Primary Angiosarcoma of Humerus – A Case Report and Literature Review

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Learning Point of the Article:
Being a rare vascular sarcoma of osseous origin, the article illustrates the various histopathological and immunohistochemical examination findings that aid in the diagnosis of the condition and also highlights the appropriate management of the tumour.

Abstract

Introduction: Osseous angiosarcoma is a very rare tumor of bone with aggressive behavior, propensity for recurrences, and distant metastasis. The etiology of osseous angiosarcoma is uncertain; however, specific risk factors have been recognized. The diagnosis of angiosarcoma of bone demands multimodality imaging in conjunction with histopathological and vascular marker evaluation to aptly differentiate them from other vascular tumors. Treatment of osseous angiosarcoma remains controversial.

Case Report: A 53-year-old male presented with pain and swelling of the right upper 1/3rd of the arm following heaviness while lifting weight for 3 months. He had a history of significant weight loss and appetite with no history of inciting trauma or irradiation in the past. On examination, a diffuse swelling was noted in the right shoulder and right scapular aspect with varied consistency and ill-defined borders and margins. The skin over the swelling was stretched and shiny with dilated engorged veins over it. The plain radiograph of the right shoulder with humerus revealed a large expansile lytic soft-tissue mass in the right proximal humerus with a wide zone of transition without sclerotic margins. Magnetic resonance imaging showed T1 hypointense, T2/PDFF hyperintense large well-defined expansile lytic lesion with multi-located cysts, and multiple blood-fluid levels involving right proximal humerus. The patient underwent an incisonal biopsy which exhibited angiosarcoma of the humerus. The patient was treated with six cycles of chemotherapy with a mesna, doxorubicin, ifosfamide, and dacarbazine regimen. The patient was still under follow-up.

Conclusion: Being a rare clinical entity, controversy exists in angiosarcoma of humerus regarding its etiology and recommended management protocols. Histopathology and immunohistochemistry remain the gold standard in differentiating osseous angiosarcoma from other osseous vascular tumors. The treatment protocol has to be standardized to decrease morbidity and to improve the functional quality of life of the patient.

Keywords: Angiosarcoma, humerus, chemotherapy.

Introduction
Primary angiosarcoma of bone is an extremely rare neoplasm of mesenchymal origin. The tumor biology exhibits an aggressive natural course with rapid malignant transformation and marked anaplasia. [1] Osseous angiosarcoma has a propensity for recurrences and distant metastasis. At a glance, about 6% of all angiosarcomas develop in the bone [2, 3]. The etiology of osseous angiosarcoma is uncertain; however, specific risk factors have been recognized [4]. The diagnosis of angiosarcoma of bone demands multimodality imaging in conjunction with histopathological and vascular marker evaluation to aptly differentiate them from hemangioma, metastatic carcinoma, renal cell carcinoma, and bacillary angiomatosis [5, 6, –7]. However, treatment is quite challenging and involves multi-modality management options such as chemotherapy, neoadjuvant chemotherapy, surgical resection, and radiotherapy [3, 8]. Here, in this article, we report a case of angiosarcoma of the humerus bone in a 53-year-old male and provide a review of the literature on the same account to outline the importance of clinico-radio-histological assessment of the patient for prompt diagnosis and management of such deceptive neoplasm.
A 53-year-old man came with pain and swelling of the right upper third arm following heaviness while lifting weight for 3 months. The pain was sudden in onset, dull aching, radiating to the scapular aspect without any aggravating or relieving factors. The swelling appeared to be a lemon-sized one which gradually progressed in the last past 3 months. The patient had associated paraesthesia, numbness, and restricted mobility of the shoulder. The patient reported a history of significant weight loss and appetite. There was no history of inciting trauma or irradiation in the past. On examination, a diffuse swelling was noted in the right shoulder and right scapular aspect measuring approximately 15 cm × 10 cm × 7 cm. Swelling showed varied consistency with ill-defined borders & and margins. The skin over the swelling was stretched and shiny with dilated engorged veins over it. No bruit was felt over the swelling.

