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Oncology

"A STUDY OF VASCULAR TUMORS OF SHORT BONES"

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ABSTRACT

Background: Epithelioid Hemangioendothelioma of bone is a rare tumor of vascular origin with borderline malignancy and pursues a clinical course intermediate between hemangioma and an angiosarcoma.

Methods: Epithelioid Hemangioendothelioma is reported more commonly in soft tissues and viscera involving lungs and liver but less so in bones. The tumor is usually multicentric with a predilection for bones of a particular anatomical area. We describe four cases of multicentric Epithelioid Hemangioendothelioma involving the metacarpals and phalanges. Histologically the tumor exhibited cords of epithelioid-appearing cells in a myxoid stroma and inflammatory cells with few mitotic figures.

Results: The patients were treated with primary surgical resection, curettage, bone grafting and K-wire fixation. Follow-up x-rays revealed good consolidation of bone graft with no signs of local recurrence.

Conclusion: Primary surgical resection, curettage and if required bone grafting and K-wire fixation is a good method to manage these vascular tumors.

KEYWORDS : Bone tumors, hemangioendothelioma, multicentric lytic tumors.

I. Introduction

Epithelioid Hemangioendothelioma is a rare tumor of vascular origin with borderline malignancy. The term was first applied in 1982 by Weiss and Enzinger1. Very few cases have been reported in the literature since then. The tumor is usually multicentric and its occurrence in ilium, skull, vertebra and femur is well-documented. It has a protracted clinical course with potential for local recurrence and biologic capacity to metastasize. Here we present a series of such rare cases of epithelioid hemangioendothelioma of short bones with multicentric involvement which were initially diagnosed as multiple enchondroma due to its clinical and radiological resemblance and was identified only after resection and histopathological examination.

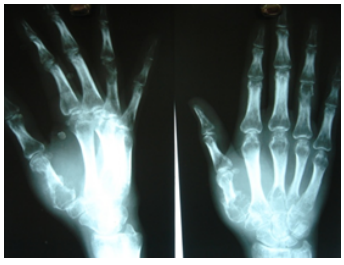
II. Materials And Methods

This is a prospective clinical study of four rare patients who presented to us with multiple swelling in the hand and foot region. All patients did not have a history of significant trauma. They presented with pain and swelling in the hand and foot. Clinical examination revealed swelling over the thumb and metacarpal region in hand and dorsal aspect of the foot and that the swelling was bony. Movements of the fingers and foot were restricted because of severe pain. Weight bearing was not possible the patients involving foot. One patient had history of diabetes and was on anti-diabetic therapy with oral hypoglycemics for more than eight years.

Another apparently healthy 41 year male presented with complaints of pain and multiple swellings in his right hand for two months. Pain was progressive and dull aching type. Swelling was of gradual onset and increasing in size for the last one month. It interfered with his daily activities as he was unable to hold and lift weight. There was no history of trauma, constitutional symptoms or pain in any other bones or joints. Patient gave no history suggestive of tuberculosis. General examination was normal with no lymphadenopathy or signs of hemangioma over skin. Examination of right hand revealed swelling over base of the thumb, dorsum of hand and proximal part of ring finger with overlying skin shiny and stretched. The swelling was tender, warm, diffuse, hard and arising from underlying bone. Movements of the thumb and ring finger were painful. There were no signs of neurovascular deficits in the hand.

RADIOGRAPHS: Plain radiographs revealed multiple osteolytic lesions in 1st metacarpal, base of 3rd metacarpal and proximal phalanx of ring finger with periarticular osteoporosis. The cortex was expansile and thinned out but intact with impending pathological fracture of the 1st metacarpal.

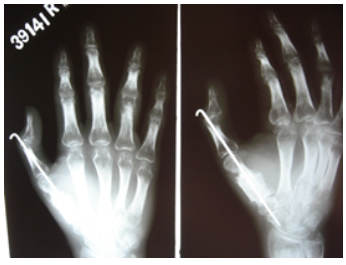
Figure 1: Pre-operative X-ray



All hematological and urine investigations were normal. Echo showed mild MVP without MR. FNAC from the swelling at the base of thumb was reported as enchondromatosis with advice for biopsy.

MANAGEMENT: A provisional diagnosis of multiple enchondro matosis was made and surgical excision was decided. Through dorsal approach, all the three lesions were resected and curettage was done. Autogenous corticocancellous bone graft harvested from left iliac crest was used to fill the defect and K-wire fixation was done to stabilize the 1st metacarpal. Post operatively the hand was immobilized in functional position.

Figure 2: Post-operative X-ray



FOLLOW-UP: The patient was then screened for similar lesions with x-ray chest, pelvis, left hand and sonography of abdomen and pelvis which did not detect any abnormality. Post operatively the patient had an uneventful recovery and follow-up x-rays revealed good consolidation of the bone graft. At 3-month follow-up the patient had a good functional recovery without any complaint and with no signs of local recurrence.

III. Results

The demographic profile of the patients is depicted in **Table 1.** Mean age of the patients was 40 years. 75% of patients were male and 25% were female. **Table 2** shows the site and side of involvement. All the patients had clinically good results at follow up of twenty weeks. All

the patients had pain till twelve to sixteen weeks.

