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CASE REPORT

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Epithelioid Hemangioendothelioma of the Right Iliac Wing in a Young Adult Male: A Case Report

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Epithelioid hemangioendothelioma (EHE) of bone is a malignant vascular neoplasm with a very low global prevalence. Nonspecific clinical and histopathological findings make the diagnosis of this tumor very challenging. In this case, we reported a 23-year-old male presented with persistent right buttock pain for 9 months (VAS 2-3). No other clinical findings were found. On plain radiographs, a lytic lesion was found on the right iliac wing. MRI showed iso-hyperintense lesions on both T1- and T2-weighted sequences with hemorrhagic components. A CT guided core needle biopsy was performed for further evaluation. The patient underwent wide excision with adjuvant chemotherapy. At six months post-surgery, lymph node metastases was found on PET scan without any clinical symptoms. A comprehensive interdisciplinary evaluation is required to establish the diagnosis of EHE.

Keywords: Epithelioid hemangioendothelioma, Pelvic neoplasm, Vascular tumor, Wide excision, Lymph node metastases.

INTRODUCTION

Epithelioid hemangioendothelioma (EHE) is a very rare vascular tumor and often multifocal. The prevalence rate was below 1 per 1 million cases annually. EHE exhibits features between benign hemangioma and malignant angiosarcoma[1-3]. The neoplasm was a malignant vascular tumour with intermediate clinical behaviour and variable metastatic potential[1]. EHE mainly develops in soft tissues and rarely occurs in bones. However, in bones, this neoplastic lesion often arises from long bones of the lower extremities. The predilection of EHE is within the diaphyseal area. It could involve the metaphysis or meta-epiphyseal region as well[2]. The clinical features and histopathological results may mimic those of other high-risk vascular tumors[4]. The nonspecific findings make the diagnosis of bone EHE particularly challenging. We presented a case of iliac bone EHE, which appeared to be among the first documented cases from Indonesia. This case report aimed to highlight the unusual presentation, diagnostic challenges, and management considerations of iliac bone EHE with early pelvic lymph-node metastasis in a young adult male.

CASE PRESENTATION

A 23-year-old male presented with right buttock pain that had persisted for the past nine months. On physical examination, the patient reported mild localized discomfort in the right buttock with Visual Analog Scale (VAS) score of 2–3. Distal sensory function was preserved and no neurological deficits were observed. The range of motion in the right hip and adjacent joints was full and pain-free. These results indicate the absence of neuromuscular function and joint disorders.

Initial plain radiographs revealed a lytic lesion in the right iliac wing (Figure 1). Subsequent contrast-enhanced pelvic MRI demonstrated an iso-hyperintense lesion on both T1- and T2-weighted sequences with hemorrhagic components (Figure 2). The lesion exhibited an approximately 2.7 × 3.7 × 2.9 cm lobulated margin at the superoposterior aspect of the right iliac wing. Imaging also indicated cortical and medullary bone destruction with surrounding bone marrow edema. Blood test and chest radiograph was within normal limit. Based on these findings, a primary malignant bone tumor of the right ilium was suspected. A CT-guided core needle biopsy was performed to

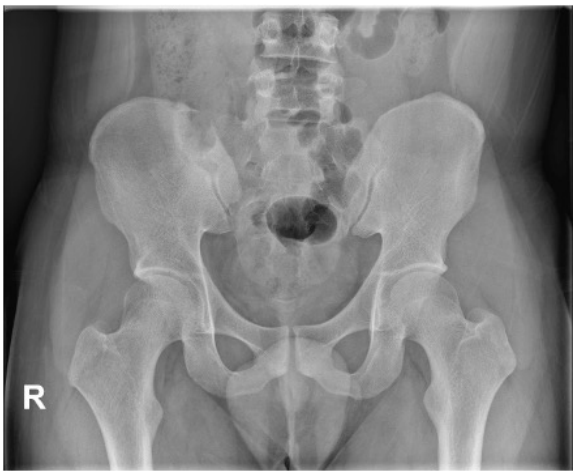

Fig. 1 — X- ray reveals lucency at right iliac wing pelvis.