The metabolic profile revealed a 10-fold rise in serum alkaline phosphatase and significant elevation of lactate dehydrogenase. Plain radiograph of the right shoulder with humerus revealed a large expansile lytic soft-tissue mass in the right proximal humerus involving both cortex and medulla with a wide zone of transition without sclerotic margins (as shown in Fig. 1a, 1b). Non-enhanced computed tomography (CT) NECT scan of the right shoulder (coronal and axial sections) revealed a large expansile lytic soft-tissue density mass lesion measuring 10.8 cm × 7.8 cm × 9.2 cm at right upper 1/3rd of the humerus and showing loss of fat planes with right subscapularis, supraspinatus, and abutting right deltoid and pectoralis major muscle (as shown in Fig. 2a, 2b). Magnetic resonance imaging (MRI) showed T1 hypointense, T2/PDFS hyperintense large well-defined expansile lytic lesion measuring 10.8 cm × 7.9 cm × 9.2 cm with multi-loculated cysts and multiple blood-fluid levels involving right proximal humerus infiltrating right subscapularis and supraspinatus muscle with hyperintensities in infraspinatus and teres minor muscles (as shown in Fig. 3a to 3e). Tc-99m methylene diphenylphosphonate (MDP) bone scan revealed abnormal tracer uptake in the right upper 1/3rd of humerus and scapula. No metastatic lesions were demonstrated with fluorine-18-fluorodeoxyglucose positron emission tomography (PET) imaging.

The radiological evidence with the clustering of multifocal lesions in a single anatomic region throws a high index of suspicion towards osseous vascular tumors. It is highly questionable to differentiate between angiosarcoma, hemangiopericytoma, and hemangioendothelioma. Angiosarcoma may be more destructive, but cellular differentiation is not reliable. The possible differential diagnosis was bone marrow metastases, multiple myeloma, osseous angiosarcoma, osseous hemangiopericytoma, and osseous hemangioendothelioma.

With the suspicion of a malignant tumor, the patient was subjected to an incisional biopsy. Grossly, the tumourigenic tissue was friable, tan red, and hemorrhagic with few areas of necrosis. Histopathologically, the tumor showed cytologically malignant cells that were epithelioid in appearance. The nuclei were vesicular and contain one or two small nucleoli and occasional macronuclei. The cytoplasm was scant to moderate, deeply eosinophilic, hold intact, and fragmented erythrocytes. Areas of vasofrization were also seen along with numerous mitotic figures. The tumor cells were arranged in solid sheets and also line the irregular vascular lumina. Along with extravasated erythrocytes scattered deposits of hemosiderin were present. A variable inflammatory infiltrates consisting of lymphocytes and neutrophils were also seen (as shown in Fig. 4).

Immunohistochemical analysis demonstrated von Willebrand factor, Vimentin, CD-31 (as shown in Fig. 5) and -34, factor 8 RA positivity which confirmed the tumor of vascular origin, and hence a confirmed diagnosis of primary angiosarcoma of osseous origin (humerus) was made.

Our patient was treated with 6 cycles (one cycle per /month) of palliative chemotherapeutic agents according to mesna, doxorubicin, ifosfamide, and dacarbazine (MAID) protocol (mesna, doxorubicin, ifosfamide, and dacarbazine) and was followed up for 6 months after the last cycle of chemotherapy. The patient presented with a decreased in the size of the mass.
Angiosarcomas are very rare, high-grade malignant tumors with a spectrum comprising hemangiomas, hemangiendotheliomas, well or poorly differentiated angiosarcomas [9]. The cell of origin in angiosarcoma is endothelial cell either vascular or lymphatic origin. It represents 1% of all soft-tissue sarcomas [5]. Angiosarcoma occurs most predominantly in the cutaneous region followed by deep tissues such as the liver, breast, bone, and spleen [5]. They exhibit hematogenous spread with the lungs being the most common site for metastasis, followed by the liver, bones, soft tissues, and lymph nodes. The pathological diagnosis of angiosarcoma depends on the grading of tumorigenic cells either of high or low grade [10]. The overall survival rate ranges from 6 to 16 months [5]. The prognosis depends on the local recurrence and distant metastasis.

The knowledge of osseous angiosarcoma on its biology and behavior are is meager in the literature because of its very rare occurrence. Osseous angiosarcoma accounts for less than <1% of all primary malignant bone tumors with the worst prognosis [3]. Angiosarcoma of bone has slight male preponderance (M:F = 2:1) with affection towards 3rd third to 5th fifth decades of life [3, 5, 11]. Most commonly it occurs in tubular long bones (60%) [(tibia [(23%)], femur [(18%)], and humerus [(13%)])], followed by axial skeleton (40%) [6]. Osseous angiosarcoma can be divided into primary being idiopathic and secondary being due to prior irradiation, lymphoedema, Paget’s disease of bone, and bony infarction [12, 13]. The two most commonly reported symptoms are pain and swelling at the local site. Vermaat et al. stated that up to 30% of malignant vascular tumors are multifocal [14]. Humeral primary angiosarcoma is a very rare tumorigenic entity and the review of the published such cases have been tabulated in (Table 1).

