Case Report

Primary Myoepithelial Carcinoma of the Clivus: A Rare Presentation

Shilpi Modi, Deepa Goel, Pawan Goyal[1], Aditya Gupta[1]

Departments of Pathology and [1]Neurosurgery, Artemis Hospital, Gurgaon, Haryana, India

Abstract

Myoepithelial tumor (MET) of bone is an unusual tumor of uncertain differentiation and histogenesis. Although its presence in various bones has been reported sparsely, the presentation in clivus as primary myoepithelial carcinoma (MEC) has never been reported. They resemble their salivary gland counterparts morphologically and immunohistochemically, but harbor distinct molecular phenotype. At present, moderate nuclear atypia is the acceptable criteria to differentiate MEC from myoepithelioma. Because of their rarity, wide histopathological spectrum, and intraosseous location, MET of bone is easily confused with a variety of primary bone and cartilaginous tumors. Application of immunohistochemistry and, if required, molecular testing are required for making a correct diagnosis. In this article, we describe an extremely rare case of a primary MEC arising from the clivus, which owing to unusual location and immunohistochemical profile was diagnostically challenging.

Keywords: *Clivus, myoepithelial carcinoma, myoepithelial tumor*

Introduction

Myoepithelial tumors (MET) are rare clinicopathological entities. These tumors bear resemblance to their salivary gland counterparts, but differ in histological criteria of malignancy and have characteristic genetic aberrations. In bone and soft-tissue MET, moderate nuclear atypia is sufficient for the diagnosis of myoepithelial carcinoma (MEC). Because of their distinctive morphological and immunophenotypical heterogeneity, they often present a diagnostic challenge. Herein, we report a rare case of primary MEC originating in the clivus. To the best of our knowledge, there is no such presentation previously reported in the literature, though a metastatic MEC in clivus has been reported.[1]

Case Report

A 47-year-old male presented to our hospital with complaints of headache along with diplopia for the past 6 months. There was no history of loss of consciousness, seizures, gait disturbance, loss of vision, or any sensorimotor deficit. There was no history of swelling or pain in any other body parts. He was hypertensive for the past 6 years. The patient was clinically examined thoroughly and was found to have left abducens nerve palsy. There was no visible or palpable lump present anywhere. There was no history of any previous surgery or other ailments. Magnetic resonance imaging (MRI) of the brain [Figure 1] reported a large lobulated clival mass extending into the sellar region measuring 37 mm × 30 mm. The lesion showed heterogeneous enhancement on postcontrast scan. The radiological diagnosis rendered was chordoma or a rare possibility of invasive pituitary adenoma. Hormonal profile was found to be normal. Chest X-ray and ultrasonography of the abdomen were unremarkable.

Endoscopic transnasal transsphenoidal excision of the mass was done under neuronavigation guidance. The tumor was soft to firm in consistency, yellowish-brown in color, involving bony compartments of sphenoid sinus and clivus with no paranasal sinuses invasion. It was found to be completely extradural and sellar dura was completely intact.

The tumor was curetted and sent for histopathology. The sections showed a tumor with lobular architecture. The tumor cells were arranged in cords, small nests, and reticular pattern. The cells were

How to cite this article: Modi S, Goel D, Goyal P, Gupta A. Primary myoepithelial carcinoma of the clivus: A rare presentation. Asian J Neurosurg 2020;15:1024-6.

Submitted: 07-Apr-2020 **Revised:** 01-Jul-2020
Accepted: 13-Aug-2020 **Published:** 19-Oct-2020

Address for correspondence:
Dr. Shilpi Modi,
H.No. - 104, Tower-2,
Malibu Towne, Sector 47,
Gurgaon - 122 018, Haryana, India.
E-mail: drshilpi.modi@gmail.com

Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_144_20

Quick Response Code:

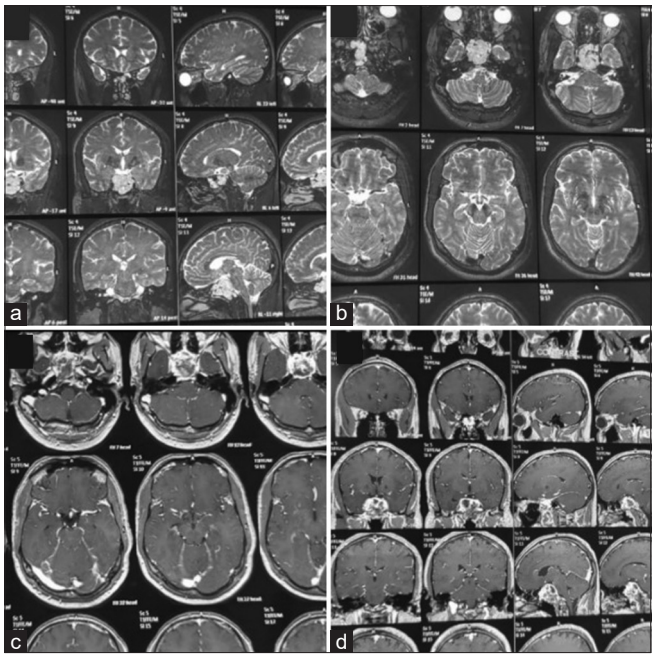

Figure 1: Magnetic resonance imaging of the patient showing a large clival mass extending superiorly into the sellar region. (a) T2 coronal and axial, (b) T2 axial, (c) T1 contrast axial, and (d) T1 contrast coronal and sagittal sections