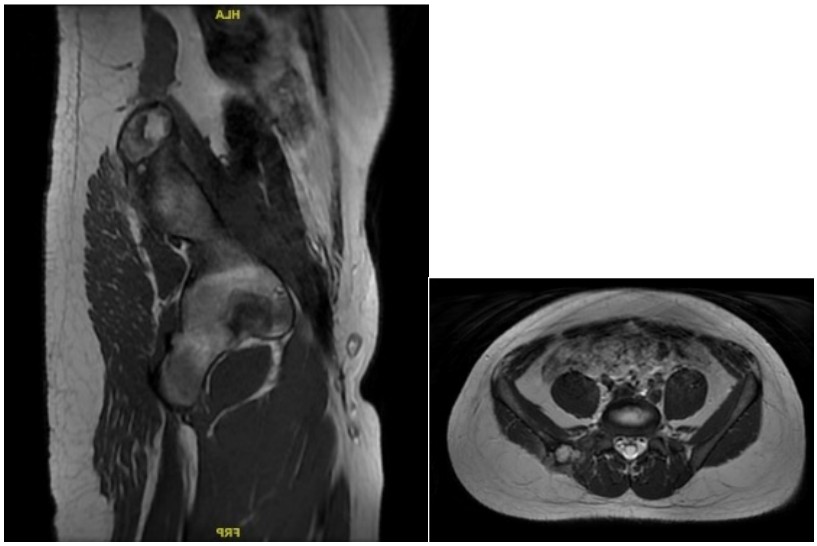

Fig. 2 — MRI pelvis with contrast demonstrated iso-hyperintense lesion on T1WI-T2WI with hemmorhagic component and lobulated margin at right superoposterior iliac wing. A: Sagittal view; B: Axial view.

obtain tissue for histopathological evaluation.

Histopathological examination demonstrated a malignant neoplasm composed of epithelioid cells with vascular differentiation. Immunohistochemical staining revealed positive expression of vascular markers CD31, CD34, ERG, and FLI1, supporting endothelial lineage (Figure 3). AE1/3 showed focal positivity, while other epithelial, muscle, lymphoid, and neural markers were negative. These findings confirmed the diagnosis of EHE.

The patient underwent wide excision of the lesion, including partial resection of the right iliac wing without subsequent bony reconstruction (Figure 4). At three months follow-up, the patient remained asymptomatic. But, later at six-months after surgery, Positron Emission Tomography (PET) scan demonstrated common iliac lymph node mass at pelvic region, presumed to reflect regional pelvic lymph-node metastasis. PET scan was performed later after the surgery due to financial and health insurance problems. The patient achieved a lower-limb MSTS score of 27 and was able to ambulate without the need for walking aids. The patient was initiated on six cycles of adjuvant chemotherapy postoperatively, consisting of: ifosfamide 2500 mg/m2 for 4 days, mesna 500 mg/m2 for 3 doses per day for 4 days, and doxorubicin 25 mg/m2 for 3 days. The present study complies with the 2013 Care Checklist guidelines[5].

DISCUSSION

The term EHE was initially introduced by Weiss and Enzinger in 1982. EHE in bone is very rare, accounting for around 1% of the prevalence of EHE among all