On conventional radiography, osseous angiosarcoma exhibits variable presentations of the lytic lesion with cortical destruction, without sclerosis. Tumors show osseous expansile and eccentric lesions without a periosteal reaction. An expansile bony lesion is observed in low-grade osseous angiosarcoma whereas cortical permeation, a geographical lytic lesion with soft- tissue infiltrations is observed in high-grade aggressive osseous angiosarcoma. Angiosarcoma of bone rarely shows calcifications. Lv et al. explained the aggressiveness of angiosarcoma in the lower extremity as an osteolytic destructive lesion with an ill-defined cortico-medullary junction [1]. In our case, a large expansile lytic soft-tissue mass in the right proximal humerus involving both cortex and medulla with a wide zone of transition with decreased pain over the right humerus at the end of 1 year. The patient was still under follow-up.

CT stands superior in evaluating the bony destruction than conventional radiographs. CT serves as a useful diagnostic tool in terms of sensitivity for lesion detection and provides a precise anatomic extent of the disease [20]. MRI is considered as the gold standard diagnostic modality in demonstrating the invasiveness of bone and soft-tissue components and assists in the staging of the tumor. MRI offers tissue characterization [4]. Lv et al. observed mottled, the increased signal on T1- weighted MRI. They observed enhanced MRI demonstrated the infiltration level of the lesion [1]. Griffith et al. observed hypointensity in T1W images within the humerus where it contains numerous serpiginous vascular channels. These vascular channels demonstrated fluid-filled levels on T2W images. On administration of contrast medium, the lesion exhibited a heterogeneous enhancement into soft-tissue component [21]. Our case showed hypointensity on T1W images and hyperintensity on T2W/PDFS images with large well-defined expansile lytic lesion with multi-loculated cysts and multiple blood-fluid levels involving right proximal humerus infiltrating right subscapularis and supraspinatus muscle with hyperintensities in infraspinatus and teres minor muscles. Osseous scintigraphy with 99mTc MDP show increased uptake by the tumor cells and helps in assessing the multifocality of the tumor [22].

Osseous angiosarcoma must be differentiated from osseous haemangiopericytoma and osseous haemangiendothelioma [23]. Hence, tissue diagnosis and immunohistochemistry are essential to confirm the diagnosis. The differential diagnosis for osseous angiosarcoma is tabulated in (Table 2). Angiosarcomas typically have ill-defined margins and exhibit a considerable degree of cellular atypia and architectural differentiation. They range from well-formed, anastomosing vessels to solid sheets of high-grade epithelioid or spindled cells without clear vasoformation. Most of the tumors show high-grade morphology with anastomosing vascular channels or more solid areas, variable nuclear atypia, mitotic activity, and coagulative necrosis. Sometimes atypia is mild and focal with cells resembling normal vascular endothelium. Marked pleomorphism is rare. Vasoformative areas are composed of ramifying channels lined by the atypical spindle or epithelioid cells, with variable endothelial multilayering, intraluminal budding, hobnailing, or papillary-like projections. The vascular channels may be poorly formed with complex dissecting patterns. Solid areas typically comprise cellular sheets of spindled to epithelioid cells with abundant eosinophilic to amphophlic cytoplasm, large vesicular nuclei, and prominent nucleoli. These may be associated with blood lakes or extensive hemorrhage and organizing hemATOMA. More rarely tumors appear morphologically low grade with well-formed vascular channels lined by minimally atypical spindled cells. Epithelioid angiosarcomas typically have a solid architecture, with diffuse,
### Table 1: Literature review for angiosarcoma humerus

| Author and year | Patient | Symptoms | Histopathology | Immunohistochemistry | Management |
|-----------------|---------|----------|----------------|----------------------|------------|
| Chen et al., (19) 2005 | 47 years male | Epitheloid sarcoma | Multilobular, osteolytic lesion with an anastomosing vascular channels | Diffuse keratin positive, CD 34 positive in 40% cases. Loss of nuclear INI-1 expression | Wide resection |
| Hasegawa et al., (16) 1997 | 46 years male | Pain in the right shoulder for 3 months | Idiopathic | Positive for factor VIII related antigen, CD31, Ulex Europeanus agglutinin I, c-kit, vimentin | Wide resection |
| Fukunaga et al., (17) 1999 | 55 years male | Deformity in right upper arm since 5 years of age | Polyostotic fibrous dysplasia | Diffuse keratin positive in carcinoma; vasoformative areas | Wide resection |
| Voggenreiter et al., (18) 2001 | 12 year male | Pain in the right arm for 7 months | Idiopathic | Positive for factor VIII related antigen, CD31, Ulex Europeanus agglutinin I, c-kit, vimentin | Wide resection |
| Mittal et al., (19) 2007 | 47 years male | Painful swelling over right shoulder after 10 years of radiotherapy for giant cell tumor | Osteolytic lesion with severe bone destruction in the humerus | Positive for nuclei-1 expression, CD 31 and non-Wellebead factor | Wide resection |
| Kernsida et al., (20) 2013 | 65 year female | Traumatic trauma to left shoulder | Idiopathic | Positive for CD31, CD34, and factor 8 | Wide resection and adjacent chemotherapy (6 cycles of methotrexate and ifosfamide) |
| Our case, 2020 | 53 year male | Pain and swelling of right upper arm following heaviness while lifting weight since 3 months | Idiopathic | Positive for vimentin, CD 31 and non-Wellebead factor | Wide resection |

