CASE REPORT

GIANT HEMANGIOMA OF FACE AND SCALP WITH INTRACRANIAL AND INTRA ORBITAL EXTENSIONS: A RARE CASE REPORT

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HOW TO CITE THIS ARTICLE:

ABSTRACT: Hemangiomas are one of the commonest soft tissue benign neoplasms, of vascular origin. They are commoner in pediatric and adolescent age groups. They are known to affect any part of the body. Giant hemangiomas are an uncommon entity. We present a case of giant hemangioma involving face and scalp extending intracranially and intra orbitally in a female geriatric patient who presented with a long standing gradually progressive painless swelling over face and scalp with proptosis. Our case is remarkable for presentation in geriatric age group with extensive intracranial and intra orbital involvements.

KEYWORDS: Giant Hemangioma, Face, Scalp, Intracranial, Intraorbital.

INTRODUCTION: Hemangiomas are one of the commonest soft tissue benign vascular neoplasms, known to affect any part of the body. Primary hemangiomas of the skull are benign lesions manifested as palpable masses or incidentally detected during an imaging evaluation. We present a geriatric female patient presenting with a long standing progressive swelling who showed extensive involvement of face and scalp by a giant hemangioma. She was evaluated with multimodality imaging workup and the typical appearances in various modalities were described.

CASE REPORT: A 67 years female presented with a long standing swelling in right lateral aspect of head and face (Fig. 1). It started as a small swelling few years before which increased in size to the present state. It was painless but heavy. There was proptosis of right eye with bogginess of right temporal region.

On general examination vitals were normal. Her systemic examination was within normal limits.

Local examination revealed a soft swelling which was compressible and warm. It was non tender. Axial proptosis of right eye was noted with anterior and inferior displacement. The swelling extended superiorly till the vertex and inferiorly upto the temporal and zygomatic region. Her laboratory tests were within normal limits.

Plain radiograph of skull- lateral view (Fig. 2) revealed soft tissue dense lytic lesion involving the parietal bone, predominantly the outer cortex and extending upto vertex. Lobulated soft tissue dense lesion was noted over the skull bones.

High resolution ultrasonography with Doppler interrogation using linear probe (Fig. 3) showed heterogeneous predominantly hyperechoic lesion showing significant internal vascularity. Augmentation on valsalva maneuver was noted. Its extensions were as described below.

Plain and contrast CT of head and brain (Fig. 4) with volume rendered images (Fig. 5) was performed which showed significantly enhancing soft tissue attenuation lesion involving right half of scalp and face. There was involvement of skull with erosion of outer cortex and diploic spaces. Bone window images showed (Fig. 6) focal areas of breech in inner cortex with intracranial extension.
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Typical radiating bony spicules were noted at places. However obvious involvement of brain was not appreciated. Also noted were extension of the lesion into sellar, right parasellar, suprasellar, right ethmoidal sinus, bilateral sphenoidal sinuses, temporal and infratemporal regions. There was near complete involvement of intra and extracranal spaces of right orbit with resultant proptosis. Histopathological examination confirmed the diagnosis of hemangioma.

DISCUSSION: Hemangiomas are one of the commonest soft tissue benign neoplasms of vascular origin. They comprise ~ 2% of all benign and 0.8% of all benign and malignant lesions of the skeletal system. They are commoner in pediatric and adolescent age groups. They are more common in women by a 3:1 ratio and are found in all age ranges. They are known to affect any part of the body.

Hemangiomas are classified histologically, according to the type of vessels in the lesion as capillary, cavernous, arterio-venous or venous.

Hemangiomas of the skull represent 0.2% of all osseous tumors and 10% of all the benign tumors of the skull. Hemangiomas arise from the intrinsic vasculature of the bone and from the diploic spaces of the skull.

Primary hemangiomas of the skull are benign lesions manifested as palpable masses or incidentally detected during an imaging evaluation. Hemangiomas can be found in any location in the skull, but they are most often seen in the frontal and parietal regions. They occur less frequently within the occipital, temporal, sphenoid, and petrous bones. Fifteen percent of skull hemangiomas are multiple. The usual clinical presentation is that of a painless, palpable scalp mass. A bruit is uncommon. Trauma may be an antecedent factor, although this is not always elicited in the patient's history.

On gross inspection, the tumor is fleshy and vascular. Prominent scalloping in the inner or outer table may be seen. Extension of the tumor into the soft tissues also may be seen. Microscopically, multiple engorged vascular elements are found interspersed among abundant trabecular bone. The vessels are thin walled and lined by a single endothelial layer.

Plain radiography is often the initial technique used and it is useful for showing the location of lesion and bony erosions caused by it. They have a characteristic sunburst or honey comb radiating spicules from a round to oval defect. The outer table is usually involved with relative sparing of inner table. The margins are non-sclerotic. Ultrasonography is useful in assessing the vascularity and extracranial extensions and involvements. CT is optimal for characterizing the bony involvement of the lesion and for evaluation of the lesion matrix.

The characteristic radiating spicules with the scalloped non sclerotic margins are better appreciated. Hemangiomas are usually isodense soft tissue attenuation masses enhancing homogenously and brilliantly on contrast administration helping in delineation of exact extent. MR imaging with its multiplanar capabilities and better soft tissue resolution is suited for knowing extensions of the lesion.

It is however inferior to CT with respect to assessing cortical margins and bony destruction or erosion. Intradiploic extension, intracranial extension, and involvement of adjacent neurovascular structures are more readily detected with MR imaging. It appears hyperintense on T1 and T2 images with intense enhancement on contrast administration. Angiography assist in the presurgical evaluation and possible embolization of the tumor.
CONCLUSIONS: Hemangiomas are common tumors in day to day practice and form differential
diagnosis for most masses in head. Multimodality approach for their diagnosis helps in complete
evaluation of extension and involvements.

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Fig. 1(A & B): Photograph of the patient showing large soft tissue mass involving scalp and right
orbit causing proptosis.
Fig 2: Lateral skull radiograph showing large lytic soft tissue mass involving parietal bone.
Fig. 3 (A & B): Ultrasonography with Doppler showing heterogeneous hyper echoic lesion with significant vascularity.

Fig. 4 (A & B): Axial contrast enhanced CT showing heterogeneous intensely enhancing lesion causing lytic destruction of outer table of skull and extending to orbit causing proptosis

Fig. 5: Volume rendered image showing soft tissue lesion involving parietal and right temporal regions of skull and right orbit.
Fig. 6: Axial CT (Bone window) showing extensive lytic destruction of outer table of skull with radiating spicules, focal areas of breech in inner cortex with intracranial extension.

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FINANCIAL OR OTHER COMPETING INTERESTS: None

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Date of Submission: 13/05/2015.
Date of Peer Review: 14/05/2015.
Date of Acceptance: 28/05/2015.
Date of Publishing: 04/06/2015.