Rare Primary Pleomorphic Adenoma in Posterior Fossa

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Key words
- Heterotopia
- Pleomorphic adenoma
- Salivary gland

Abbreviations and Acronyms
CN: Cranial nerve
CNS: Central nervous system
CT: Computed tomography
MRI: Magnetic resonance imaging

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BACKGROUND: **Pleomorphic adenoma is a benign neoplastic tumor of the salivary gland. Salivary gland tumors in the intracranial cavity are generally restricted to the pituitary gland and sellar region. To our knowledge, there has been only 1 previous case report of a primary central nervous system pleomorphic adenoma outside of the sellar region. In that case report of a posterior fossa pleomorphic adenoma, typical myxochondroid stroma was not identified on histology, and its pathogenesis was not explored.**

CASE DESCRIPTION: **A 71-year-old woman presented with a 6-week history of occipital headache and unsteadiness. Contrast-enhanced computed tomography and magnetic resonance imaging studies revealed a solitary large posterior fossa tumor in the left cerebellopontine angle measuring 47 × 43 × 45 mm. The tumor resulted in moderate hydrocephalus and significant mass effect with compression of the pons and medulla. She underwent a stereotactic right ventriculoperitoneal shunt insertion followed by a stereotactic craniotomy and complete excision of the tumor. The operation went uneventfully, and the patient had an uncomplicated recovery. Histopathologic examination revealed a benign pleomorphic adenoma (benign salivary gland tumor) with a classic appearance comprising an admixture of ductal epithelial cells, myoepithelial elements, and nodules of myxochondroid stroma. No extracranial source has been identified despite extensive investigation and 8 years of follow-up.**

CONCLUSIONS: **This case study illustrates a classic primary central nervous system pleomorphic adenoma in an unusual intracranial site. Its pathogenesis is postulated to involve salivary gland heterotopia.**

INTRODUCTION

Pleomorphic adenoma is a benign neoplastic tumor of the salivary gland.[1,2] It is the most common neoplasm of the salivary gland,[3,4] accounting for approximately 65% of all salivary gland neoplasms and 70%–80% of all benign tumors of the major salivary glands.[5] Of pleomorphic adenomas, 65%–85% arise in the parotid gland; 8% arise in the submandibular gland; and only 6%–7% arise in the minor salivary glands of the oral cavity, nasal cavity, and paranasal sinuses.[4] It is usually solitary and manifests as a painless, slow-growing, well-demarcated nodular mass in middle-aged patients.[5] It can recur after resection and invade normal adjacent tissue.[6] Although it is classified as a benign tumor, pleomorphic adenomas may undergo malignant transformation to form carcinoma ex pleomorphic adenoma.[5] Metastasis of a benign pleomorphic adenoma without malignant transformation, also known as metastasizing pleomorphic adenoma, is rare. Bone, lung, and lymph nodes are the most common sites of metastatic disease.[7] Cranial metastasis is very rare.[5,8] Patients with metastasizing pleomorphic adenoma invariably have a history of having a pleomorphic adenoma removed, and they typically have had multiple recurrences before metastatic involvement.[5,8] In this report, we describe an unusual case of a primary posterior cranial fossa classic pleomorphic adenoma in a 71-year-old woman with no previous history of salivary gland neoplasia and no other lesions identified despite extensive investigations. An 8-year follow-up examination showed no extracranial source and no recurrence of the tumor. Primary central nervous system (CNS) pleomorphic adenoma was first reported by Yano et al.[9] in 1997. To the best of our knowledge, our case study is the second case report of a primary CNS pleomorphic adenoma outside of the sellar region and the only one with unequivocal classic histologic features. We also review the literature and discuss the possibility of salivary gland heterotopia as its pathogenesis. This represents a case of a classic pleomorphic adenoma (salivary gland tumor) in an unusual intracranial site.

CASE REPORT

History and Examination

A 71-year-old woman presented with a 6-week history of occipital headache and unsteadiness. She remained well otherwise and had no significant past medical history. On examination, she was unsteady on her feet and tended to veer to the left. Romberg signs were negative. There were no other abnormal neurologic signs, including intact cranial nerve (CN) V, CN VII, and CN VIII. A contrast-enhanced computed tomography (CT) scan of the

head revealed a well-circumscribed mass lesion in the left posterior fossa. Magnetic resonance imaging (MRI) studies confirmed a solitary large posterior fossa tumor in the left cerebellopontine angle (Figure 1A–C). It measured 47 × 43 × 45 mm. There was no compromise of the adjacent transverse or sigmoid sinuses. The tumor resulted in moderate hydrocephalus and significant mass effect with compression of the pons and medulla.