### Table 2: Differential diagnosis of angiosarcoma on the basis of histopathology

| Parameters | Epitheloid sarcoma | Epitheloid hemangioendothelioma | Metastatic carcinoma or melanoma |
|------------|-------------------|--------------------------------|---------------------------------|
| Cellular morphology | Resembles epitheloid angiosarcoma | Cords and small nests of round epithelial endothelial cells with abundant eosinophilic cytoplasm; No vasoformative areas | Resembles epitheloid angiosarcoma; Lack vasoformative areas |
| Immunohistochemistry/mutation | Diffuse keratin positive, CD 34 positive in 40% cases. Loss of nuclear INI-1 expression | Recurrent t(1;3) involving CAMTA-1 and WWTR-1 | Diffuse keratin positive in carcinoma |

**MAID:** mesna, dexamethasone, ifosfamide, and dacarbazine.

**INH-1 expression**

**Willebead factor**

**CAMTA-1 and WWTR-1**

**Diffuse keratin positive in carcinoma**
Epitheloid angiosarcoma

- Cells with epithelioid cell-like nuclei
- Multiple large nucleoli and coarse chromatin
- Multinucleation common
- Presence of intracytoplasmic lumina

Table 3: Difference between histological variants of osseous angiosarcoma

| Spindle cell angiosarcoma | Epitheloid angiosarcoma |
|---------------------------|-------------------------|
| Atypical cells with oval or spindle hyperchromatic nuclei | Cells with epithelioid cell-like nuclei |
| Fine to coarse chromatin pattern | Multiple large nucleoli and coarse chromatin |
| Multinucleation rare | Multinucleation common |
| Vasoformative features | Vasoformative features |
| Cytoplasm with microvacuolization, especially on Diff-Quik-stained slides | Cytoplasm with microvacuolization, especially on Diff-Quik-stained slides |

Intracytoplasmic lumina absent

Presence of intracytoplasmic lumina

sheet-like patterns of large, atypical epithelioid or polygonal cells with ovoid vesicular nuclei, prominent large central nucleoli, and abundant cytoplasm [23, 24, 25]. The difference between histologic variants of osseous angiosarcoma was tabulated in (Table 3) [26]. Immunohistochemistry confirms the diagnosis of angiosarcoma with positivity for endothelial cell surface markers (CD31, CD34, factor 8 RA, Ulex europaeus agglutinin I, FLI-1, and thrombomodulin), epithelial markers (cytokeratins & and epithelial membrane antigen) Vimentin, and von Willebrand factor [27, 28].

(Tables 2): Differential diagnosis of angiosarcoma on basis of histopathology

Due to the rarity of the tumor, the management options are yet to be explored. No specific treatment options for osseous angiosarcoma have been mentioned in the available literature. Yamashita et al. recommended that surgical modality coupled with immediate post-operative radiotherapy prove to be a viable treatment option for cases where complete resection is possible [7]. Various researchers have investigated multiple combinations of chemotherapeutic agents like such as cisplatin + doxorubicin, cisplatin + paclitaxel, and cisplatin + doxorubicin + paclitaxel [29, 30, –31]. With the existing plausible literature, it has been understood that angiosarcoma responds well with a chemotherapeutic agent like anthracycline. Baliaka et al. described the distinct histological features of primary angiosarcoma of bone which were defined by malignant spectrum (mitotic figures and nuclear atypia) of osseous vascular tumors. The worst prognostic markers were more than 3 mitoses per /10 HPF, the presence of a macronucleus, and the presence of fewer than five 5 eosinophils per /10 HPF. With all these histopathological features in the osseous angiosarcoma, the 5-year survival rate is 0%. D2-40 marker lymphangiogenic differentiation displays the more aggressive behavior of the tumor [33]. Our case was still under our follow-up.

Conclusion

Being a rare clinical entity, controversy exists in angiosarcoma of humerus regarding its etiology and recommended management protocols. Histopathology and immunohisto-chemistry remain the gold standard in differentiating osseous angiosarcoma from other osseous vascular tumors. The treatment protocol has to be standardized to decrease morbidity and to improve the functional quality of life of the patient.

Clinical Message

Being a rare vascular sarcoma of osseous origin, histopathology, and immunohistochemistry examination aids in diagnosis and its appropriate management of the tumor.

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