Table 1: Demographic profile of patients

Age (yrs)	Male (%)	Female (%)	Total (%)
30-40	02 (50%)	00 (00%)	02 (50%)
40-50	01 (25%)	01 (25%)	02 (50%)
Total	03 (75%)	01 (25%)	04 (100%)

Table 2: Site and Side of involvement

Site affected	Right (%)	Left (%)	Total (%)
Hand	02 (50%)	01 (25%)	03 (75%)
Foot	01 (25%)	00 (00%)	01 (25%)
Total	03 (75%)	01 (25%)	04 (100%)

Histopathological examination of the resected tissue exhibited thin trabeculae with cords and nests of plump epithelioid-appearing cells around blood vessels. The cells embedded in a myxo-hyaline stroma possessed abundant acidophilic cytoplasm with characteristic intracytoplasmic vacuoles and a large vesicular nucleus. Focal infiltration of inflammatory cells with many eosinophils and few mitotic figures were noted. Histology was consistent with diagnosis of epithelioid hemangioendothelioma of bone.

Complications: One patient(25%) with diabetes had wound healing complication. One patient(25%) developed pathological fracture which was treated by K-wire fixation after open reduction and bone grafting.

IV. Discussion

The term "Epithelioid Hemangioendothelioma" was first used in 1982 by Weiss and Enzinger1 to a soft tissue vascular tumor of borderline malignancy pursuing a clinical course intermediate between hemangioma and an angiosarcoma. These tumors are well recognized in soft tissues, lungs[2], liver[3], skin but less so in bone. In the bone, it was first reported by Tsuneyoshi[4] in 1986 and involvement of the bones like ilium5, vertebra, skull and femur[6] have been well-documented. One of its variants, the epithelioid and spindle cell hemangioma is described to occur in small bones of hands and feet[7].

Epithelioid endothelial cell tumors have been a source of controversy because of their unusual morphology, poorly understood histogenesis and unpredictable biologic behaviour. Currently these tumors are classified by WHO as – epithelioid hemangioma (angiolymphoid hyperplasia with eosinophilia), epithelioid hemangioendothelioma and epithelioid angiosarcoma[8]. Epithelioid hemangioendothelioma of bone usually presents as pain in the involved region, and gradually increasing swelling with pathological fracture may be seen in few cases. 50% of tumors are multicentric with a predilection for bones of a particular anatomical area[9]. Our patient had this classical presentation.

Radiological findings of lytic lesion with thinning of cortex are similar as seen in cartilaginous tumors like enchondroma. Histopathological examination of the tumor clinches the diagnosis. Epithelioid endothelial cells possessing intracytoplasmic vacuoles and abundant eosinophlic cytoplasm are arranged in cords and form rudimentary vascular spaces within a myxoid stroma. Ultrastructure reveals specific Weibel-Palade bodies. Immunohistochemical reactivity to Factor VIII-related antigen supports the vascular nature of these neoplasms10. Q-bend 10 (CD 34) i.e., hematopoietic progenitor cell antigen detected in endothelial cell is positive.

Surgical resection is the primary treatment for these tumors and radiotherapy may be useful for the surgically inaccessible ones. In few cases intensive combination chemotherapy[4,5] (with vincristine, adriamycin, cyclophosphamide and etoposide) was tried but proved ineffective to halt the progression of the disease. Metastasis if occurs is usually to the lungs and liver where it destroys the involved organ, resulting in death by respiratory compromise and hepatic failure respectively. Mortality is 35% after liver metastasis and 65% in lung metastasis[4].

Conclusion

The clinical course and outcome of epithelioid hemangioendothelioma varies greatly and may not be predicted by its histological features since few reports demonstrate the aggressive behaviour and metastatic potential of histologically low-grade tumor[4]. Local recurrence is possible. Hence a thorough clinical and radiological evaluation of all patients with epithelioid hemangioendothelioma of bone is advocated at presentation and follow-up to screen for local recurrence and metastatic disease.

REFERENCES

1. Weiss SW, Enzinger FM. Epithelioid Hemangioendothelioma: a vascular tumor often mistaken for a carcinoma. Cancer 1982;50:970-981.
2. Dail DH et.al. Intravascular, bronchiolar and alveolar tumor of the lung. Cancer 1983; 51:452-464.
3. Ishak KG et.al. Epithelioid Hemangioendothelioma of liver. Hum Pathol 1984;15:839-842.
4. Tsuneyoshi M, Dorfman HD, Bauer TW: Epithelioid Hemangioendothelioma of bone. Am J Surg Pathol 1986;10:754-764.
5. Bollinger et.al. Epithelioid Hemangioendothelioma with multiple site involvement. Cancer 1994;73:610-615.
6. Evans et.al. Vascular tumors of bone. Relationship of hemangioendothelioma of bone, epithelioid hemangioma, epithelioid hemangioendothelioma and high-grade angiosarcoma. Hum Pathol 2003;34:680-689.
7. Keel SB and Rosenberg AE. "Hemorrhagic Epithelioid and Spindle cell Hemangioma. A newly recognized unique vascular tumor of bone". Cancer 1999;85:1966-1972.
8. Schajowicz F, Ackerman LV, Sissons HA. Histologic typing of bone tumors. Geneva: World Health Organization, 1993.
9. Kleer CG, Unni KK, McLeod RA; Epithelioid Hemangioendothelioma of bone. Am J Surg Pathol 1996;20:1301-1311.
10. Weiss SW et.al. Epithelioid Hemangioendothelioma and related lesions. Semin Diagn Pathol 1986;3:259-287.