Operation and Postoperative Course

The patient first underwent a stereotactic right ventriculoperitoneal shunt insertion for obstructive hydrocephalus, as the shunting procedure is a safe and effective option to allow gradual normalization of intracranial hypertension with a low risk of intraperitoneal tumor dissemination.[10,11] A postoperative CT scan of the head confirmed the shunt was in a satisfactory position. She was then readmitted 2 weeks later for a stereotactic craniotomy (unilateral left suboccipital approach) and resection of the large posterior fossa tumor. During the operation, the tumor was noted to have a fibrous consistency and was found to arise intradurally in the left cerebellopontine angle. It was attached to the adjacent petrous ridge just posterior to the petrosal vein (porus acusticus). CN VII and CN VIII were found anterior to the tumor and were visualized and preserved. CN V was not seen because this tumor was posterior and superior to the petrosal vein. This finding explains why CN V, CN VII, and CN VIII were intact on examination. The tumor was completely excised, and she made a good recovery postoperatively. A postoperative CT scan of the head was satisfactory and confirmed total resection of the tumor. After the operation, her gait remained unsteady, for which she underwent intensive rehabilitation. At the 6-month follow-up examination, her gait had recovered completely, and MRI of the head was satisfactory. She was assessed by an ear, nose, and throat surgeon for further investigations to look for a possible primary source of the salivary gland tumor. No primary lesion has been identified despite extensive examination and investigations, including salivary gland ultrasound; CT scan of the chest, abdomen, and pelvis; MRI of the head and neck; and positron emission tomography scan. She was evaluated in April 2016, 8 years after her surgery, and no evidence was found of extracranial lesion and intracranial recurrence on brain MRI (Figure 1D–F). She is currently still working as a part-time cleaner at the age of 78.

Histopathologic Findings

Formalin-fixed, paraffin-embedded hematoxylin-eosin–stained sections of the tissue specimen revealed a partially encapsulated tumor composed of duct-like structures admixed with myxochondroid stroma (Figure 2). The ducts were lined by inner luminal cuboidal to columnar epithelial cells and surrounded by an abluminal layer of myoepithelial cells. Occasional mucous cells were also present in the lining epithelium of the ducts. Luminal eosinophilic secretory material was seen (Figure 3A). There was an intimate association of the duct-like structures with the myxochondroid stroma; aggregates of plasmacytoid myoepithelial cells were seen to spread from the environs of the duct-like structures and "melt" into the adjacent stroma (Figure 3B). No carcinomatous change was seen. Tongues of tumor protruded into the capsule, and the capsule was deficient in other areas. Immunohistochemistry demonstrated strong staining of the inner

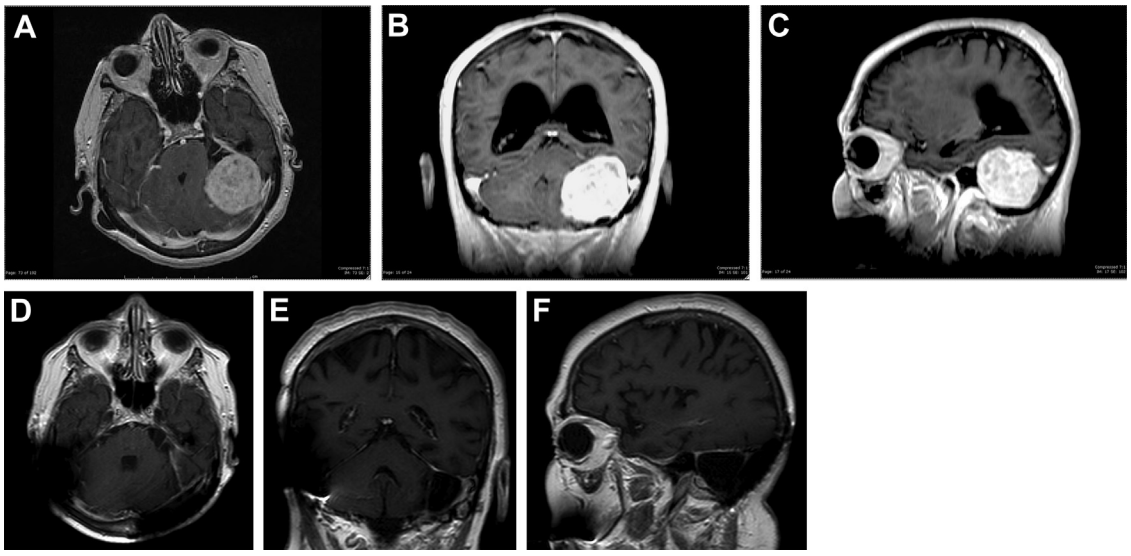

Figure 1. Neuroimaging. (**A**–**C**) Preoperative magnetic resonance imaging T1-weighted sequences with gadolinium (axial, coronal, and sagittal views) revealed a 4.7 × 4.3 × 4.5 cm posterior fossa tumor in the left cerebellopontine angle with marked mass effect and moderate hydrocephalus. The tumor demonstrates heterogeneous but prominent enhancement. (**D**–**F**) Follow-up magnetic resonance imaging T1-weighted sequences with gadolinium (axial, coronal, and sagittal views) obtained 8 years after surgery show the tumor has been completely resected with no evidence of recurrence. There was an artifact obscuring some of the right cerebellum secondary to the presence of a ventriculoperitoneal shunt over the right occipital bone.