Liposclerosing Myxofibrous Tumor of the Cranial Vault

Jillian Ploof DO[1], Hamza Shaikh MD[2,3] , Jenny Melli MD[5], George Jour MD[4], Alan Turtz MD[2]

PHILADELPHIA COLLEGE OF OSTEOPATHIC MEDICINE

[1] Department of Neuroscience, Philadelphia College of Osteopathic Medicine; [2]Department of Neurosurgery, Cooper University Hospital; [3]Department of Radiology, Cooper University Hospital, Camden, NJ; [4]Department of Pathology, MDAnderson Cancer center at Cooper, Camden, NJ; [5]Department of Medicine, Cooper University Hospital, Camden, NJ

PCOM

Abstract

Background and Importance**:** Liposclerosing myxofibrous tumors (LSMFT) are rare benign fibro-osseous tumors most frequently occurring in the proximal femur. We report the first case of this rare tumor occurring within the calvarium.

Clinical Presentation: Our patient presented with a 2-year history of enlarging, painless, fixed mass over the left forehead. She underwent surgical resection and the mass was histologically confirmed to be a liposclerosing myxofibrous tumor.

Conclusion: LSMFT is a rare tumor that should remain on the differential for calvarium lesions. When diagnosed, this lesion can be removed with the goal of gross total resection and excellent cosmesis can be achieved.

Introduction

Liposclerosing myxofibrous tumor is a rare benign fibro-osseous bone lesion which most frequently occurs in the proximal femur. Eighty-to-ninety percent of these tumors are seen within the intertrochanteric region of the femur with other common locations being the tibia, humerus, and rib. LSMFT commonly presents in the fourth decade of life, with an age range of 15-80 years. The incidence is equal in men and women.

In this case report, we describe a young patient with LSMFT of the left frontal bone. We found this to be the first reported case of LSMFT of the skull. We report the clinical presentation, radiographic features, histological findings, and review the literature.

References

1. Campbell K, Wodajo F. Case Report: Two-step Malignant Transformation of a Liposclerosing Myxofibrous Tumor of Bone. 2008;466(11):2873.
2. Deel C, Hassell L. Liposclerosing Myxofibrous Tumor. Arch Pathol Lab Med. 2016 05;140(5):473-6.
3. Jung Woo Choi, Young Seok Lee, Ju Han Lee, Han Kyeom Kim, Bom Woo Yeom, Jong Sang Choi, Hong Chul Lim, Chul Hwan Kim. Liposclerosing Myxofibrous Tumor in Tibia: A Case Report and Review of the Literature.. Korean Journal of Pathology. 2005;39(3):207.

Figures

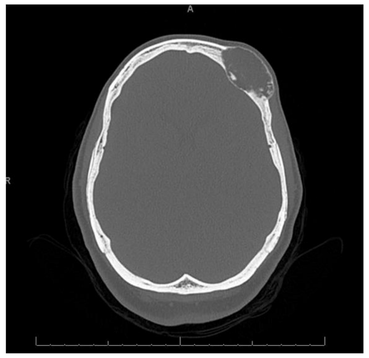

Figure 1. Axial CT head demonstrating a mixed density ldiploic space lesion of the left frontal bone. Note thinning of the inner and outer tables, scalloping of the endosteum with marginal sclerosis, and scattered peripheral and ossific densities

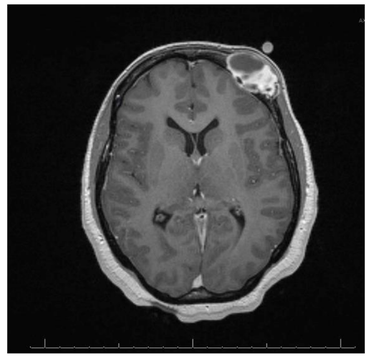

Figure 2: Axial T1 brain MRI with post gadolinium administration demonstrating a heterogeneously enhancing ldiploic space lesion.

