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The Diagnostic Role of Ultrasound and MRI with IHC in a Rare Case of Epithelioid Hemangioendothelioma of Soft Tissue and Management

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Abstract:- Epithelioid Hemangioendothelioma (EH) is a rare malignant vascular tumour that is considered to be intermediate grade between benign Hemangioma and malignant Angiosarcoma originates from vascular endothelial or Pre endothelial cells. EH can occur anywhere in the body most commonly affects the Liver, Lungs, Bones, and although involves the Pleura, Mediastinum, Spleen, Skin, Breast, Head and neck area, Brain and Meninges and Lymph nodes. It often involves either superficial or deep soft tissue, visceral organs and less commonly, medium-size or large veins. The aetiology is unknown and is usually diagnosed at young adult, being rare in children. EH is locally aggressive, heterogenous and represents less than 1% of all the vascular tumours and capable of metastasis. Prevalence is 1 in 1 million.

A 45Years old male presented with soft to firm subcutaneous swelling since 7years later it progressed rapidly in size from 1year in the Right upper anterior chest. On examination: A non-tender, multilobulated exophytic lesion in the subcutaneous plane showing few erythematous nodules of varying size and were firm in consistency. High frequency Ultrasound.(US) of Right Supraclavicular Mass, and MRI Thorax modalities suggest features of Supraclavicular soft tissue neoplasm showing vascular components and concern for Hemangioendothelioma.

Management includes wide excision of the soft tissue lesion in the supraclavicular region and followed by excisional biopsy and histopathological confirmation. US and most sensitiveMagnetic Resonance Imaging(MRI) modality features along with pathological techniques of Histopathology and Immunohistochemistry(IHC) techniques confirms the vascular nature of tumour. Followed by the wide excision the patient has undergone adjuvant radiation therapy to decrease the risk of local recurrence and distant metastasis.

Keywords:- *CT Computed Tomography, Epithelioid Hemangioendothelioma, Immunohistochemistry, Magnetic Resonance Imaging, Out Patient Department, T1 Weighted Images, T2 Weighted Images, Ultrasound.*

I. INTRODUCTION

The term "EH" was proposed by Weiss and Enzinger, to explain a category ofsoft tissue vascular tumours composed of epithelioid appearance endothelial cells with intermediate clinical course between benign hemangioma and malignant angiosarcoma[1].

EH, which is a rare vascular tumour with an epithelioid and histiocytoid appearance, originating from vascular endothelial or pre-endothelial cells[2].

It has a prevalence of 1 in a million . It is often misdiagnosed and not suitably treated leading to a poor prognosis in many cases[6]. It usually affects middle-aged patients, although cases in children and elderly people.

Moreover, many patients are asymptomatic at the time of diagnosis[4].

The aetiology of EH remains a dilemma. At the molecular level, various angiogenic stimulators may act as promoters of endothelial cell proliferation. Recently it had been reported that monocyte chemo-attractant protein-1 is required for EH proliferation and might promote the event of those lesions by stimulating the angiogenic behaviour of endothelial cells[3].

It occurs in any a region of the body are often affected, but the foremost common sites are liver alone 21%, liver plus lungs 18%, lungs alone 12%, and bone alone 14%[5] but the prognosis on visceral organs EH is worse.

The definitive diagnosis of EH requires Histopathological correlation. The pattern of solid growth and thus the epithelioid appearance of the endothelium frequently leads to the mistaken diagnosis of metastatic carcinoma. The tumour are often distinguished from a carcinoma by the shortage of pleomorphism and mitotic

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activity in most instances and by the presence of focal vascular channels[1].

II. CASE PRESENTATION

2.1 Clinical History

A 45year old male presented to Surgery OPD, with soft to firm subcutaneous swelling since 7years later it progressed rapidly in size from 1year in the right upper anterior chest with no history of pain.