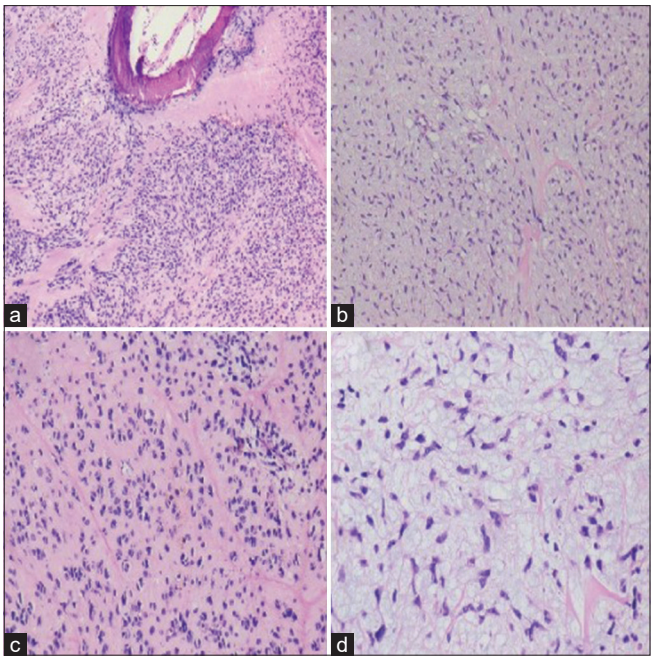

Figure 2: Microphotograph showing (a) lobulated tumor (×100), (b) spindled tumor cells in the myxoid background (×200), (c) epithelioid tumor cells with clear cytoplasm (×200), and (d) mild–to-moderate nuclear atypia (×400). H and E, original magnification (H and E)

spindled to epithelioid with mild-to-moderate nuclear atypia and eosinophilic to clear cytoplasm. Mitoses were rare. The cells were embedded in variably collagenous, myxoid, and chondromyxoid stroma [Figure 2]. The tumor had infiltrative margins.

On immunohistochemistry (IHC), the tumor cells were strongly positive for Vimentin, S100, and h-caldesmon and weakly positive for epithelial membrane antigen (EMA) [Figure 3]. Cytokeratin, glial fibrillary acidic protein (GFAP), p63, smooth muscle actin (SMA), CD31, Fli1, CD56, and synaptophysin were negative. INI1 showed retained expression. In view of typical histomorphological features and reactivity for myoepithelial and myogenic markers, it was diagnosed as a case of MEC of clivus. Postoperative MRI revealed residual disease and the patient was offered radiation therapy.

Discussion

METs of bone have a wide age distribution and show an almost equal sex distribution. The tumors usually present in long bones, but can also occur in small tubular bones and axial skeleton. By imaging, MET of bone is lytic expansile or sclerotic tumors. They may show bone destruction with cortical erosion, breach, and soft-tissue extension.[2]

Grossly, METs are variable in their consistency. Cut surface is usually glistening, myxoid, or gelatinous. Microscopically, these tumors display a range of architectural patterns, cell types, and intervening stroma. Some tumors are composed of spindle cells arranged in bundles, whereas other tumors show epithelioid cells and clear or vacuolated cells forming cell nests and cords, and therefore can be confused with chordoma or chondrosarcoma. Background stroma can be fibrous, myxoid, myxohyaline, or chondromyxoid. Metaplastic cartilage or bone formation and calcification is also frequently seen. Malignant MET can show nuclear atypia, mitoses, areas of necrosis, and infiltrative margins. At least moderate nuclear atypia is sufficient for the diagnosis of MEC.[3]

As a result of their biphenotypic nature, METs express a range of IHC markers, including epithelial and myoepithelial markers. EMA positivity has been reported within the range of 20% and 100%. S100 has been reported within the range of 72% and 100%. P63 positivity is seen between 23% and 70% of tumors and GFAP expression within 27% and 60% METs. The most commonly expressed myogenic marker in these tumors is calponin, which is reported in 86%–100% of cases, followed by SMA which is documented in 36%–64% of cases and desmin in 0%–20% of cases.[4-7]

In our case, the patient presented with diplopia due to sixth nerve palsy, which typically points toward a clival mass. Because of its axial location and histomorphology, the major differentials apart from MEC were chordoma, chondrosarcoma, and epithelioid hemangioendothelioma.

