

Primary Extracranial Meningioma Presenting as a Cheek Mass

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Meningioma is well known as common disease of the central nervous system, whereas primary extracranial meningioma is rare, representing 1% to 2% of all meningiomas. We have experienced a case of primary extracranial meningioma presenting as a right cheek mass. The tumor was completely excised via a right lateral rhinotomy incision. Histopathologic examination confirmed the diagnosis of primary extracranial meningioma.

Keywords. Meningioma, Face, Neoplasms

INTRODUCTION

Meningioma, the second most common benign tumor of the central nervous system, accounts for 13% to 26% of all intracranial neoplasms. About 20% of intracranial meningioma has extracranial disseminations to the orbit, middle ear cavity, nasal cavity, nasopharynx and paranasal sinuses, *et al.* Primary extracranial meningioma (EM), not connected to the underlying intracranial meningioma, represent 1% to 2% of all meningiomas [1]. We describe a case of primary EM at the right cheek, including the main findings of the imaging examination.

CASE REPORT

A 66-year-old woman presented with a non-tender mass of the right cheek, which slowly increased in its size over 3 years. Three years ago, she had visited with the same lesion. However, she had been lost to follow-up after computed tomography (CT), and the mass had gradually expanded over the time. Physical examination revealed a hard, mobile round mass with about 2 cm diameter. The overlying skin and cheek sensation was normal. She had

a history of left frontal bone and orbital wall fracture due to traffic accident 7 years ago. Findings of fine needle aspiration biopsy showed clusters of epithelioid cells suggesting benign tumor such as salivary gland tumor or skin adnexal tumor. Nasal cavity was normal on endoscopic examination. Preoperative CT scan revealed a heterogeneously enhanced mass in the subcutaneous fat layer of the right cheek, with well defined margin (Fig. 1). Tumor diameter has increased about 2 times compared to that of 3 years ago. There was no evidence of any intracranial mass lesion except encephalomalacia of the frontal lobe.

Under general anesthesia, she underwent complete mass excision via lateral rhinotomy incision. Intraoperative frozen biopsy was compatible with benign lesion. The tumor was surrounded by unclear and thin fibrous capsule, and tethered to the surrounding soft tissue. The relationships of the tumor with the infraorbital nerve was not identified. There was no postoperative morbidity such as cheek sensory defect. She was discharged three days after surgery. She had shown no signs of recurrence during a follow-up period of 18 months.

Microscopic examination revealed a poorly-defined tumor which is haphazardly admixed with mature adipose tissue and normal skeletal muscle. The tumor was composed of uniform, epithelioid cells showing delicate chromatin pattern, and characteristic meningotheelial whorls were frequently observed (Fig. 2). There were several foci of psammoma bodies within meningotheelial whorls. Neither mitotic figures nor significant nuclear pleomorphism was identified. Immunohistochemical studies revealed that tumor cells were reactive for epithelial membranous antigen (EMA), focally positive for S-100, but negative for HMB-

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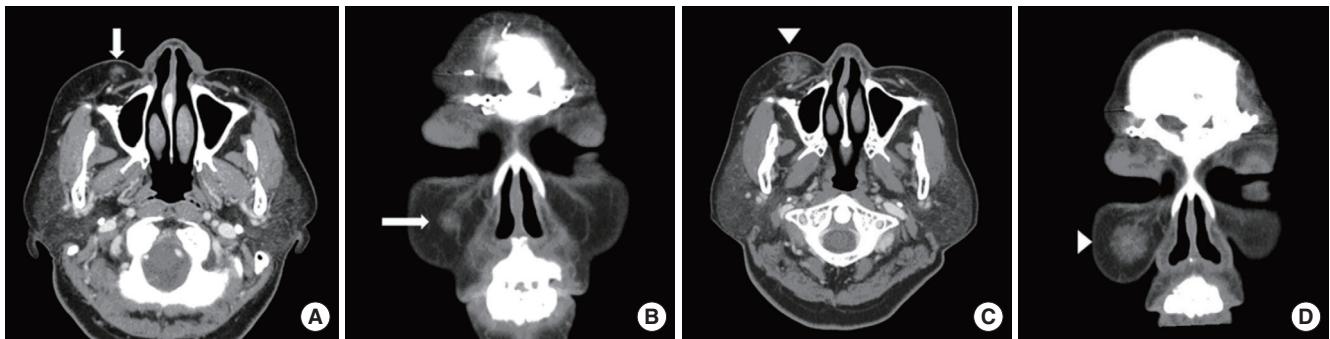