On physical examination showed non-tender, multilobulated exophytic lesion in the subcutaneous plane showed erythematous nodules of varying size and were firm in consistency(Fig. 1). Cutaneous changes over the lesion noted.

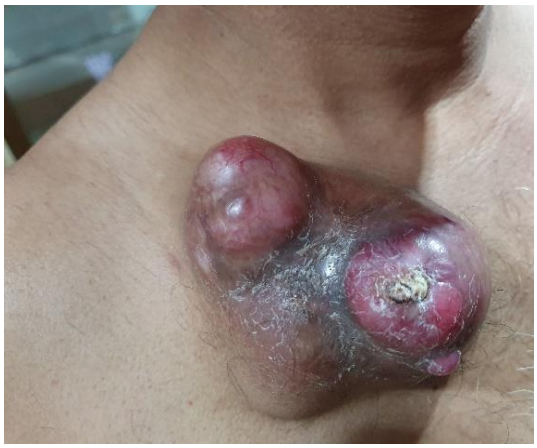
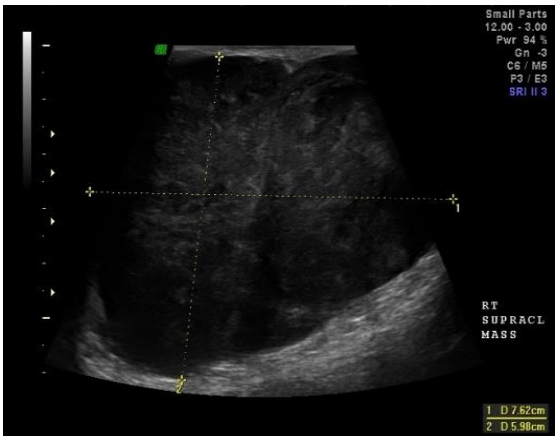
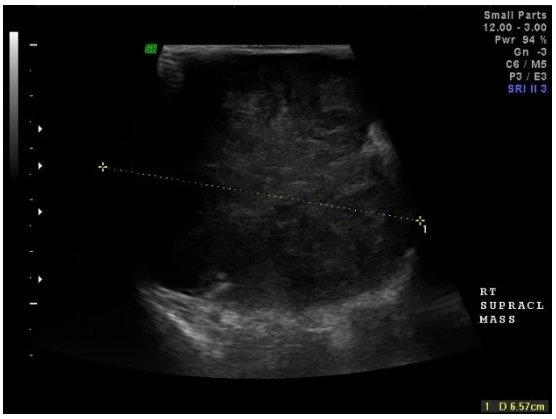


Fig. 1:Multilobulated exophytic lesion in the subcutaneous plane of Right Supraclavicular region.

2.2 On High frequency Ultrasound of Right Supraclavicular Region showed well defined Lobulated nodular heterogenous predominantly hypoechoic subcutaneous soft tissue mass noted anteriorly to and about the right clavicle extending to the skin surface measures about 7.6x5.9x6.6cm(Fig. 2).There was no evidence of infiltration of the lesion into the underlying pectoral muscles.



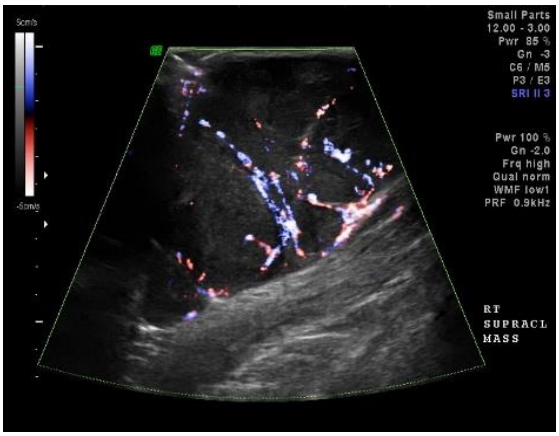
(a)



