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CASE REPORT

EPITHELIOID HEMANGIOENDOTHELIOMA OF CHEST WALL: CASE REPORT

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ABSTRACT

Epithelioid hemangioendothelioma (EHE) is a rare vascular tumor, first described by Sharon Weiss and Franz Enzinger in 1982 as an intermediate condition between hemangioma (benign) and angiosarcoma (malignant) both clinically and pathologically. However, with the advent of cytogenetics, EHE is now considered as a distinct entity with distinct disease defining genetic alteration. Because of its rarity and varied anatomical locations, EHE has got no standard treatment plan. With the treatment options being surgical resection, chemotherapy, radiotherapy, measures inhibiting angiogenesis, etc. We report a case of chest wall epithelioid hemangioendothelioma, a solitary lesion with easy accessibility for curative resection, making it the best treatment option for the same.

Keywords: Epithelioid hemangioendothelioma, Chest Wall

1.INTRODUCTION

EHE was originally described as vascular tumor arising from endothelial cells most commonly in the small veins of the extremities (upper and lower limbs) and the liver and lungs. It has since been described in organs throughout the body.

OTHER SITES OF EPITHELIOID HEMANGIOENDOTHELIOMA[3].

Head and neck	Breast
Lymph nodes	Mediastinum
Diaphragm	Brain and meninges
Mastoid	Infundibulum
Clivus	Cerebellopontine angle
Spine	Skin
Peritoneum	Stomach
Retroperitoneum	Ovary
Prostate	Eyelid

We report a case of EHE arising from chest wall, a rare site for a rare tumor and its review of literature.

2.CASE REPORT:

A 82 years old male presented with swelling over right scapular region for 9 years, which was insidious in onset and gradually progressing. But following a trauma to the same, 1 year back it has rapidly increased in size. His past history was unremarkable except for excision of similar swelling at 10 yrs of age over right lumbar region. On examination, he had a single huge non compressible swelling of size 40 * 30 cm over right scapular region. It was a freely mobile, fluctuant swelling with mixed consistency (predominantly cystic) and presented with skin excoriations (Fig 1). Clinically, soft tissue sarcoma was suspected and magnetic resonance imaging was done. Fine Needle Aspiration Cytology was done for tissue diagnosis.

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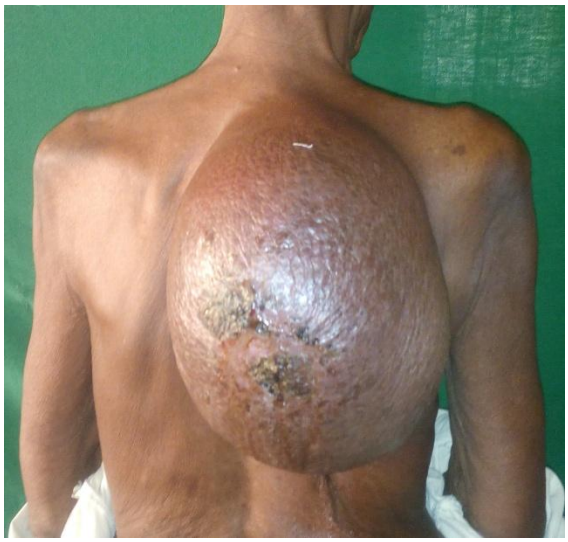

