

Myxoma of the Frontal Bone

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Abstract

Primary myxoma of skull bone is a rare benign tumor. A 34-year-old female presented with diffuse tender swelling in the right frontal bone. After investigations, she underwent craniectomy and excision of mass. Histopathology and immunochemistry confirmed it to be a primary myxoma.

Keywords: Myxoma; Frontal Bone; Bone Destruction

Introduction

Myxoma are benign tumors that arise from mesenchymal tissues located anywhere in the body. It most commonly arises from heart, skin, bone and subcutaneous tissue [1]. Myxoma of the head and neck region are rare, occurring more commonly in the bones than in soft tissues [2]. In skull myxoma usually arise from skull base bones especially mastoid, sphenoid or ethmoid. Frontal convexity involvement has not been reported in the literature, hence being reported [3-5].

Case Report

A 34-year-old female presented with a diffuse bulge over the right frontal region of 1-month duration and giddiness of 15 days' duration. There was no history of fever, trauma or decreased appetite. On examination there was diffuse bulge, tender with ill-defined margins and firm in consistency. Computed tomography (CT) revealed a lytic lesion in the frontal bone (Figure 1a). Magnetic resonance imaging (MRI) revealed an isointense mass on T1WI and hyperintense on T2WI (Figure 1b and c) X-Ray skull done in this hospital revealed irregular osteolytic lesion in the right frontal bone (Figure 1d). Echocardiography was normal.



Figure 1(a) (b) (c) (d)

Figure 1 (a): Pre-Op Contrast Enhanced CT Scan Showing Lytic Lesion in The Right Frontal Bone.

Figure 1 (b and c): MRI Showing Bony Lesion with Hypointense Signal on T1WI (B) and Hyperintense Signal on T2WI (C).

Figure 1 (d): X-Ray Skull Lateral View Showing Irregular Osteolytic Lesion in the Right Frontal Bone.

Craniectomy around the lesion revealed a moderately vascular mass involving the bone and indenting the dura. Lesion was removed en mass. While separating the tumor from underlying dura, it got breached and was repaired primarily with pericranium. Histopathology revealed stellate cells scattered with in myxoid matrix and bone spicules. (Figure 2a,2b). There was no evidence of mitosis and necrosis. Immunochemistry was positive for Vimentin and negative for keratin, confirming the diagnosis of myxoma. CT scan in the follow up period showed no evidence of residual bone destruction (Figure 2c).

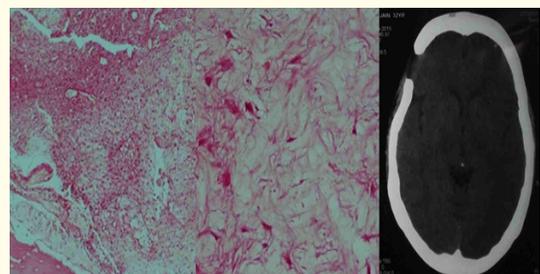


Figure 2(a) (b) (c)

Figure 2 (a): Section Showing Bony Trabeculae with Stellate Tumor Cells with Myxoid Matrix (H and E, X 100)..

Figure 2 (b): Vimentin Positive Stellate Cells (Vimentin, X 250).

Figure 2 (c): Post-op CT Scan Showing no Residual Disease.

Discussion

Myxoma commonly arise from the mesenchymal tissue of the heart and may involve neural tissue by thromboembolic phenomenon [6,7]. However rarely it may arise primarily from bone and sinuses of skull.

Usually the skull bone and sutures are involved and involvement of convexity is seen rarely. In all cases echo must be done to rule out possibility of primarily atrial involvement. Usually the

tumor develops in the marrow, expands the cortex and destroys the bone by aseptic pressure necrosis [1]. This was evidenced by our case as well. In our case soft tissue mass was visible under the area of bony erosion mimicking a granuloma or meningioma.

As myxoma is a benign lesion, preferable treatment is complete excision which may not be feasible in the cases involving skull base or vital structures and recurrence may require redo as radiation has no effect on the tumor. These tumors are common in third and fourth decade and may occur in any sex. Differential diagnosis includes myxomatous meningioma, sarcoma with myxoid degeneration and metastatic myxoma. Immunohistochemistry is helpful in distinguishing these lesions. True myxoma will stain positive for vimentin and negative for cytokeratin and S-100 as seen in our patient as well. Myxomas are reported as hypodense to isodense with variable enhancement.

Pathologically the neoplasm is composed of stellate cells set in loose mucoïd stroma [8]. Cranial base erosions can be extensive and the gelatinous nature of these lesions compromises radical operative treatment especially if they cannot be resected en bloc with surrounding tissue. Recurrence rate is reported to be 25% if they cannot be removed radically [9]. Osterdock, et al used preoperative embolization because of extensive vascularity of the tumor in their case [10].

The etiology of the primary temporal bone myxoma which is most commonly involved in skull remains unclear, but it is thought that it arises from primitive mesenchyme which regresses when pneumatization is complete [11]. Recurrence is most common within the first 2 years after excision making imaging and clinical surveillance of paramount importance.

Conclusion

Myxoma of the frontal bone though rare should be kept as a remote possibility in differential diagnosis in an expansile/osteolytic well circumscribed enhancing mass lesion in a young adult. An attempt should be made to excise it totally as radiotherapy has no role in its management.

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