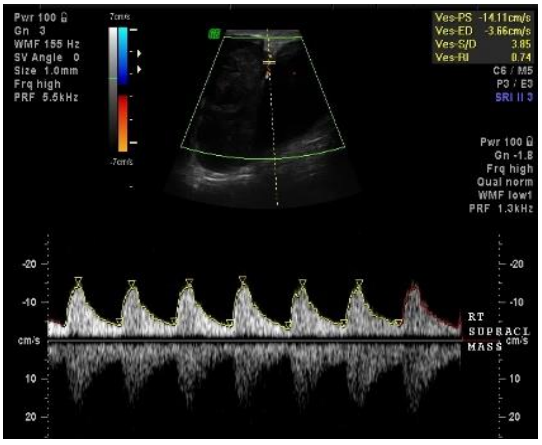
(b)

Fig. 2 (a) and (b) Grey scale Ultrasound images showing lobulated soft tissue mass anteriorly to and about the right clavicle.

2.3 On Colour doppler, lesion was hypervascular showed low resistance flow in the arteries. Veins showed normal flows. No admixture flow are seen in Fig. 3.US revealed the features of differentials of Hemangioendothelioma or Soft tissue Sarcoma, however in view of lack of infiltration of underlying muscle planes,Sarcoma was unlikely. However there was no admixture flow hence angiosarcoma was ruled out.



(a)



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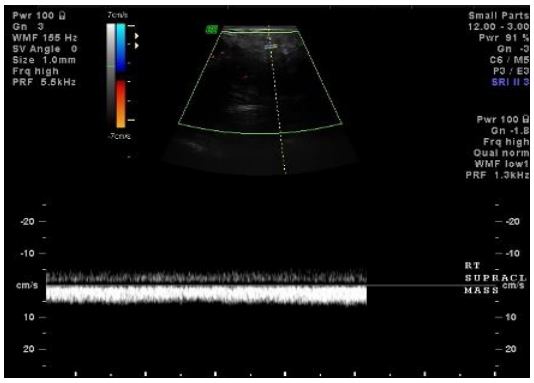


(c)

Fig. 3(a), (b) and (c): Colour Doppler images of Right Supraclavicular mass showing hypervascular lesion with no admixture flow.

2.4 On Computed Tomography(CT) Thorax(Plain) showed well defined soft tissue attenuated lesion was seen in Fig. 4 involving the right supraclavicular region extending to the skin surface. No evidence of calcification within the lesion.

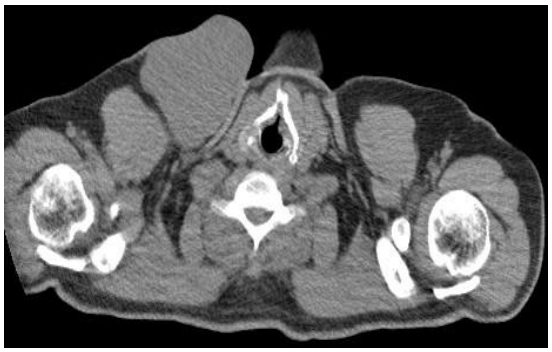
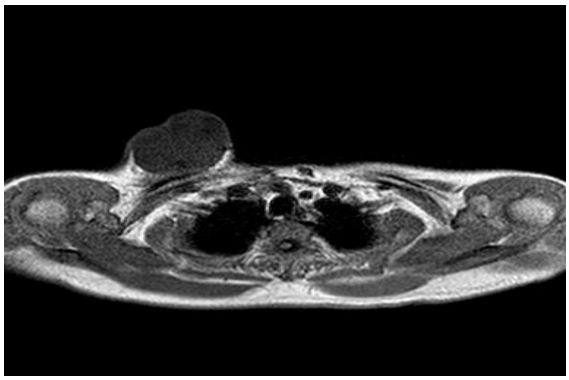