Fig. 1. Computed tomography scan. Axial and coronal images show an enhancing mass in the subcutaneous fat layer of the right cheek. (A, B) Computed tomography scan of 3 years ago shows about 1 cm-sized mass (arrow). (C, D) Tumor diameter has increased about 2 times compared to that of 3 years ago (arrowhead).

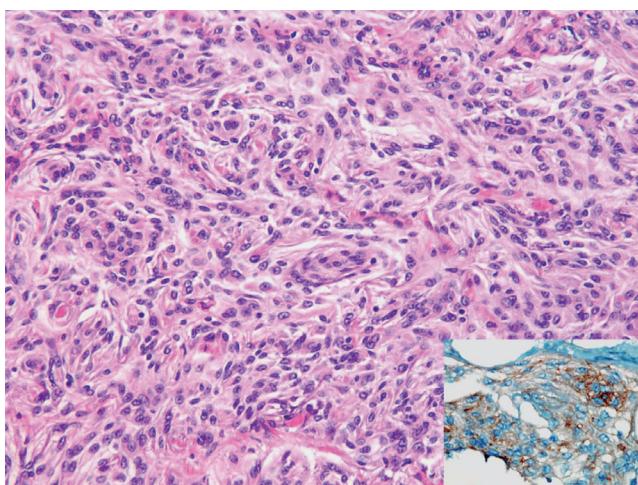


Fig. 2. Microscopic finding of the tumor. The tumor is composed of epithelioid cells forming meningothelial whorls, characteristic of meningioma (H&E, $\times 200$). Tumor cells are reactive for epithelial membranous antigen (lower right).

45, smooth muscle actin, CD31, CD34, and CD68. Additionally, Ki-67 was found to have low expression index. These histological and immunohistochemical findings were compatible with meningothelial meningioma. This case report was approved by Institutional Review Board of Hallym University Sacred Heart Hospital.

DISCUSSION

Meningioma arises from meningothelial arachnoid cells, which are derived from the neuroectoderm. There are three types of meningiomas according to malignancy grades: benign (WHO grade I), atypical (WHO grade II), and anaplastic (malignancy; WHO grade III) meningiomas. About 80% of all meningiomas are benign. The most common histological variants are meningothelial, fibrous, and transitional meningiomas [2].

Possible mechanism of primary EM is the proliferation of perineurial cells or ectopic arachnoid tissue along the cranial nerve. Another origin of primary EM is thought to be misplaced embryonic rests of arachnoid cells and multipotent mesenchymal cells [3], which can be applicable in this case.

Primary EM in the head and neck area has been reported in the facial bone, orbit, temporal bone, middle ear cavity, nasal cavity and paranasal sinuses. In the soft tissue of the head and neck, primary EM has been reported in the tonsil, scalp, soft palate and eyelid [4]. To our knowledge, the present report is the first case of primary EM located in the cheek soft tissue.

Immunohistochemical studies are essential for diagnosis of primary EM. Most extracranial meningiomas show immunohistochemical profile with variable immunoreactivity for vimentin, EMA, cytokeratin, S-100, desmoplakin and desmin. More than 95% of all meningiomas are immunopositive for EMA and vimentin [5]. Complete surgical excision without adjuvant therapy is the modality of choice for primary EM, since meningioma is radioresistant and recurrence is very rare after adequate surgical resection.

In conclusion, primary EM should be considered in patients with slowly-growing tumor in the head and neck area, and complete surgical excision is the treatment of choice.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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