Fig 1: Pre operative image

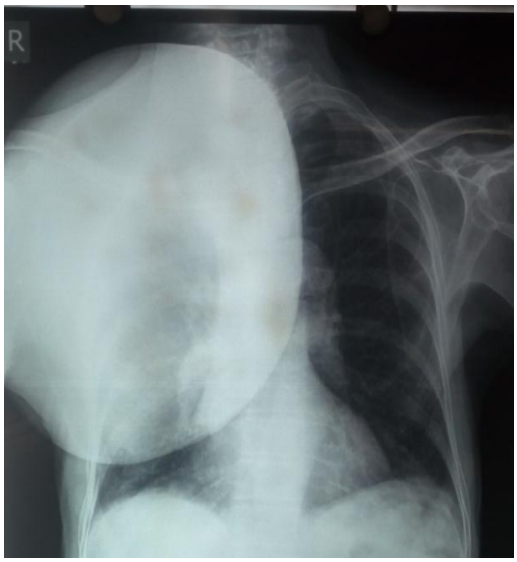

Fig 2: X-ray image

MRI showed a well encapsulated, heterogenous, predominantly cystic lesion with multiple internal septations, eccentric fatty components and right trapezial infiltration. Metastatic imaging work up turned out to be negative. FNAC was inconclusive. Wide local excision was done including the tumor with 1.5 litres of hemorrhagic fluid within it and the involved skin and part of right trapezius muscle. Post operative recovery was uneventful. Patient was in follow up for 6 months without any signs of recurrence or metastasis.

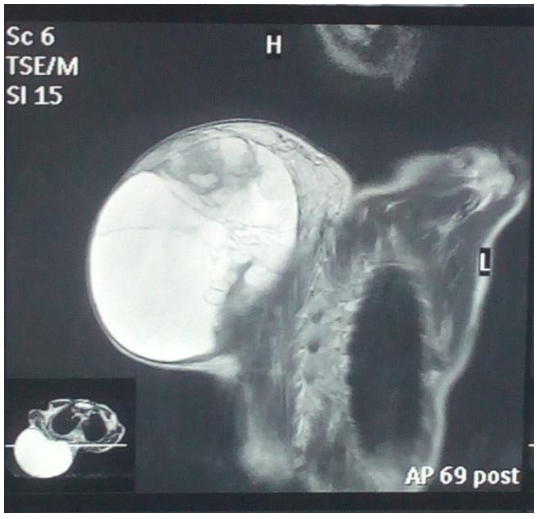

Fig 3: MRI image

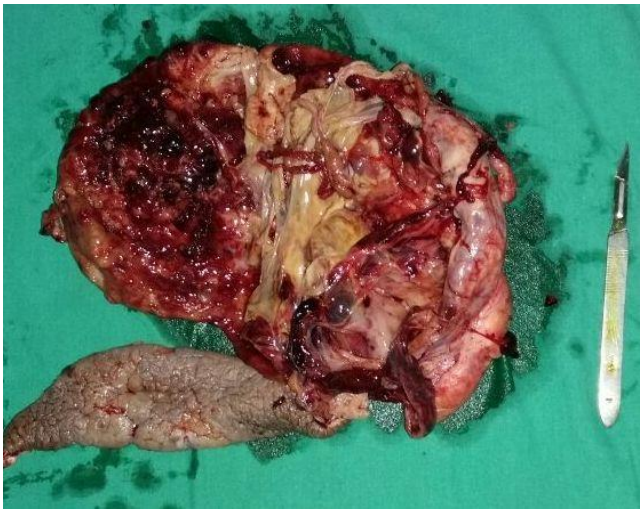

Fig 4: Post operative specimen

3.DISCUSSION:

Epithelioid hemangioendothelioma is a rare vascular tumor with an epithelioid and histiocytoid appearance, originating from vascular endothelial or pre-endothelial cells. It represents less than 1% of all vascular tumors and was described for the first time in 1975 by Dail and Liebow as pulmonary EHE (P-EHE). The term "epithelioid hemangioendothelioma" was introduced in 1982 by Weiss and Enzinger to describe a vascular tumor of bone and soft tissue showing features between hemangioma and iosarcoma (Epithelioid hemangioendothelioma,2014).

The recent World Health Organization (WHO 2002) classification describes EHE as lesions that fall into the category of locally aggressive tumors with metastatic potential[6][7]. The estimated prevalence of EHE is less than one in 1 million(Epithelioid hemangioendothelioma,2014). According to (WHO) classification of Tumors of bone and soft tissues (2002), one half to two thirds of them originates from a vessel (such as a small vein). Exceptionally, it may originate from a large vein or artery.