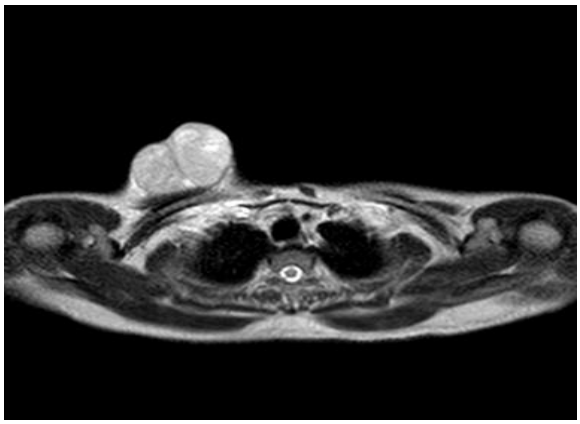
Fig. 4 : CT Thorax showing no calcification within the lesion.

2.5 On MRI Thorax with Contrast

On T1 Weighted Images(T1WI) the lesion was seen in Fig. 5(a) a large well defined multilobulated Hypointense to the muscle and in Fig. 5(b) on T2 Weighted Images(T2WI) there was central hyperintense with peripheral thin hypointense rim measures 66 x 63 x 77mm (AP x TR x CC) which was anteriorly extending into the skin surface and posteriorly lies anterosuperior to the underlying pectoralis major muscle and medial one third of the clavicle.



(a)

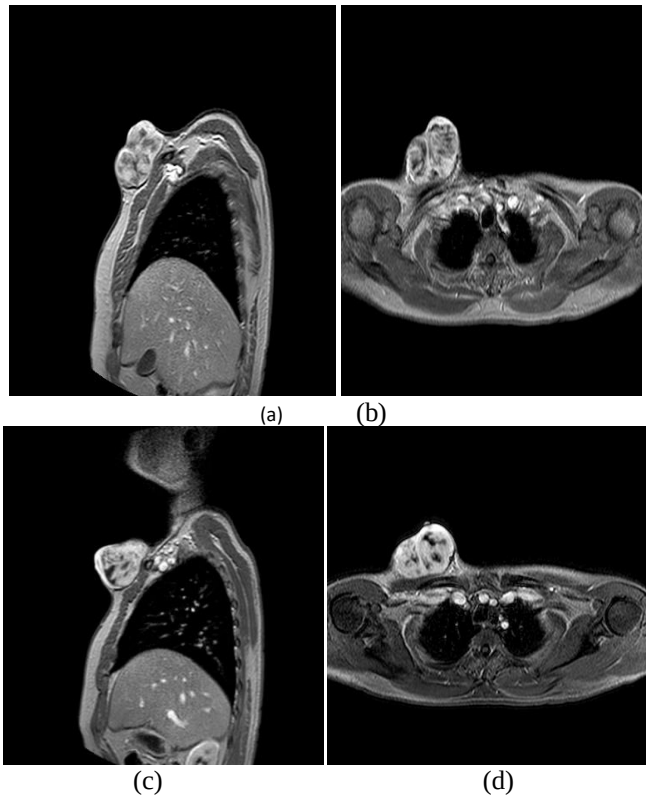


(b)

Fig. 5: MRI THORAX showing (a) Hypointense lesion on T1 WI and (b) Hyperintense on T2 WI with no extension into the underlying structures clavicle and pectoralis major muscle.

On Post-contrast T1WI the lesion showed "Target Pattern" of hypointense centrally with thick enhancing inner peripheral rim and thin non enhancing outer peripheral rim(Fig.6).

Tubular like hypointense are noted within the lesion on Post Contrast T1WI suggestive of Intratumoral flow voids (Fig.6).



(a) (b)

(c) (d)

Fig. 6(a), (b), (c) and (d) : MRI THORAX of Axial and Sagittal sections of Post-contrast T1 Weighted Images showing Non homogenous enhancement, "Target Pattern" of hypointense centrally with thick enhancing inner peripheral rim and thin non enhancing outer peripheral rim with intratumoral flow voids.