Epithelioid and clear cells in MEC along with myxoid background may show a striking resemblance to chordoma. However, the tumor cells in chordoma are larger and more vacuolated (physaliferous). IHC for brachyury is required if there is difficulty in differentiating the two.

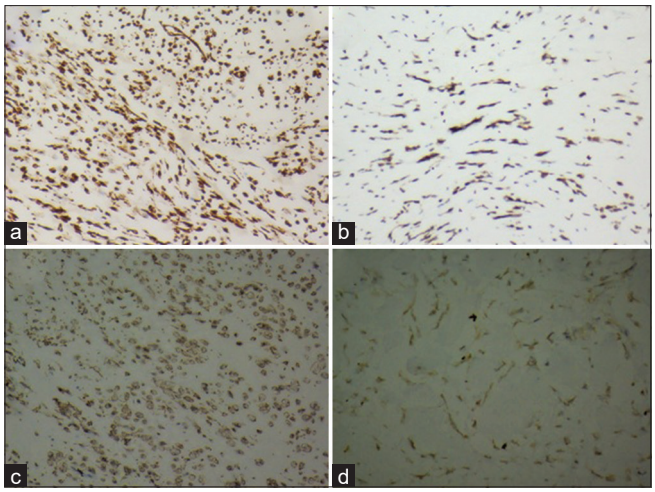

Figure 3: Microphotograph showing immunohistochemical results. (a) Vimentin positivity (diaminobenzidine, ×200), (b) S100 positivity (diaminobenzidine, ×200), (c) h caldesmon positivity (diaminobenzidine, ×200), and (d) epithelial membrane antigen positivity (diaminobenzidine, ×400)

Chondrosarcomas can have a myxoid background and can mimic MEC, but lack epithelial markers.

The presence of myogenic marker ruled out the possibility of chordoma and chondrosarcoma in our case.

Epithelioid hemangioendothelioma is composed of cords and nests of epithelioid endothelial cells in myxohyaline stroma by which it may resemble MEC. Epithelioid hemangioendothelioma can present in axial bones and may express epithelial markers. However, endothelial markers will render a correct diagnosis, which was negative in our case.

Furthermore, MEC in clivus can be confused with metastatic carcinoma. One such case has been reported previously.[1] However, clinical and radiological evaluation ruled out the possibility of metastatic carcinoma in our case.

Recent studies have unraveled certain genetic aberrations underlying METs of soft tissue and bone, including Ewing sarcoma breakpoint region 1 gene rearrangement as the most common genetic alteration, noted in approximately 45%–50% of these cases.[7]

Complete surgical excision is the treatment of choice in MET. Adjuvant radiation therapy is recommended in recurrent or malignant cases. The role of chemotherapy is unclear.

Conclusion

Primary MEC of bone is an extremely rare tumor with the variable histomorphological spectrum and divergent immunophenotype. This is the first presentation of primary MEC in clivus to the best of our knowledge; hence, it should be considered in the differential diagnosis of clival mass.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Bohnstedt BN, Tomcik M, Eads T, Hagen MC, Shah M. Metastasis of soft-tissue myoepithelial carcinoma to clivus. J Neurosurg Pediatr 2012;9:161-4.
2. Song W, Flucke U, Suurmeijer AJ. Myoepithelial tumors of bone. Surg Pathol Clin 2017;10:657-74.
3. Verma A, Rekhi B. Myoepithelial tumor of soft tissue and bone: A current perspective. Histol Histopathol 2017;32:861-77.
4. Kurzawa P, Kattapuram S, Hornicek J, Antonescu CR, Rosenberg AE, Nielsen GP. Primary myoepithelioma of bone: A report of 8 cases. Am J Surg Pathol 2013;37:960-8.
5. Kilpatrick SE, Hitchcock MG, Kraus MD, Calonje E, Fletcher CD. Mixed tumors and myoepitheliomas of soft tissue: A clinicopathologic study of 19 cases with a unifying concept. Am J Surg Pathol 1997;21:13-22.
6. Hornick JL, Fletcher CD. Myoepithelial tumors of soft tissue: A clinicopathologic and immunohistochemical study of 101 cases with evaluation of prognostic parameters. Am J Surg Pathol 2003;27:1183-96.
7. Rekhi B, Joshi S, Panchwagh Y, Gulia A, Borges A, Bajpai J, *et al*. Clinicopathological features of five unusual cases of intraosseous myoepithelial carcinomas, mimicking conventional primary bone tumours, including EWSR1 rearrangement in one case. APMIS 2016;124:278-90.