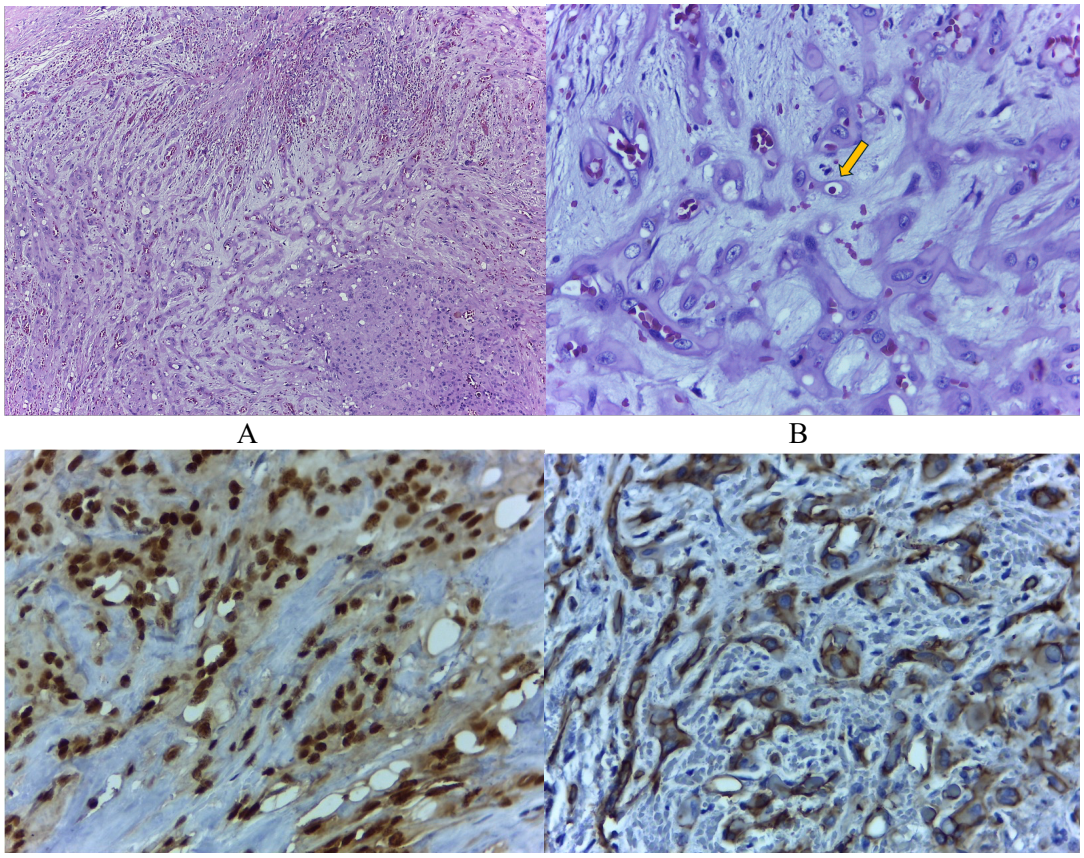

Fig. 3 — Hematoxylin-eosin stain showing epithelioid .cells arranged in cords, aggregates or nests with pale to densely eosinophilic cytoplasm and intracytoplasmic vacuoles/blister cells, (×100); B: Intracytoplasmic vacuoles (blister cells) contain erythrocytes (arrow) and myxoid to hyaline stromal matrix (H&E,×400); C & D: Diffuse nuclear reactivity for ERG and membranous staining for CD31 (400x)

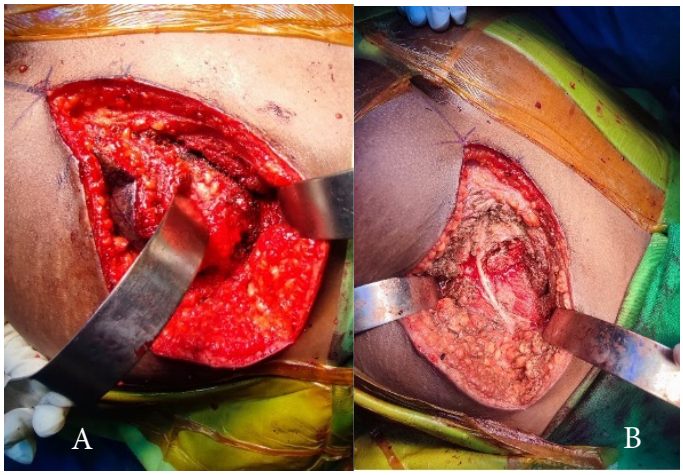

Fig. 4 — A & B Wide excision without any reconstruction of the bone.

vascular bone neoplasms. Witte et al. conducted a study examining five cases of EHE. Three of them had lesions in the lung and thoracic regions, yet no bone lesions were observed[6]. EHE predominantly affects men aged 20 to 30, although individuals of any age may be at risk of developing EHE. The symptoms reported by patients with EHE are nonspecific. Typically, patients experience pain and swelling in the affected region[7]. In our case, we reported a patient presenting with mild persistent pain in the right buttock region with no other inflammatory symptoms. There were no typical malignancy complaints of fever, weight loss, or systemic symptoms.