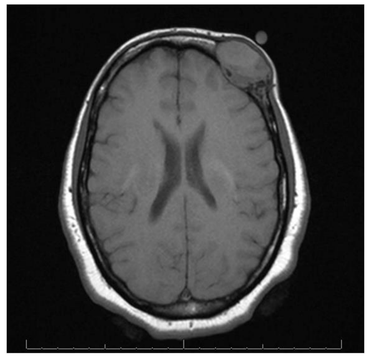

Figure 3: Axial T1 brain MRI without gadolinium demonstrating a diploic space lesion with expansion of the inner and outer tables of the skull

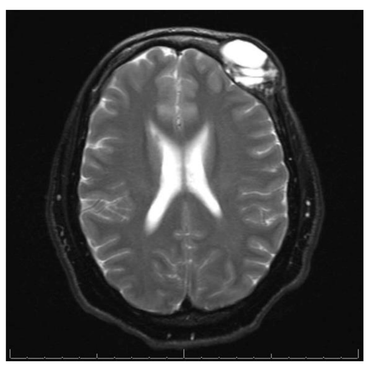

Figure 4: Axial T2 MRI brain demonstrating a heterogeneous lesion with both cystic and solid components

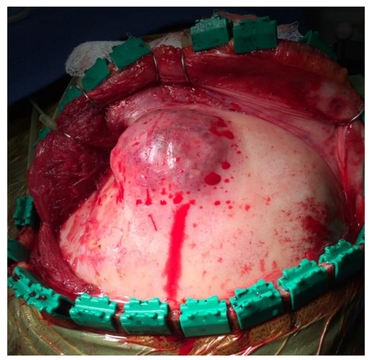

Figure 5: . Intraoperative image of the lesion showing the thin mantle of bone and the underlying bluish hue of the lesion

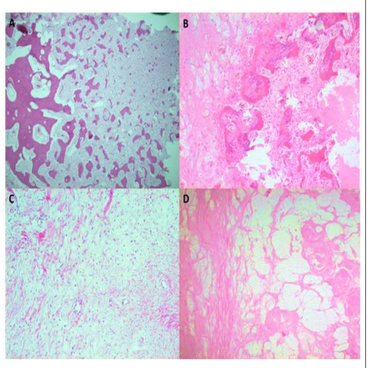

Figure 6: H&E staining. A) central fibrous dysplasia-like area with peripheral native cortical bone. B&C) Cystrophic calcifications and myxoid areas, respectively. D) Areas of cystic degeneration and lipomatous differentiation.

Publication

*This case report was accepted for publication in *Neurosurgery*, March 2018.

Case Report

A 30-year-old female was referred to the neurosurgical clinic for a two-year history of an expanding, painless left frontal bone growth. She denied neurologic symptoms. On physical exam, she did not have focal neurologic deficit or facial weakness. There was a visible left forehead mass that was firm and painless. The overlying scalp appeared normal and freely mobile.

A head CT scan demonstrated an expansile, mixed density lesion centered in the left frontal bone diploic space measuring 4.4 x 2.3 x 4.8 cm (Fig. 1). The lesion showed thinning of the inner and outer tables, scalloping of the endosteum and scattered peripheral calcific and ossific densities (Fig. 1). MRI of the brain with and without gadolinium identified a large, solitary, well-demarcated heterogeneously enhancing lesion in the diploic space (Fig. 2-4). The lesion was partially cystic with expansion of both the inner and outer tables.

She was taken to the operating room for resection of the lesion. An incision was made through all the layers of the scalp over normal appearing bone and reflected in a single layer over the mass. The periosteum appeared uninvolved with the mass. Superficially we observed a very thin mantle of bone that was soft and depressible with an underlying bluish hue (Fig. 5). A craniotomy was used to cut circumferentially around the lesion, and it was removed in *en bloc*. There were some residual remnants of abnormal yellow tissue over the orbital roof and this was drilled until normal bone appeared. The bony defect was reconstructed with a polyetheretherketone synthetic plate that was manufactured prior to surgery based on the pre-operative thin slice CT scan.

Histological examination showed a fibro-osseous lesion with a variety of patterns including myxoid and fibrous tissue with areas of cystic degeneration, curvilinear woven bone with and without osteoblastic rimming, areas of lipomatous differentiation and foci of dystrophic calcification (Fig. 6). This was consistent with liposclerosing myxofibrous tumor.

This patient did well post-operatively. She was discharged home in good condition on post-operative day four and returned to work within six weeks of surgery. At her eight-month routine follow up evaluation, she had complete function of all activities of daily living and was working full-time. She had no evidence of recurrence on imaging at this visit. She will continue to be followed with annual surveillance scans.

Summary and Conclusion

LSMFT is most commonly identified in the intertrochanteric region of the femur. There have been reports of this pathology found in other bony areas, but upon review of the literature we did not find cases of LSMFT identified within the calvarium. Diagnosis of this rare, benign lesion depends on the combination of clinical course, radiographic features, and histologic appearance. Location can be helpful in guiding diagnosis, however, LSMFT should not be ruled out when the location is unusual. Biopsy is ultimately needed for definitive diagnosis.