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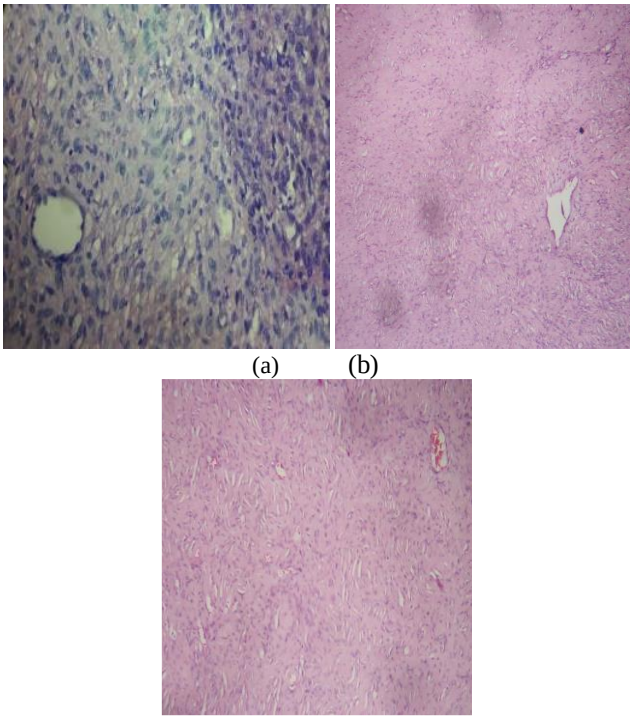
No evidence of extension of lesion into the underlying structures clavicle and pectoralis major muscle. Lung parenchyma appeared normal without mass lesion or infiltration. There was no mediastinal mass and mediastinal or hilar lymphadenopathy.

2.6 Histopathological findings:

After wide excision of a tumour the sample sent to the HPE which showed the deep dermis were comprised of epithelial and endothelial cells arranged in cords and small nest pattern. Individual tumour cells shows abundant eosinophilic cytoplasm with some intracytoplasmic vacuolesFig. 7. All margins of the excised lesion was free from tumour deposits. The features were suggestive of EH of Anterior Chest wall.

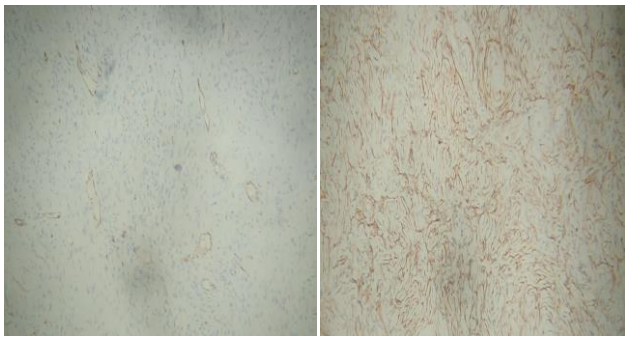


(a) (b)

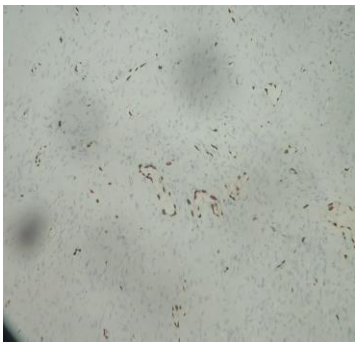
(c)

Fig. 7(a), (b) and (c): Histopathological slides of the obtained tissue from the wide excision of lesion.

2.7 IHC : The cells were immunoreactive for CD31, CD34 endothelial markers and ERG revealed the endothelial nature of the cells(Fig. 8).



(a) (b)



(c)

Fig. 9(a), (b) and (c) : Immunohistochemical staining shows tumour cells express CD31, CD34 and ERG.