A soft tissue mass and lytic lesion of bone destruction

with cortical destruction could be observed on plain radiography. A lesion could either be well-defined or inadequately marginated[8]. The MRI demonstrates iso intensity on T1-weighted imaging, hyperintensity on T2-weighted imaging, and significant enhancement on contrast-enhanced images[9]. In evaluating metastasis, 18F-FDG-PET/CT is particularly important[10]. We initially suspected that our patient had a round cell tumor with metastatic bone disease (MBD) as a differential diagnosis based on the plain radiological image and atypical clinical presentation. The presence of bone damage and adjacent bone marrow edema led us to hypothesize that the lesion was a primary bone malignancy. A histopathology test was needed to confirm the diagnosis. Biopsy results typically exhibit extensive eosinophilic cytoplasm, moderate nuclear pleomorphism, and well-formed thick-walled vessels that are lined by epithelioid cells. Tumor cells show positive staining for vimentin, Factor VIII, CD31, and CD34, accompanied by infrequent mitotic activity[11].

Our patient presented with a localized lesion. Therefore, surgery was chosen as the primary treatment option as surgical resection remains a preferred approach in unifocal localized disease[12]. Reconstruction was not always required for all primary pelvic tumours. As peripheral iliac involvement typically does not compromise pelvic stability, opting against reconstruction can further reduce the associated morbidity[13]. Early intervention has also been linked to a better prognosis[14]. In our case, the surgery was successful and did not result in any significant complications. The patient's complaints improved postoperatively and no observed disruption in daily function at the six-month follow-up. In the case series reported by Kamal et al., surgical resection of malignant pelvic tumors yielded moderate functional outcomes (mean MSTS score of 16.5). Inferior outcomes were noted in cases involving more extensive resections and larger tumour sizes (>20 cm)[15]. The role of active surveillance in asymptomatic cases should not be underestimated before initiating treatment. European Society for Medical Oncology (ESMO) had released a consensus statement supporting the option of active surveillance in regional and systemic metastatic cases[16].

In a study by Frezza et al., anthracyclines, paclitaxel and pazopanib showed limited efficacy with median overall survival (OS) of 14.3 months, 18.6 months, and 8.5 months, respectively[17]. Meanwhile, Kyriazoglou et al. study demonstrated stable disease (SD) results after completing six treatment cycles of doxorubicin and olaratumab combination in two metastatic hepatic EHE cases[18]. There is a lack of clear evidence supporting the benefits of chemotherapy. However, chemotherapy may be administered in metastatic and multifocal cases to stabilize the disease[19-21]. Our patient was given a combination of ifosfamide, mesna, and doxorubicin following PET scan evaluation of lymph node metastases.

Radiotherapy might be beneficial as a primary treatment when surgery is not feasible. To reduce the risk of local recurrence, radiotherapy can be addressed as adjuvant therapy post-surgical intervention[22]. Both chemotherapy and radiotherapy may as well be administered postoperatively to mitigate complications arising from incomplete resection[23].

Epithelioid hemangioendothelioma cases in bones are considered as very rare. As far as we are aware, there was only one other study that has previously reported the occurrence of EHE in Indonesia, in which, specifically addressed EHE in the hand area in pediatric cases[24]. In this case, the patient was diagnosed with EHE and the diagnosis was established through multidisciplinary collaboration. Although, EHE has a relatively lesser ability to spread. We assumed that the lymph node lesion in our case originated from the primary EHE lesion that metastasized.

CONCLUSION

Epithelioid hemangioendothelioma of the bone is a rare occurrence. However, the combination of physical examination, radiological evaluation, and histopathological analysis is crucial for a proper diagnosis. Differentiating between benign and malignant bone tumors is crucial. Hence, immunohistochemistry testing can be utilized to assist in the diagnosis of EHE and to exclude other primary bone tumors.

Limitations

The Ki-67 index and molecular testing for CAMTA1/TFE3 were not performed in this case. The duration of follow-up was limited to only six months. Long-term observational data could not be obtained. Lymph-node metastasis in the present case was identified solely through radiological imaging, as biopsy confirmation was not available.

Patient consent: Written informed consent covering all procedures and subsequent publication was obtained from the patient, following the regulatory and ethical guidelines of Fatmawati Central General Hospital.

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Conflict of interest: The authors reported no conflict of interest.

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