Parasagittal myoepithelioma: A Case Report and Review of Literature

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Background

• Myoepithelial neoplasms were originally described in the salivary glands but have since been described in other locations; however its’ occurrence in the CNS is exceedingly rare [1].

• These tumors have varying proportions of myoepithelial and epithelial cells that behave in a spectrum ranging from benign to malignant (myoepithelioma to myoepithelial carcinoma)[2].

• They are thought to arise from ectopic glandular tissue or embryological remnants of vestigial glandular structures.

Keywords: Myoepithelioma [ME] STA [superficial temporal artery] ECA [external carotid artery] IMAX [internal maxillary artery]
Case

- 18/male with a parasagittal mass with extensive extracalvarial extension; PE showed no other palpable masses, especially in the salivary glands; s/p left parietal craniotomy, gross total excision of a malignant round cell neoplasm at 9 years of age.

Figure 1. Pre-operative photos showing the extensive extracalvarial growth of the tumor and the involvement of the overlying skin of the scalp.
• MRI with contrast: large, lobulated, homogeneously enhancing parasagittal tumor—6.0 x 4.4 x 2.9 cm; extracalvarial component—17.4 x 15.1 x 16.7 cm, with bone erosion.

• Figure 2. T1-weighted axial, coronal and sagittal contrast enhanced images.
- Cerebral catheter angiogram: hypervascular mass centred in the left posterior parietal calvarial region crossing the midline, supplied by: occipital arteries [ECA], parietal branches of the STA, and middle meningeal arteries [IMAX].

Figure 3. Cerebral angiogram A-C. Supply from the branches of the left ECA. D-F. Supply from the branches of the right ECA.
Figure 4A. Patient in prone position for surgery. Bilateral parieto-occipital craniectomy, excision of tumor with primary closure was done. 4B. Post-operative photos. Devascularization done by ligating the ECA supply with no preoperative embolization.
• Histopathology (Figure 5. A. Low power. B. High power view. C. IHC): myoepithelial cells in cords and nests; uniform architecture with myxoid stroma; no atypia. IHC of the initial tumor and the recurrence showed both to be vimentin [+]; negative for CD99, desmin, GFAP, LCA, SMA, MSA, S-100, synaptophysin. The case was signed out as a myoepithelioma [ME].

• Post op CT with contrast showed complete excision. No adjuvant treatment has been given due to the benign histology of the tumor, and the patient has since been well.
Discussion

• Variable appearance of myoepithelial cells and their phenotype, makes diagnosis challenging [4]; there is also variable immunophenotype—combinations of cytokeratin, S-100, EMA, SMA and GFAP expression [3].

• Rare; literature review reveal only 5 other cases in the CNS (cavernous sinus, orbital apex-middle cranial fossa, interhemispheric fissure, right cerebral hemisphere & cranial dura—falcine) [5].

• Explanation for the case above is that the tumor originated from salivary gland cell rests in the scalp and subcutaneous area which gradually grew to invade the parasagittal area and involve the intracranial dura [6].

• Benign myoepithelial tumors have a 20% recurrence rate and only rarely metastasize, while high-grade features can be associated with aggressive disease [6]
Conclusion

• The case above show the successful management of a very extensive parasagittal tumor with extracalvarial extension without preoperative embolization in a low income country setting.

• By applying the basic tenets in neurosurgery, even complex cases as shown above can be managed accordingly.

• In a patient presenting with a parasagittal tumor with extracalvarial extension, there can be a multitude of differential diagnoses (*meningioma, hemangiopericytoma*), and histopathologic and immunohistochemical analyses are invaluable.
References