ETIOPATHOGENESIS: Many things have been speculated as potential causes of EHE, including hormonal effects, oral

contraceptive pills, industrial exposures (vinyl chloride) and trauma among others. None of these associations has been confirmed. Fortunately, significant progress has been made very recently. Two separate research groups published findings in late 2011 indicating a specific chromosomal translocation that is virtually diagnostic for EHE. This swapping of genetic material between chromosomes 1 and 3 is seen in almost all EHE tumor cells and can be considered 'disease defining. This genetic alteration results in the production of a fusion gene and a resulting fusion protein. It has part of a protein that is normally seen only in endothelial cells and a other part seen only in brain. This abnormal fusion protein presumably causes the cells to grow without normal regulation.

A new, suggestive hypothesis of the pathogenesis of this disease refers to a causal relationship between chronic *Bartonella* infection and the development of this rare vascular tumor. The unique Bartonella's capability of invading and inducing long-lasting intraerythrocytic and intraendothelial infections, together with the ability of at least three *Bartonella* spp. (*B. henselae*, *B. quintana*, *B. bacilliformis*) of inducing vascular endothelial growth factor-mediated vasoproliferation as they upregulate mitogen and proinflammatory genes resulting in cytoskeletal rearrangement and suppression of endothelial cell apoptosis, suggest that these bacterial pathogens might contribute to the development of vascular tumors(Mascarelli et al., 2011).

CLINICAL FEATURES:

The clinical presentation of this rare vascular tumor is as heterogeneous as its clinical localization can be:

SITE	CLINICAL FEATURES	TREATMENT
FOREHEAD	Nodular skin leision, Visual disturbance	Surgery
ABDOMINAL WALL	Swelling	Not done
MASTOID	Hearing loss, Vertigo	Surgery + RT
RETROPERITONEUM	Abdominal discomfort	Surgery
PROSTATE	Perineal pain, night sweats, rectal fullness, urgency	Surgery
OVARY	Pelvic pain	Surgery
EYELID	Swelling	Surgery
FOREARM	Mass, pain, weakness	Surgery
LIVER	Altered blood count	Liver transplantation
PLEURA	Pleural effusion, chylous ascites	Chemo (carboplatin + etoposide)

HISTOLOGY: When EHE was identified, electron microscopy showed a typical image of endothelial cells similar to those composing medium-size vessels or a large vein, arranging in nests or cords, while immunohistochemistry revealed Weibel-Palade bodies in the cytoplasm of their cells as a typical finding[9]. A variety of endothelial proteins may be useful to identify EHE. The *Fli-1* protein is expressed by the endothelium as well as the T-cells and megakaryocytes: this nuclear protein has proven useful in identifying vascular neoplasm including EHE, showing a better combined sensitivity and specificity than the endothelial markers CD31 and CD34. CD34 is reported to be expressed by more than 90% of vascular tumors, so this marker has poor specificity as a variety of soft tissue tumors also express it. In contrast, CD31 is regarded as a relatively specific vascular tumor marker.

INVESTIGATIONS: Blood counts and biochemical profiles are usually normal. The radiology examination with MRI are necessary to obtain morphological data: the degree of the neoformation, the relations with the surrounding tissues, a potential cleavage plan(Gherman and Fodor,2011).

TREATMENT: Because of its rarity, EHE has no standard for treatment and actually few therapeutic options are available. If the proposed association of *Bartonella* spp. infections were confirmed, it would be plausible that eradicating the bacterial infection or interrupting *Bartonella*-induced angiogenic and proliferative cell signals could slow the tumor progression and improve patient outcomes(Gherman and Fodor,2011).

Many cases of hemangioendothelioma are asymptomatic and are treated with a 'wait and see' approach. It is sometimes considered prudent to wait until the hemangioendothelioma declares its intentions, before starting to fight back. However, there are times when treatment may be indicated.

PROGNOSIS: Though there is little experience with epithelioid sarcoma-like hemangioendothelioma, it appears to have in a relatively indolent fashion. Local recurrence and soft tissue metastasis has been reported but not lymph node or distant metastasis. No tumor related deaths have been reported to date, except for lung tumours resulting in respiratory failure(Mascarelli et al., 2011).

4.CONCLUSION:

If you have isolated disease of hemangioendothelioma, ie a tumor in one place where it can be removed, then surgery to remove the one tumor can be the best chance for a cure and this is almost certainly the way to go.

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