After the 2 weeks of wide excision of the supraclavicular soft tissue the patient was referred to the radiotherapy unit and advised to take the local radiotherapy to reduce the risk of local recurrence after the primary surgery and the patient underwent the PET CT scan in which there was no uptake of FDG in the previously operated right supraclavicular region, suggestive of no residual tumour with no distant metastasis. Then Patient underwent the adjuvant radiation therapy to decrease the risk of local recurrence.

III. DISCUSSION

EH may be a rare vascular tumour with an epithelioid and histiocytoid appearance, originating from vascular endothelial or pre-endothelial cells[2]. Epithelioid hemangioendothelioma of soft tissue is most often a solitary lesion, in either the superficial or deep tissue with uncertain behaviour and prognosis.

The tumour impacts on both sexes equally and no predisposing factors are recognized. The neoplasm usually presents as solitary, rarely multiple, slightly painful erythematous papules, nodules, plaque and nonhealing ulcer[7].

Although epithelioid hemangioendothelioma is capable of causing regional and distant metastasis, it does thus far less frequently than the typical angiosarcoma. In a recent study the rate of metastases in epithelioid hemangioendothelioma is found to be 22%[8].

Because of its rarity, EH has no standard treatment. The available treatment options are surgical resection, adjuvant chemotherapy and/or radiotherapy[8],[9]. Histopathological examination remains the mainstay of diagnosis for this rare tumour. The final diagnosis to be assisted by the use of immunocytochemical techniques. The more commonly used antisera are CD 31, CD34, ERG and factor VIII-related antigen. Radiotherapy after surgical resection is chosen for localized EH to control residual disease[8],[9].

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The prognosis of EH is uncertain as the mortality rate for EH of the liver is 35% and lung is 65%. It would seem that the prognosis in primary cutaneous lesions may be good[7].

In diagnosing the tumour in this case, ultrasonography demonstrates tumour characterisation and vascularity which helps in making differential diagnosis. Hemangioendothelioma / Soft tissue Sarcoma/Angiosarcoma, however in view of lack of infiltration of underlying muscle planes, Sarcoma is unlikely. There was no admixture flow oncolour doppler, Angiosarcoma had been ruled out. Suggest features of Supraclavicular soft tissue neoplasm showing vascular components and concern for Hemangioendothelioma. Magnetic Resonance Imaging(MRI) Thorax of contrast modality has major role in diagnosing epithelioid hemangioendothelioma showing the characteristic Target pattern of the lesion with minimal delayed capsule enhancement and intralesional flow voids.

However CT Thorax is done as the ancillary to the MRI has the MRI is most sensitive modality to evaluate the soft tissue structures and the soft tissue lesions to delineate its characteristics by the extent of the lesion and its relationship to the adjacent structures and provides an excellent contrast resolution.

The patient underwent the wide excision of the right supraclavicular lesion with skin grafting and the sample sent to the HPE showing deep dermis are comprised of epithelial and endothelial cells with abundant eosinophilic cytoplasm with some intracytoplasmic vacuoles consistent with Epithelioid Hemangioendothelioma and was confirmed by IHC in which tumour cells were immunoreactive for CD31, CD34 endothelial markers and ERG, revealed the endothelial nature of the cells.

Wide excision of the supraclavicular soft tissue was done and after 2 weeks the patient was referred to the radiotherapy unit and advised to take the local radiotherapy to reduce the risk of local recurrence after the primary surgery and the patient underwent the PET CT scan in which there was no uptake of FDG in the previously operated right supraclavicular region suggestive of no residual tumour with no distant metastasis. Then Patient underwent the adjuvant radiation therapy to decrease the risk of local recurrence.

IV. CONCLUSION

The importance of distinguishing epithelioid vascular tumor on suspecting malignant epithelioid vascular tumor on imaging and further characterization into subgroups, using Ultrasound and most sensitive MRI modality features are confirmed with pathological techniques of Histopathology and Immunohistochemistry and to arrive at a confirmation of diagnosis followed which patient underwent the wide excision with adjuvant radiation therapy to decrease the risk of local recurrence and distant metastasis.

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