative MRI showed an irregular enhancing soft tissue mass at the base of right middle cranial fossa with extension to the paracavernous sinus region on post-contrast coronal T1WI with fat saturation, suggestive of residual tumor (arrow). Decreased bulk of the right masticator muscle with contrast enhancement, suggestive of postoperative change with denervation atrophy (arrowhead).

**DISCUSSION**

PPS neoplasms are rare, accounted for less than 0.5% of the head and neck neoplasms. About 80% of the parapharyngeal neoplasms are benign [1]. PA is the most common primary neoplasm of the parapharyngeal space, accounted for about 40% of PPS neoplasms [2]. PPS PA is considered to have two origins: first, primary PA arising in the parapharyngeal space, and second, PA arising from the deep lobe of the parotid gland and extending medially into the parapharyngeal space. About 80% of parapharyngeal PAs are of the latter origin [6], which frequently appear dumb-bell shaped as they bulge through the stylomandibular tunnel, or appear as mass lesion without a cleavage plane with the biopsy which suggested pleomorphic adenoma.
deep lobe of the parotid gland [2, 7-9].

Typically, parapharyngeal PA is well-defined and displaces the pharyngeal mucosa medially, the carotid sheath posterolaterally, and the masticator muscles laterally without invasion [3-4, 10]. Skull base remodelling is seldom, and only one case of parapharyngeal PA with a small tongue-like extension to the skull base have been described [5]. To our knowledge, our case is the first reported of a benign giant parapharyngeal PA with large skull base remodelling and bulky intracranial extension demonstrated with MRI.

PA, also known as benign mixed tumor, is a benign neoplasm with mixed epithelial and mesenchymal components in variable proportions. Histopathologically, the epithelial component may give rise to ductal structures or myoepithelial cells, and the mesenchymal component may give rise to myxoid, hyaline, cartilaginous, or osseous stroma [11]. PA may arise in salivary glands or extra-salivary tissues. Of the salivary gland origin, PA is the most common salivary gland neoplasm, accounting for 63% of all parotid gland tumors, 59.5% of submandibular gland tumors, and 42.9% of minor salivary gland tumors along the upper aerodigestive tract or in the parapharyngeal space [12]. Sporadic cases of PA arising the soft palate, lip, paranasal sinuses, larynx, epiglottis, and trachea have also been reported [13]. Regarding the extra-salivary origin of PA, the most common site is the lacrimal gland [14]. Other reported unusual extra-salivary sites include the external auditory canal [15], middle ear and mastoid [16], posterior cranial fossa [17], skin, breast, and vulva [11].

PA commonly affects middle-aged patients, with a mean age of 46 years, and has a mild female-to-male predominance of 1.4:1 [12]. The size of the reported parapharyngeal PA ranged from 1.2 to 11 cm (median size, 5.2 cm) [2]. The typical clinical presentation of parapharyngeal PA is an asymptomatic mass displacing the oropharyngeal structures medially, followed by ear pressure or pain, a change in voice, trismus, cranial nerve deficits, obstruction of the Eustachian tubes, and rarely obstructive sleep apnea [6]. Our patient was a 69-year-old man with a 7.4 cm parapharyngeal PA, which was larger than the reported median size. The patient presented with right ear fullness due to middle ear effusion that did not respond to grommet insertion, probably secondary to compression of the Eustachian tube by the tumor.

The typical MRI characteristics of PA include a sharply marginated mass of intermediate/low T1 signal intensity and of bright T2-weighted signal intensity from the myxoid stroma [11, 18-19]. The contrast enhancement pattern varies according to the cellular and myxoid components of the tumor. As seen in our case, the tumor was predominantly bright on T2WI with scattered hypointense streaks and exhibited heterogeneous contrast enhancement, corresponding to the histopathological findings with abundant myxoid stroma interspersed with myoepithelial tissue. A clear fat plane between the tumor and parotid gland was observed on MRI, indicating that the tumor originated from ectopic salivary rests in the parapharyngeal space instead of the deep lobe of the parotid gland. Of note, our case demonstrated unique MRI findings of rare presentations of benign parapharyngeal PA: wide skull base remodelling plus direct bulky intracranial extension into the cavernous sinus and middle cranial fossa.

The differential diagnosis in this case primarily included minor salivary gland malignancies, neurogenic tumors, and paragangliomas [3, 18, 20]. They may be differentiated from each other by their inherent soft-tissue MRI appearance or the direction in which the parapharyngeal space fat or carotid sheath structures are displaced. Minor salivary gland malignancies, including adenoid cystic carcinoma, adenocarcinoma, and mucoepidermoid carcinoma [13], may show intracranial extension via perineural spread, but they generally have an infiltrating margin or show invasion of adjacent structures [3, 5, 18]. Neurogenic tumors are usually fusiform enhancing masses that cause smooth remodelling of the skull base. However, they arise from the poststyloid compartment and typically displace the ICA anteriorly [3, 5, 18]. Paragangliomas are generally vividly enhancing masses with a salt-and-pepper appearance and anterior displacement of the ICA, and they usually cause irregular, shaggy-appearing bony margins rather than smooth remodelling [3, 5, 18]. Other diagnoses to be considered include meningioma, metastatic disease, and sarcomas. Although the gross skull base erosion and intracranial extension in our case raised the suspicion of malignancy, particularly carcinoma ex pleomorphic adenoma, no malignant component was found in the specimen of our tumor excised en bloc from the extracranial portion. Additionally, the patient experienced no pain, trismus, or cranial nerve deficits, which are frequently associated with malignant tumors. There was no evidence of tumor regrowth in the 2-year follow-up.

Treatment for parapharyngeal PA is primarily surgical excision. The main surgical approaches are transcervical and transparotid-transcervical, with mandibulotomy reserved for very large or extensive tumors [6, 20]. The prognosis is good for completely resected tumors. The overall recurrence rate was 8% in a 30-year review study [6].

REFERENCES